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Peer review is the heart of scientific publication. The Editor wishes to place on record the contributions of the following VIMS Faculty who have provided their time for peer review of the submissions :

Dr. Pranamita Ray (Associate Professor, Dept. of Pathology)

Dr. Debjani Sinha Ray (Assistant Professor, Dept. of Radiology)

Dr. Suman Das (Visiting Surgeon, Maxillofacial Unit).

Dr. Saikat Sengupta (Senior Consultant, Dept. of Anaesthesiology, Perioperative Medicine & Pain, Apollo Multispecialty Hospitals, Kolkata)

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Editorial

Climate Change and Healthcare

Climate change is one of the greatest health threats of the 21st century. At the COP27 talks, the Government of India committed to reaching a net zero carbon emission economy by 2070, reducing greenhouse gas emissions 45% by 2030 and achieving 50% energy from renewables by 2030. The health sector has a considerable carbon footprint, generating between 8% to 10% of all greenhouse emissions in the USA. In the UK it is estimated that the National Health Service generates about 25% of all public sector emissions.

The chief source of greenhouse gas emissions in healthcare is, not surprisingly, generated by surgical units and operating theatres. Surgery is a resource intensive activity utilising expensive equipment. Sterilisation procedures, advanced technology and life support systems use a large amount of energy and consumables, with the production of copious waste.

All anaesthetic gases are greenhouse gases, and minimally metabolised. A study of several large North American healthcare facilities (Macneill, Lillywhite & Brown : The impact of surgery on global climate : a carbon footprinting study of operating theatres in three health systems. Lancet Planet Health 2017;1:e381–e388) estimated that the use of desflurane for anaesthesia contributed 51% to 63% of the total carbon footprint. The use of alternative such as sevoflurane or isoflurane, regional and total intravenous anaesthesia can significantly reduce theatre emissions. Heating, ventilation and air conditioning systems (HVAC) account for 90% to 99% of theatre energy use. There is scope for considerable savings by simply switching off overnight all but the emergency theatres.

There are no published studies on the carbon footprint of my specialty, Otolaryngology/Head Neck surgery, although there is a paper from Chile looking at the carbon footprint of a plastic surgery unit there performing Rhinoplasty. One would imagine that the greenhouse impact of surgery in India is less than that in the West, and there is a paper comparing phaco cataract surgery in an Indian hospital with that in a UK hospital. The Indian procedure generated 6kg CO_2 equivalents compared with 180kg CO_2 equivalents for the UK procedure.

Healthcare consumable manufacturing is another source of emissions. Eckelman M et al (Comparative life cycle assessment of disposable and reusable laryngeal mask airways. Anesth Analg 2012; 114: 1067–72) calculated the carbon footprint of a single use laryngeal mask airway (LMA) to be 11.3kg CO_2 equivalents, compared to 7.4kg CO_2 equivalents for a reusable LMA. However, reusable equipment may not always have a smaller carbon footprint.

At our individual levels we should be constantly challenging ourselves – do we really need to fully gown for all procedures? Do we always need to use disposable gowns? Are there good locally produced consumables which may be used in place of imported ones? One simple measure is to reduce patient follow ups – if all we need is to find out if the patient is feeling better or not, is it feasible to follow up by telephone?

Climate change ultimately affects the health of everyone. As physicians we have a duty to improve public health. As part of a sector which contributes significantly to climate change we have a responsibility to minimise that impact as much as possible.

A common question I am asked is whether the Journal is indexed. At present the answer to that question is, in a word, No. While we are a peer reviewed journal, we are not registered with any of the indexing agencies. In this respect you will be happy to know that the decision has been taken to index the Journal with The Directorate of Open Access Journals (DOAJ). This is recognised by the National Medical Commission, hence once it is achieved, authors may cite the Journal papers for meeting faculty promotion norms.

DOAJ recognition requires certain changes. The first, visible, change you will see is in the composition of the Editorial Board. We welcome Professor Atul Kumar Gupta, Principal, Ramakrishna Mission Seva Pratishthan Vivekananda Institute of Medical Sciences, as our new Editor in Chief. The Editorial Board has been recast, and I am grateful for the support of the new members, namely : Professor Sanjay Mohan Bhattacharjee, Professor Sukanta Misra, Professor Sudip Chatterjee and Professor Soumitra Kumar.

The Journal will move to its own website, and the individual papers will be available as digital files. As an Open Access journal we hope to attract submissions from all specialties, so please encourage your colleagues and juniors to contribute.

As I write this editorial the Hospital is gearing up for the Annual Scientific Conference: the abstracts will appear in our next issue.

Ranjan Raychowdhury, Executive Editor; Professor, Department of ENT & Head Neck Surgery, RKMSP, VIMS

Original Article

Apnoeic Oxygenation for Paediatric Emergency Intubation – Historic Review and Evidence for Paediatric Practise

Guruprasad Hassan Shankar, Srilakshmi Hassan Shankar

Abstract :

Apnoeic oxygenation, which is the process of providing oxygen supplementation during the period of apnoea when intubation is ongoing, is slowly emerging as a standard of care adopted by anaesthetists, intensivists and emergency physicians during elective or emergency intubations. While evidence in adult population is robust, paediatric work is ongoing and has begun to demonstrate similar benefits. Safety of the technique has been adequately established while more research is needed to clarify the pros and cons of different delivery methods and flow rates. Though the principle of apnoeic oxygenation and technique itself have been known for long, with a rich historical background deeply connected with development of mechanical ventilation itself, challenges remain in the incorporation of the relatively new practise across units. In this paper, we review the historical background, physiology and evidence for practise in the specified context of paediatric emergency intubation. We briefly discuss the practical aspects of the implementation of this useful practise for improving the safety of paediatric emergency intubations in routine care.

Key words :

Apnoeic oxygenation; paediatric emergency intubation

Introduction :

"Primum non nocere". First, do no harm, is a doctrine as old as medicine itself.[1] Actively anticipating and preventing harm is a vital part of any disciplined approach to the practise of medicine. Tracheal intubation is a very often performed intervention in emergency rooms and intensive care units. However, it is associated with a substantial risk of hypoxemia (48% of difficult and 15% of non-difficult intubations in paediatric intensive care unit (PICU)[2]). Hypoxemia during the process of intubation contributes significantly to adverse events including dysrhythmias, hypoxic brain injury, seizures, surgical airway, or cardiac arrest with profound hypoxemia being defined as oxygen saturation of < 70%.[3] While the process of pre-oxygenation prior to endotracheal intubation is standardised, research is ongoing about additional methods to increase the safety of and prevent critical events during intubation. One of these techniques involves providing oxygen supplementation even as the process of intubation is ongoing. This method, called apnoeic oxygenation (AO), is the process by which oxygen moves by mass flow through the upper airways into the alveoli in the absence of any respiratory effort, consisting of administration of oxygen during apnoeic period of intubation to extend the safe apnoea time.[4] Apnoeic oxygenation during intubation may be particularly beneficial in patients who are at risk of rapid desaturation, and in patients for whom airway management may be difficult or prolonged.

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Apnoeic oxygenation : historical aspects

The history of apnoeic oxygenation preceded the development of anaesthesia itself. Prior to the safe administration of anaesthetic agents thereby causing apnoea, it was important to establish that it is possible to keep animals and then human beings alive by artificial oxygenation as well as artificial ventilation. This fascinating story begins in 1666, in a published account of the Philosophical Transactions of the Royal Society of London,[5] of an experiment made by M. Hook, of keeping alive animals by blowing into the lungs using bellows. Since it was then believed that "the motion of the lungs is necessary to life, upon the account of promoting the circulating of the blood", Hook performed an experiment of keeping a dog alive for some time by continuously blowing air into its lungs without any respiratory movements. After a long hiatus, in 1908 came a similar description by Volhard.[6] Oxygenation of paralysed dogs was performed using an glass tube in the trachea, but without any respiratory movements. This only worked when 100% oxygen was provided, which was later confirmed by others.[7]

Through the following years, more aspects of this phenomenon were discovered. Draper et al., in 1947 described "diffusion respiration"[8] where in the alveoli can continuously capture oxygen in the absence of ventilatory efforts provided cardiac output remains adequate, oropharyngeal patency is maintained, haemoglobin is in the normal reduced state, denitrogenation and replacement with oxygen occurs, and a continuous oxygen insufflation is performed after the cessation of diaphragmatic movement. However, it was thought that alveolar oxygen attaching to the haemoglobin-oxygen pump provided an inward suction pressure leading to the diffusion of oxygen from the pharynx to the alveoli. On a similar note, the term 'aventilatory mass flow'[9] was coined by Bartlett, Brubach, and Specht (1959). By usage of whole body plethysmography, they demonstrated observable en masse movement of ambient air into the lungs. They postulated that this mass flow is due to the decrease in intrapulmonary pressure caused by the combination of long airway and tissue CO_2 retention. Since CO_2 washout was thought at that time to be equal to oxygen extraction from the alveoli, it provided an explanation for the observed mass flow.

These theories were disproved by Frumin et al. in 1959[10] who suggested to adopt the term apnoeic oxygenation as originally suggested by Nahas.[11] They demonstrated that extraction of oxygen from the alveoli into the blood was not equal to addition of CO_2 into alveoli from the blood. They proposed that a reduction in the barometric pressure gradient between the upper airway and the alveoli due to poor CO_2 diffusion into the alveoli from the blood as well as denitrogenation is the mechanism of apnoeic oxygenation. More physiologic depth and detail was added in the following studies conducted in the following years. Weitzner et al[12] (1959) studied arterial oxygen saturation at different periods of apnoea and showed that it drops to dangerous levels by about 1½ minutes. This paved the way for the concept of safe apnoea time. Millar et al[13] (1961) showed a significant sympathetic surge occurring during apnoeic oxygenation. Heller et al[14] in 1964 became the first team of researchers to perform apnoeic oxygenation in humans. In a group of healthy volunteers, after paralysis with thiopental or succinylcholine, bag and tube ventilation with 100% oxygen for 4 minutes, they induced apnoea

and measured polarographic oxygen tension in arterial blood every minute for 5 minutes. This experiment was performed under two conditions, one with endotracheal tube left open to room air versus connected to an oxygen reservoir bag. The study concluded that hypoxia can be avoided in respiratory arrest for specific periods of time through apnoeic oxygenation. Cardiopulmonary effects of apnoeic oxygenation were studied by Fraioli[15] in 1973. They quantified the degree of fall of PaO_2 and rise in $PaCO_2$ during AO, observed a slight decrease in FRC during the process of AO and recorded arrythmias at the same time (only PVCs were observed). Importantly, they observed that rate of desaturation during AO was dependent on ratio of FRC to body weight. The provision of AO at different flow rates were reported by Mackenzie et al in a study on dogs in 1991.[16]

The physiology of apnoeic oxygenation

With cumulative contributions from the above studies, the current understanding about the mechanism of AO is as follows. During regular breathing, where the volume of oxygen entering the blood from alveoli is nearly equal to the amount of CO_2 moving out of blood into alveoli. During apnoea, in contrast, though 250 ml/minute of oxygen moves from the alveoli to the blood, in the opposite direction, only 8-20 ml/minute of CO_2 moves from blood into the alveoli.[17] The rest of the CO_2 produced in the tissues gets buffered.[18] Due to this discrepancy, the alveolar pressure becomes sub-atmospheric and draws the air from pharynx to alveoli through passive diffusion. By giving apnoeic oxygenation, the pharyngeal space is filled with an oxygen rich reservoir which in its turn moves in and fills the lungs and helps to improve arterial oxygen saturation and prolong the safe apnoea time.

Summary of evidence for apnoeic oxygenation from paediatric emergency room or ICU studies

The studies performed in the cohort of paediatric patient undergoing emergency intubations in the emergency department (ED) or ICU are largely in the form of quality improvement (QI) initiatives undertaken by units to incorporate the practise of AO for presumably reducing the risk of adverse events. Some studies conducted in this context are summarised in table 1.

Discussion :

Due to the obvious physiologic advantage of continued oxygenation during apnoea in reducing the risk of hypoxemia and prolonging safe apnoea time, the high risks associated with hypoxemia during an emergency intubation, demonstrated efficacy in other cohorts under more controlled conditions, as well as the ease of application of the technique without reported adverse events, AO can be proposed as a standard of care in paediatric emergency intubations. Though evidence for efficacy of this practise has not been strictly demonstrated in high quality large randomised control trials (RCT) conducted in the paediatric population in the ED/ICU setting, work in adults and in the paediatric anaesthetic cohort is supportive.[26-29]

Several methods of providing apnoeic oxygenation have been described in adult literature, including nasal prongs, high flow nasal cannula (HFNC), nasopharyngeal catheter, endotracheal and endobronchial catheters and laryngoscope with oxygen channel incorporated in the design of the blade.[30] While the pros and cons of each method are debatable and not demonstrated in children in comparative studies, the simplest and the most common method used has been the nasal cannula. Flow rates have

varied in different studies, but approximately 5 L/min for infants, 10 L/min for children < 10 years and 15 L/min for older children > 10 years have been used more commonly[19, 23-25] and can be suggested. Flow rates and delivery technique require standardisation. Though, generally flow rates through nasal cannulae are limited to 6 L/min for lack of humidification, this may not be a practical impediment to use for short periods of time. Practically, if respiratory support in the form of HFNC is already being provided immediately prior to intubation, it may be useful to continue the same for pre as well as apnoeic oxygenation for RSI with FIO_2 increased to maximum allowed, though again, comparative studies of high versus low flow systems have not been performed. Simple nasal cannula is generally not an impediment to bag and mask ventilation in modified RSI, should that become necessary, but the HFNC cannula may be more difficult in this regard and should be removed if proving so. Furthermore, risk of gastric distension may be theoretically increased with AO, though this particular aspect has not been studied. It is again important to practically have two sources of oxygen during any emergency intubation, one for apnoeic oxygenation and the other connected to AMBU bag so that precious time is not wasted for changing the oxygen from one delivery method to another during an emergency. Multiple factors may come to consideration in assessing the risk of hypoxemia during an emergency intubation including the number of attempts, experience of the provider, adequacy of pre-oxygenation, presence of comorbidities like obesity, difficulty of the airway in both anatomic and physiologic terms etc. But it may be postulated that apnoeic oxygenation may be useful across the spectrum of paediatric intubations, with greater usefulness in more difficult contexts.

There may be several hindrances to the regular incorporation of this practise in routine care.[24] Lack of awareness of the practise, resistance to incorporating new practices, difficulty in remembering to provide in the context of an intense high-risk situation, perceived ease of intubation and reduced risk of hypoxemia, unanticipated difficulty in the context of selective usage, lack of standardisation of delivery technique, can all contribute to underutilisation. Therefore, as the above quoted studies have demonstrated, performing a QI initiative with a multipronged approach to improving utilisation can be highly effective in incorporation as a standard of care.

Conclusion :

Though new research is necessary to provide high quality evidence for efficacy in the specific context of paediatric emergency intubation, multiple studies have demonstrated safety and trends toward reduced risk of hypoxemia. Standardisation of delivery methods and flow rates needs more study. We can however conclude that apnoeic oxygenation is an easily applied, well tolerated technique with minimal adverse effects to help improve the safety of paediatric emergency intubations.

Authors/year of publication/n (sample size)	Study design	Method of providing apneic oxygenation	Outcomes
Mortimer et al[19]/2016/44 (58 intubation attempts)	Prospective case series	Nasal cannula, 5 L/min for <4 years, 10 L/min for 4 to 12 years, and 15 L/min for 12 to 18 years	AO was well tolerated in critically ill children and was not associated with adverse events.
Long et al[20]/2017/117 (46 post and 71 pre implementation of QI initiatives)	Prospective quality improvement study	AO using positive airway pressure/ PEEP via face mask/t-piece, and nasal cannula oxygenation during laryngoscopy.	Fewer hypoxic adverse events in post intervention cohort, all occurring only with multiple attempts. (multiple interventions designed to improve peri-intubation outcomes performed including AO)
Vukovic et al[21]/2018/149	Observational study	4 L/min for < 2 years, 6 L/min for 2-12 years, 8 L/min for > 12 years	Apneic oxygenation is an easily-applied intervention associated with decreases in hypoxemia during pediatric intubation. Nearly 50% of children not receiving AO experienced hypoxemia
Overmann et al[22]/2018/305	Retrospective observational study	Simple nasal cannula: 2 L/min for < 3 years, 4 L/min for 3-8 years, 6 L/min for > 8 years	AO was not associated with a lower risk of desaturation during RSI
Napolitano et al[23]/2019 / 1373 (661 pre- and 712 post implementation of AO)	Prospective pre-post observational study. Implementation of AO as QI initiative in academic PICU	Regular nasal cannula : 5 L/min for infants < 12 months, 10 L/min for children from 1–7 year, 15 L/min for children above 8 years	Moderate and severe oxygen desaturation fewer in post implementation phase with AO. It is feasible to implement AO as a QI intervention in an academic PICU.

Jen Heng Pek et al[24]/2020/47 (22 pre and 25 post QI implementation)	Prospective QI study	Nasal cannula: 5 L/min for < 1 year, 10 L/min for 1–10 years, and 15 L/min for > 10 years	Successful implementation of care bundle to incorporate AO as standard of care for pediatric emergency intubations
Napolitano, Polikoff et al[25]/2023/ 6549 (2554 pre and 3995 post implementation phase)	Prospective multicentric QI interventional study	Nasal cannula: 5 L/min for < 1 year, 10 L/min for 1–7 years, and 15 L/min for =8 years	Use of AO was associated with lesser tracheal intubation associated adverse events, but this result may be explained by patient, practise and provider factors.

Table 1 : Summary of evidence for apneic oxygenation from pediatric emergency room or ICU studies

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Original Article

Mucosal Hemangiomata of The Maxillary Antrum -- Our Experience

Basundhara Chakraborty[1], Amitava Maity[2], Ranjan Raychowdhury[3], Ushirin Bose[4], Jnanabrata Ray Chowdhury[5]

Abstract :

Maxillary hemangiomata are rare lesions and were originally described by Smith in 1959. They can be central (osseous) or mucosal in origin. Surgical excision is the mainstay of treatment and diagnosis can be challenging. We present 4 patients with mucosal antral hemangioma. Symptoms included nasal obstruction, epistaxis and fullness of cheek. All patients underwent contrast CT scan. Surgical excision was achieved endoscopically in 3 while one patient underwent open surgery. Complete excision was achieved in each case with minimal blood loss. Embolization was not required. Thus, it can be concluded that contrast CT is helpful; and majority can be treated endoscopically. Intra-operative hemorrhage is minimal.

Key words :

Maxillary sinus, Hemangioma, Epistaxis, CT scan, Endoscope.

Introduction :

Maxillary hemangiomata were originally described as benign vascular neoplasms causing bony expansion and remodelling.[1] They may be capillary or cavernous depending on the dominant vessel size. They are usually congenital, however a history of trauma may be elicited.

Dahlin reported that hemagiomata are quite common in the head and neck but infrequent in the sinonasal cavity.[2] Choukas divided maxillary hemangioma into osseous or mucosal types.[3] Their clinical picture and radiological features often confuse the diagnosis and may lead to inappropriate treatment.

In the five year period (2013-2018) we came across 4 such patients all of whom were successfully treated surgically.

Case Report :

Case-1

A 45 year old lady presented with a 1 year history of right sided nasal obstruction followed by sero-sanguinous discharge from the same side. The general examination was unremarkable. Anterior rhinoscopy revealed a polypoidal mass filling the right nasal cavity which on nasal endoscopy turned out to be a friable bluish polypoidal mass arising from the middle meatus. A contrast enhanced CT (CECT) scan showed a heterogenous mass filling the right maxillary antrum, extending to the nasal cavity (Fig.1). As the diagnosis was in doubt, informed consent was obtained for examination under general anesthesia; if the mass bled profusely, a biopsy would be obtained, if not total excision would be attempted. At surgery, bleeding was minimal and the entire mass could be removed endoscopically via a large middle meatal antrostomy.

Case-2

A 42 year old lady presented with intermittent epistaxis from her left nostril and fullness of the left cheek for 4 months. Nasal endoscopy

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revealed a polypoidal lesion in the left nasal cavity filling the middle meatus. It was friable and bled to touch. The CECT showed a heterogenous mass within the left maxillary antrum with a small focus of calcification. At surgery, the mass was endoscopically excised piece-meal with minimal bleeding (Fig.2).

Case-3

A 35 year old male presented with left sided nasal obstruction and intermittent epistaxis of 6 months duration. Anterior rhinoscopy and nasal endoscopy showed a bluish polypoidal mass in the left nasal cavity. The CECT showed a soft tissue mass in the left antrum causing bony expansion and remodeling of the sinus walls. The mass was excised endoscopically.

Case-4

A 19 year old male presented with intermittent painless bleeding from the right nostril for 5 years. Anterior rhinoscopy and nasal endoscopy showed an irregular polypoidal lesion filling right nasal cavity with possible origin from right lateral wall. The CECT showed a soft-tissue non-enhancing mass in right maxillary antrum extending into the nasal cavity with no widening of the pterygopalatine fossa. Under general anesthesia, the mass was debulked and tissue sent for histopathology; this was reported to be an inflammatory nasal polyp with stromal spindle cell proliferation. The patient was lost to follow up for 2 years, and then re-presented with similar complaints. The patient underwent endoscopic assisted right medial maxillectomy; the final histopathology report was consistent with hemangioma with bone involvement.

Discussion :

Smith in 1959 reviewed a series of intra-osseous hemangiomata of the maxilla and mandible, reporting profuse hemorrhage during surgery.[1] Choukas in 1963 described a case of sclerosing cavernous hemangioma of the maxillary antrum which was removed surgically with minimal bleeding.[3]

The precise mechanism of development is still debatable: theories include congenital malformation, vascular hamartomas, haemodynamic instability and local trauma, while hormones have been shown to affect their growth.[5] Head neck hemangioma can originate from skin, mucosa and even deeper structures like bone, muscle and gland (Batsakis 1984).[6]

The osseous variety has been divided into 2 types: central and peripheral. As opined by Smith, peripheral haemangiomata arise from vessels in the periosteum with secondary involvement of the bone itself; while central ones develop within the spongiosum.[1] Out of the 2 histological subtypes, capillary hemangiomata are more common and composed of capillary sized vessels with flattened epithelium and cavernous ones contain large, endothelium lined vascular spaces.[7] Among sino-nasal hemangiomata, those arising from the septum and inferior turbinate are predominantly cavernous while those arising from the maxillary antrum and middle turbinate are capillary in nature. Choukas stated that maxillary hemangioma may be central (osseous) or mucosal in origin.[3] As previously noted, the intra-osseous variety is prone to severe hemorrhage whereas minimal hemorrhage is encountered in the mucosal variety.

In their retrospective study Kim and Kwon (2017) stated the common symptoms to be nasal obstruction, epistaxis, rhinorrhea, and postnasal drip[8] which agrees with our findings. The

differential diagnosis includes entities like angiofibroma, nasal polyposis, fungal balls, organising hematoma and malignancy. An organising hematoma usually erodes the medial maxilla and ethmoid cavity, while angiofibroma usually affects the pterygopalatine fossa extending to the nasal cavity, nasopharynx, and sphenoid sinus.[8] Fungal balls are localized and may show calcifications on non-enhanced CT, while sinonasal malignancy usually exhibits invasive and destructive characteristics on CT and MRI.[9]

Kim et al (2017) in their study of 37 patients with sinonasal mucosal hemangioma, only 2 out of the 27 underwent CT scans showed heterogenous enhancement in the maxillary sinus.[10] In our case series, antral wall expansion and calcification were noted. Two cases of Kim's series underwent MRI which showed iso-signal intensity on T1 and high signal intensity on T2 weighted images with low-signal line. None of our patients underwent MRI.

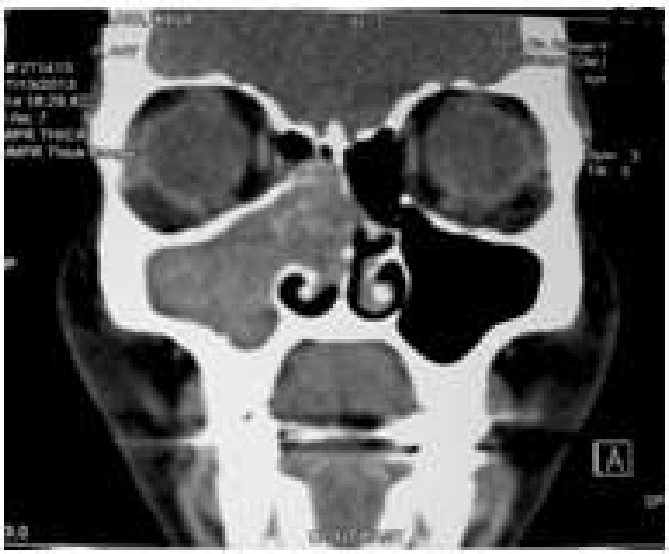
Since the majority of antral hemangiomata are capillary in nature, pre-operative embolization is rarely required. None of our patients underwent pre-operative embolization and no significant per-operative bleeding was noted.

The treatment of these lesions has changed markedly with time. Curettage, radiotherapy or sclerosant injections are of historical interest only. Open surgery by a lateral rhinotomy approach has been gradually replaced by completely endoscopic procedures. Those having extensive attachment to the lateral wall of nose may still require an external approach. Three of our patients underwent purely endoscopic excision while one had to be addressed with a lateral rhinotomy.[10]

Acknowledgements :

Secretary, Ramakrishna Mission Seva Pratishthan for permission to use hospital data.

Figgure 1A : Coronal CT scans showing a heterogenous mass filling the right maxillary antrum, extending to the nasal cavity.



Figgure 1B : Axial CT scans showing a heterogenous mass filling the right maxillary antrum, extending to the nasal cavity.

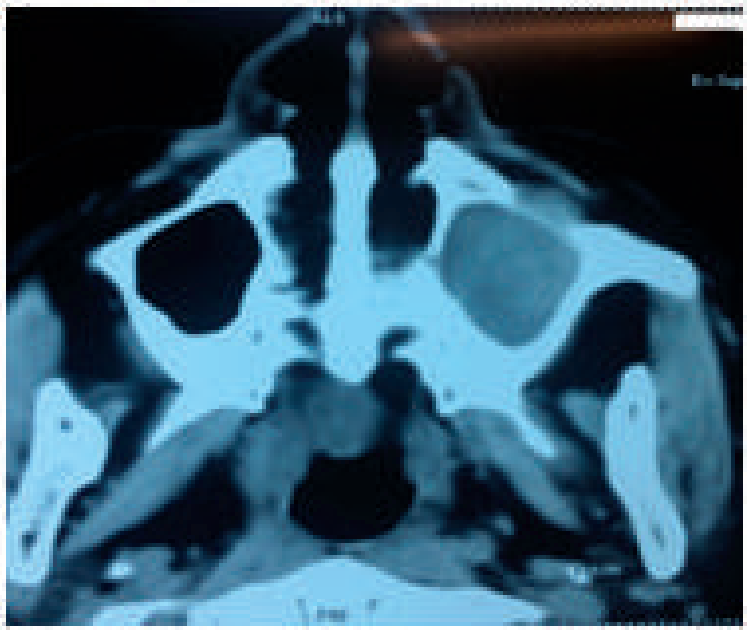
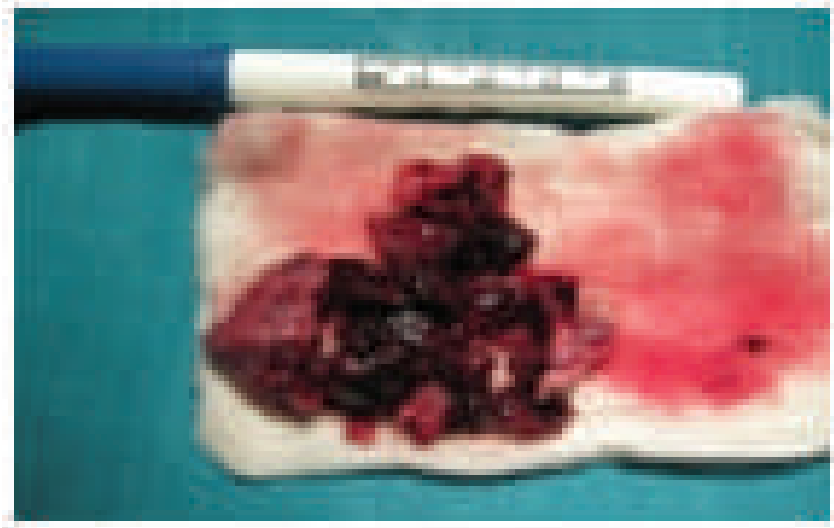


Figure 2 : Excised antral mass



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Case Report

Diabetic Mastopathy Mimicking Breast Carcinoma

Suranjana Dutta[1], Soumit Dey[2], Chhanda Datta[3], Pranamita Ray[4]

Abstract :

Diabetic mastopathy is a rare benign breast disease characterised by periductal and perilobular lymphocytic aggregation with stromal fibrosis. This type of lesion usually presents as unilateral or bilateral palpable breast mass in a patient with type 1 diabetes mellitus or autoimmune diseases but may also present in non diabetic patient and type 2 diabetes. We report a case of a diabetic mastopathy who presented clinically as an indeterminate breast lump. Ultrasonography, FNAC, trucut biopsy could not exclude the suspicion of malignancy so a simple mastectomy was done showing the typical features of Diabetic Mastopathy.

Introduction :

Diabetic mastopathy is a collection of clinical, radiological and histological features found in dense fibrous masses of the breast first described by Soler and Khardori[1] in 1984. The disease is associated with insulin dependent type 1 diabetes mellitus and some autoimmune disease but also seen in type 2 diabetes mellitus and some non diabetic patient.[2,3] Patients usually present with unilateral or bilateral palpable, hard, painless, irregular masses. Clinical, imaging and cytology findings are inconclusive and often misdiagnosed as breast carcinoma.

Case Report :

The patient described is a 62 year old Indian female, presented with a lump in her left breast of 4 months duration. There was no associated pain but she noticed an increasing size gradually. She has been treated for type 2 diabetes mellitus for last 16 years. She is also a patient of Chronic kidney disease and on hemodialysis thrice a week and associated with hypertension for last 14 years.

Physical examination revealed unilateral, hard, irregular, painless mass in the upper quadrant of the left breast. There was no nipple discharge or nipple retraction, skin abnormality or axillary lymphadenopathy.

The left breast ultrasonography examination showed a hyperechoic mass measuring 4cmx3cmx1.2cm and was reported as BIRADS 3 lesion.

FNAC was done outside of this institution. Microscopical examination showed the features of suspicious of Ductal Carcinoma of Breast (C5 Lesion). Slides were reviewed in our department and it was reported as Atypical Ductal Hyperplasia of breast (ADH) (C4 Lesion). Cytological report was downgraded due to poor cellularity of the smear.

Thereafter a Trucut biopsy was performed. Histologic evaluation revealed a dense fibrous stroma associated with some periductal and perilobular lymphocytic infiltration (fig. 1&2).

The mass was increasing in size and also there was a discrepancy between the cytology and histology reports so, a Simple Mastectomy was done.

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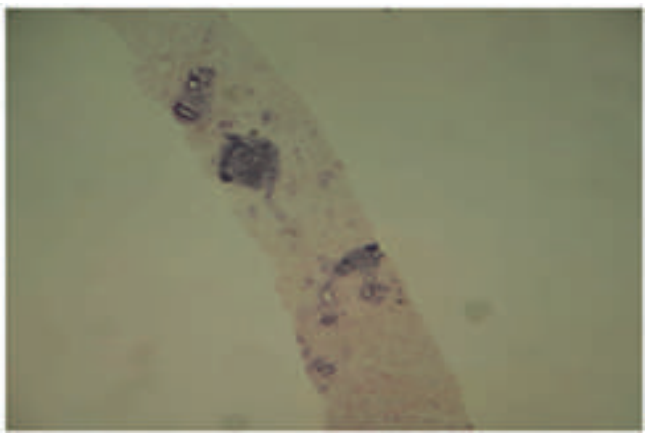
Gross pathology of the excised specimen demonstrated anterior aspect of left breast (fig.3) showed an unremarkable skin, no ulceration was present, nipple areola retraction was not there. After loafing a whitish, irregular, firm, partly circumscribed fibrotic mass was seen measuring 9cmx5.2cmx2.5cm (fig.4,5).

Histologically, the lesion showed dense stroma with duct and lobules (fig.6), lymphocytes aggregation around ducts, lobules and vessels, keloid like dense stroma (fig.7, 8, 9, 10) and some enlarged, scattered stromal Myofibroblasts (fig.11). Foci of dilatation of ducts (fig.8, 9) and Apocrine changes are present (fig.10). Some differential diagnosis could be Fibromatosis, non neoplastic fibrous lesion of Breast, Low grade metaplastic carcinoma. To confirm the diagnosis some Immuno-histochemical (IHC) Markers were applied.

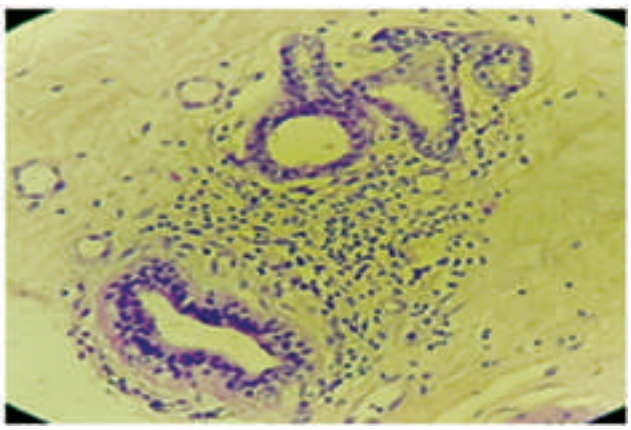
Pan cytokeratin was negative (fig.12). P63 was negative (fig.13), ruling out fibromatosis type of metaplastic carcinoma. Beta-catenin was negative in the lesion (fig.14), making fibromatosis less likely.

This case met all the histologic parameters fitting the diagnosis of Diabetic Mastopathy (DMP). The patient currently remains well and is on follow-up.



← Figure 1 : H&E, 40X

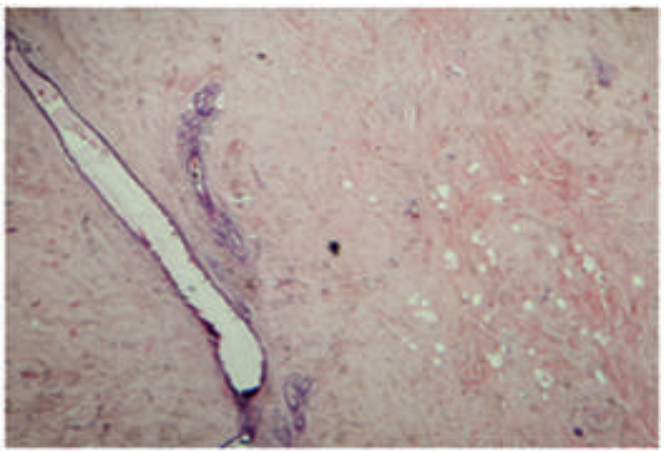
Figure 2 : H&E, 100X→



← Figure 3

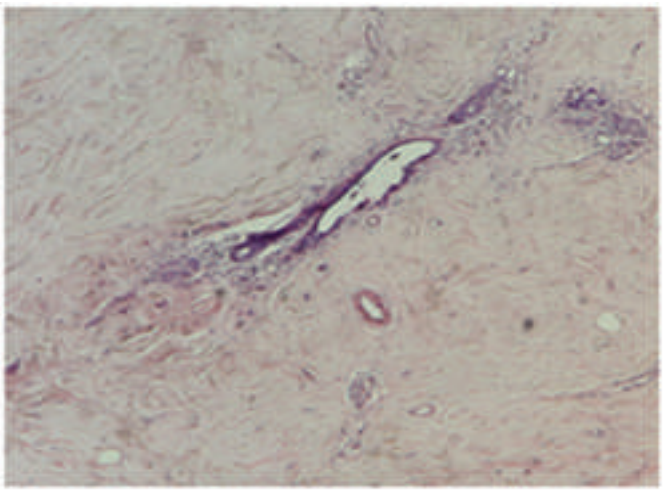
Figure 4 →

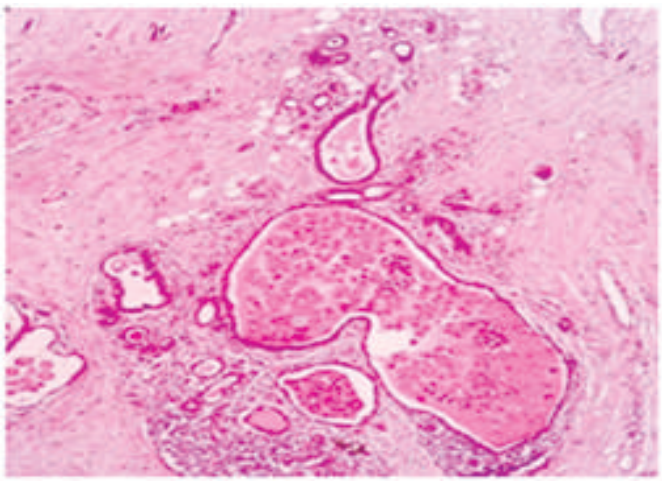
← Figure 5



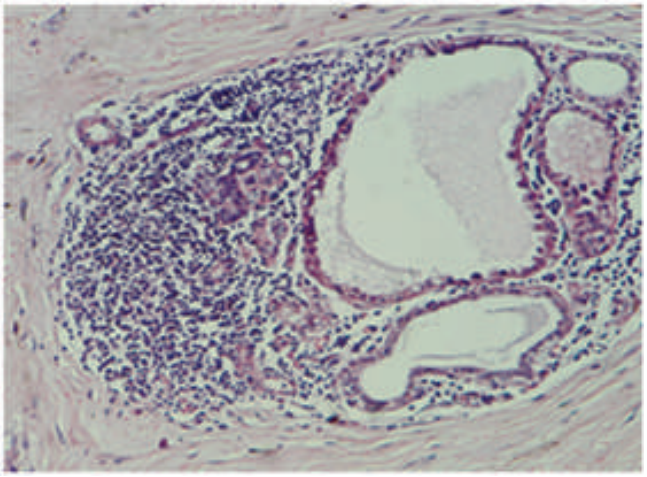
← Figure 6 : H&E, 40X

Figure 7 : H&E, 100X →



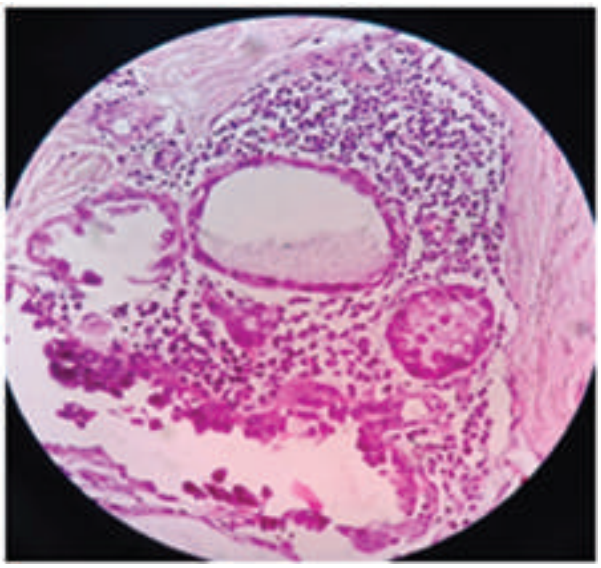


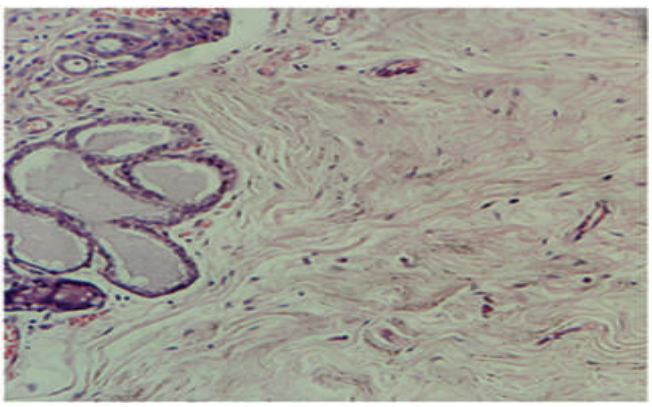
← Figure 8 : H&E, 100X



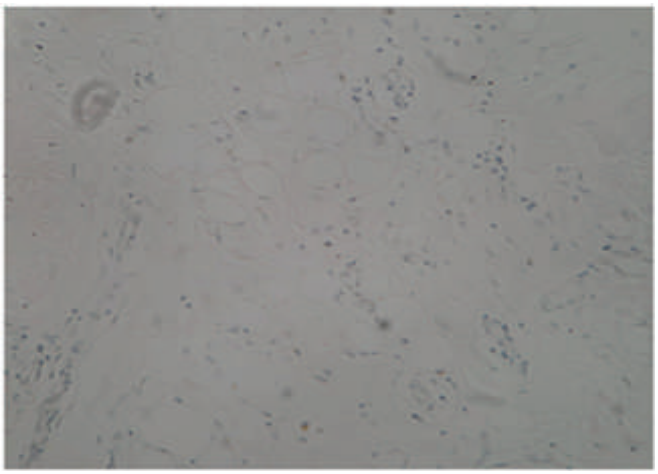
← **Figure 9 : H&E, 200X**

Figure 10 : H&E, 400X →



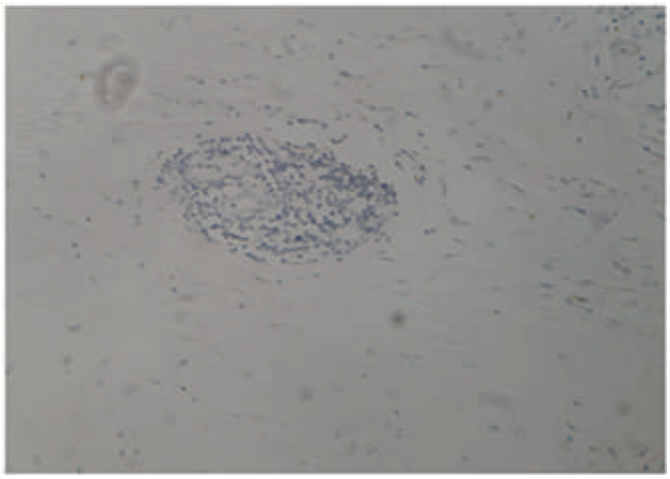


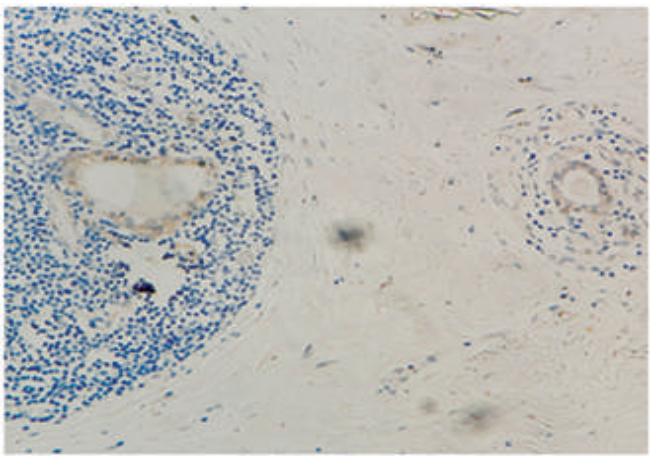
← **Figure 11 : H&E, 200X**



← Figure 12 : PAN-CK Negative

Figure 13 : P63 Negative, 200X →





← Figure 14 : Beta-catenin Negative, 200X

Discussion :

Diabetic Mastopathy (DMP) is a rare entity of self limiting fibroinflammatory disease of the breast associated with type 1 diabetes mellitus. The prevalence has been found to be less than 1% of benign breast diseases, but can range from 0.6% to 13% in type 1 diabetic women.[1] Most patients with DMP have an complication arising from diabetes such as retinopathy, neuropathy, nephropathy.[1, 4]

Clinical findings include hard, irregular, easily movable, painless breast masses. It can be solitary or multiple, unilateral or bilateral. These characteristics raised the suspicion of carcinoma.[1,5,6]

Tomaszewski et al.[3] found that certain microscopic features like epitheloid cells in the fibrous stroma were specific for Insulin dependent diabetes mellitus (IDDM).

Seidman et al.[7] proposed a combination of histologic features to confirm diabetic mastopathy. These were a collagenous stroma with keloid like features with a slightly increased concentration of stromal spindle cells and mature lymphocytes clustered circumferentially around small blood vessels as well as in and around lobules and ducts.

Even though many reports have been published on DMP in radiology, pathology and surgical literature, this clinical condition is poorly recognized since breast examination is not routinely performed in young diabetic patients.

The pathogenesis of DMP is not completely understood, but many theories have been proposed considering a multifactorial etiology.

Sternberg et al.[8] found that the glycosylation and increased intermolecular cross linkages in diabetics render collagen resistant to degradation. This leads to accumulation of connective tissue disorders in diabetics, including mastopathy.

Lymphocytic mastitis in DMP is usually associated with B cell lymphocytes.

Some researchers who studied the prognosis of DMP found that these lesions are prone to single or multiple recurrence in the same or contralateral breast.[9] Long standing IDDM in young women hence warrants a routine clinical examination of the breast.

FNAC can be used to monitor patients with recurrent lesions in a proven case of DMP, as it can show ductal epithelial cells in clusters, lymphocytes and epitheloid fibroblasts which are readily identified in connective tissue fragments.[9]

Conclusion :

Recognizing DMP requires an awareness of the existence of this entity and a careful correlation of the patient's clinical history with the physical, radiological, and pathological examinations. As the disease can be managed conservatively but may recur, an accurate diagnosis is essential to avoid unwanted multiple surgical biopsies. Monitoring of the patients by FNAC would be sufficient once the pathologic diagnosis is confirmed.

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Case Report

A Case of Soft Tissue Form of Rosai Dorfman Disease-Lump Over Knee

Ishita Ghatak[1], Soumit Dey[2], Pranamita Ray[3], Mir Hassan[4], Chhanda Datta[5]

Abstract :

Rosai Dorfman Disease (RDD) is a benign proliferation of histiocytes and classically occurs in lymph nodes. Extranodal involvement in systemic form may occur involving multiple sites: skin soft tissue, organs like-lung, liver, kidney and central nervous system. Soft tissue involvement may mimic inflammatory myofibroblastic tumour, organising lesion or Langerhans Cell Histiocytosis. Immuno-histochemistry is the cornerstone for diagnosis. A case of soft tissue lesion over knee joint in an adult is described here, which was evaluated and diagnosed to be RDD.

Keywords :

Histiocytosis, emperipolesis, Rosai-Dorfman Disease, S100

Introduction :

In 1969 Rosai and Dorfman, described 'sinus histiocytosis with massive lymphadenopathy'. It was a new entity, which is to be distinguished from 'malignant reticuloendotheliosis'.[1] They had four cases, all children, manifesting as massive cervical lymphadenopathy, fever, leucocytosis, and hypergammaglobulinaemia. The involved nodes showed dilatation of subcapsular and medullary sinuses by non-histiocytes. Focally the lesion caused total effacement of the architecture. The histiocytes often had intracytoplasmic lymphocytes ('emperipolesis').

Rosai-Dorfman disease is a rare proliferative disorder of histiocytes. The pathogenesis is not known yet. The interpretation is difficult in extranodal sites. It is commonly not a suspected lesion when it originates in bone and soft tissue. The less-characteristic histomorphology in comparison with nodal disease makes the diagnosis more challenging. The definitive diagnosis needs the presence of the characteristic S-100-positive histiocytes in tissue demonstrating emperipolesis. Bone and soft tissue lesions show less number of characteristic histiocytes and less conspicuous emperipolesis, areas of fibrosis or storiform spindle cells similar to fibrohistiocytic lesions. Here a case of an adult male is described.

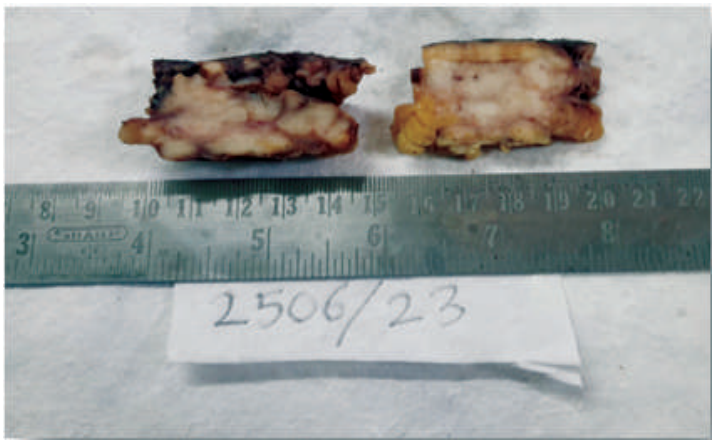
Case History :

A 46 year old, male patient presented with a lump over his right knee persisting for 6 months, which was soft to firm in consistency. The lesion was gradually progressive in size. The patient also had inguinal lymphadenopathy. An ultrasound scan suggested a solid nodular lesion. The clinical impression was that of lipoma. The patient underwent surgical excision after necessary work up.

Gross Features :

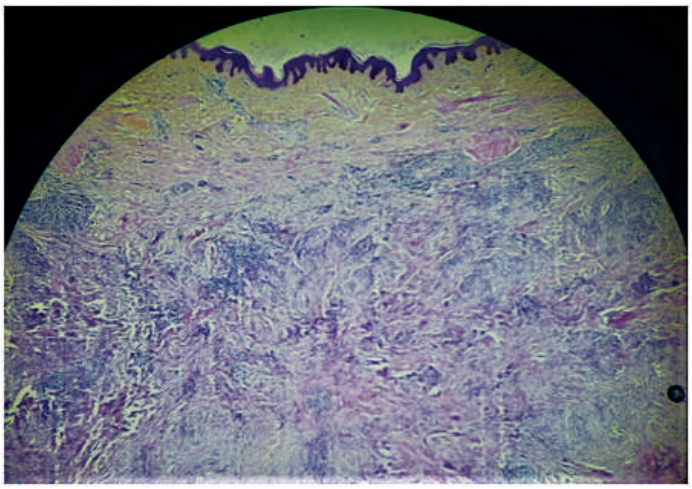
Grossly the resected specimen of lump showed a multinodular gray white lesion encroaching on to the deep resection margin. The lesion measured about 5 cm in maximum dimension (Fig.1).

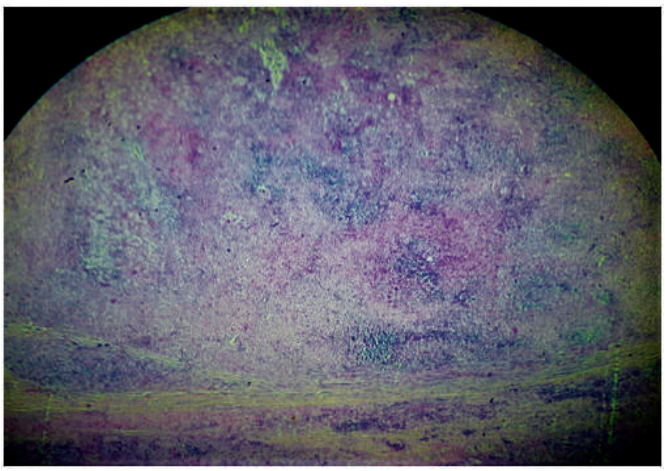
[1]Junior Resident, [2]Assistant Professor, [3,4]Associate Professor, [5]Professor and Head — Department of Pathology; RKMSP, VIMS



← Figure 1 : Gross Specimen

Figure 2 : Scanner View 4X →





← Figure 3 : Low Power View 10X

Microscopy :

Scanner view (4x) showed an ill defined lesion deep in dermis (Fig.2). The overlying epidermis was mostly unremarkable with a lesion free zone in between. The lesion showed dark staining areas admixed with some pale stained areas with some giant cells.

Low power (10x) view from dark areas showed rich lymphoplasmacytic infiltrate. Intermadiate magnification (20x) of the pale areas showed large foamy cells with pale eosinophilic staining admixed with lymphoplasmacytic infiltrate.

High power magnification (400x) revealed foamy histiocytes with engulfed lymphocytes and cell debris–features of emperipolesis (Fig.4). The overall histomorphology was consistent with Rosai-Dorfman disease.

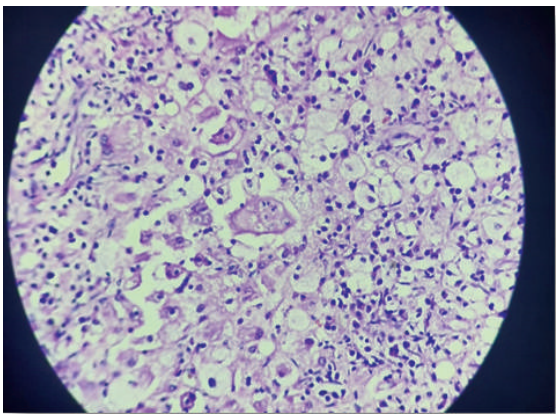


Figure 4 : High power view 400X

Discussion :

Rosai-Dorfman disease (RDD) is a benign rare histiocytic proliferative disorder characterised by massive lymphadenopathy. Multiple extranodalsite involvement can occur in generalised RDD; isolated soft tissue RDD (STRDD) is extremely uncommon. STRDD may occur as a part of a generalised process involving the lymph nodes or may occur at extranodal sites independent of lymph node status.[2]

In an extensive review, Komaragiri *et al.* diagnosed only 36 STRDD cases.[3] Two large case series of STRDD from the USA have been reported. The first described 17 cases of STRDD, of which four presented with lymphadenopathy. The second showed only one of 18 cases of STRDD to be associated with lymph node involvement.[4,5] In both series, STRDD was more common in the trunk and proximal extremities and had rapid growth.

Immunohistochemistry is the key to differentiating soft tissue lesions into the correct diagnosis. The differentials in this case included inflammatory myofibroblastic tumour, organising inflammatory lesion, Langerhans cell Histiocytosis with Rosai Dorfman Disease. The lesion showed a proliferation of histiocytes – mild to moderate atypia with nuclear pleomorphism. Large polygonal histiocytes showed emperipolesis predominantly of lymphocytes, sometimes plasma cells, erythrocytes and neutrophils. Marked plasmacytosis of polyclonal plasma cells, lymphocytosis predominated the histology.

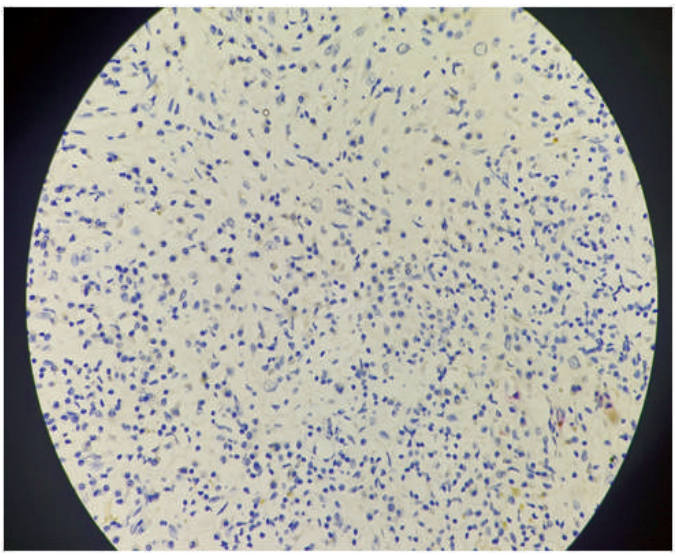
Some authors reported frequent spindling of the histiocytes, but others have claimed this phenomenon is uncommon.

RDD histiocytes are consistently positive for S100 proteins and CD68-KP and negative for CD1a and ALK. Negative ALK in this case excluded Inflammatory myofibroblastic tumor. CD1a negative status ruled out Langerhans cell Histiocytosis. The strong positive expression of S100, CD 68-KP and emperipolesis confirmed the diagnosis of RDD.

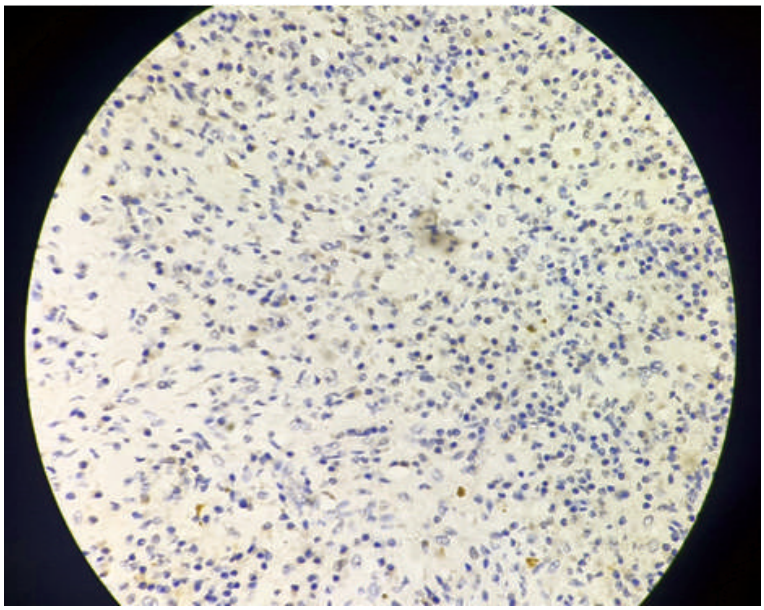
This case was diagnosed as the soft tissue form

of RDD (STRDD) and the diagnosis was based on light microscopy and immunohistochemistry of tissue sections from the specimen obtained by primary resection.

After surgery patient is on follow up in Surgery Outpatients clinic. No other lump, systemic symptoms or recurrence has occurred so far. Till date patient is doing well and hence no further management is being considered.



← Figure 5 : CD1a and ALK - both negative



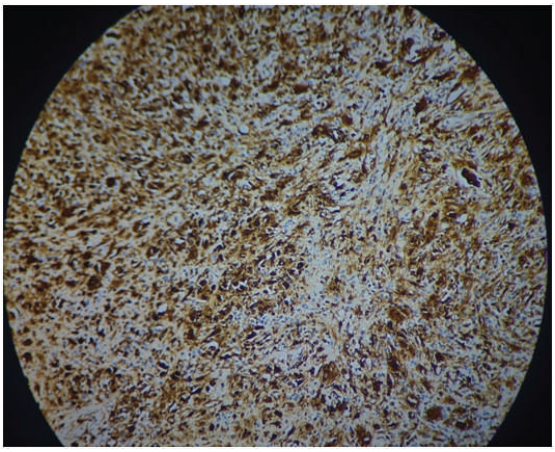


Figure 6 : Strong Expression of S100

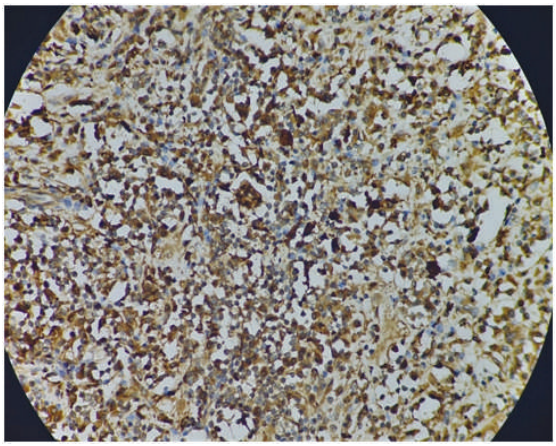


Figure 7 : CD 68-KP, Strong Expression

Conclusion :

RDD is a rare benign histiocytic proliferative disorder. The soft tissue variant is even rarer. Morphologically it may mimic an inflammatory myofibroblastic tumour, sarcoma or Langerhans cell histiocytosis. It is important that STRDD is included in the differential diagnosis of soft tissue lesions.

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Case Report

The Protean Manifestations of Mitochondrial Disease : A Case Report of ECHS 1 Deficiency

Prohlad Koley[1], Tapabrata Chatterjee[2], Aparna Mitra[3], Nisha Bhattacharya[4], Guruprashad H S[5]

Abstract :

Mitochondrial diseases are rare, involving almost all organs and all age groups. Clinical presentations depend uponthe rate and extent of genetic mutations. Sometimes clinical manifestations are exposed in stressful conditions like infections, starvation, dehydration etc. We present a case with High Anion Gap Metabolic Acidosis (HAGMA) with high lactate both in blood and Cerebrospinal Fluid (CSF), precipitated by infection and shock but lactic acidosis persisted after correction of precipitating causes. At this point with a back ground history of Global Developmental Delay and Hypothyroidism we had a suspicion that it might be a case of Inborn Errors of Metabolism or Mitochondrial Disease. Subsequently with relevant investigations we found it to be a case Mitochondrial Short Chain Enoyl-Coa-Hydratase 1 Deficiency (ECHS1). As the clinical presentations are varied, treating clinicians should have suspicion to pick up the diagnosis early.

Key words :

Lactic acidosis, mitochondrial disease

Introduction :

Mitochondrial disease is a heterogeneous group of inherited or acquired disorders affecting all age groups. The mitochondrion is the power house of a cell and present in all organs except Red Blood Corpuscles. So mitochondrial diseases involve almost all organs and may have very varied presentations.[1] We present a case of a 1 year 9 month old girl with global developmental delay, previously undiagnosed to be due to Mitochondrial disease. A metabolic crisis was precipitated by an intercurrent infection leading to lactic acidosis disproportionate to and unexplained by the clinical condition of the child led to the suspicion of an underlying metabolic disorder.

Case Descriptions :

Our case was 1 year 9 months old second born girl child born out of non-consanguineous marriage with background history of late preterm, Small for Gestational Age/Intra Uterine Growth Restriction, hypothyroidism (on regular treatment, controlled), failure to thrive and global developmental delay, presented with fever, loose stools, vomiting of 2 days duration, and respiratory distress for 1 day. On admission the baby was lethargic, with Glasgow Coma Scalescore of 12/15, tachypnoea (acidotic breathing), features of compensated shock and hepatomegaly. She subsequently suffered an upper gastrointestinal bleed.

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Table No. 1. Salient investigations and their interpretations

Investigations	Comments
Blood gas analysis-HAGMA (24) High lactate-9.5 Blood CSF lactate ratio -1	Suggestive of mitochondrial disease
Alanine transaminase (ALT)-96 U/L Aspartateaminotransferase (AST)-297 U/L International normalized ratio (INR) -2.1 Activated partial thromboplastin time (APTT)-39 Platelet counts-68000/cmm	Acute liver failure,disseminated intravascular coagulation (DIC)
Total leucocyte count 35100 Neutrophil 80% C reactive protein -209/L Blood culture – Escherichia coli growth	Sepsis likely precipitating for metabolic acidosis
Visual Evoked Potentials (VEP)-Optic pathway dysfunction	Retinopathy /optic neuropathy/deficiency of retro chiasmal pathway
Magnetic resonance imaging (MRI) - abnormal signal changes in basal ganglia	CNS involvement suggestive of Leigh disease
Tandem mass spectrometry (TMS) –increased levels of Malonylcarnitine	Nonspecific, may suggest mitochondrial respiratory chain disease

For confirmation of diagnosis Whole Exome Sequencing was done, which revealed **Mitochondrial Short Chain Enoyl-Coa-Hydratase 1 Deficiency** and also **Mitochondrial Complex 1 Deficiency Nuclear Type 19**.

The child recovered to baseline neurologic status on conservative management with fluid resuscitation, respiratory support, antibiotics, Fresh Frozen Plasma (FFP) transfusion, bicarbonate administration along with other basic supports and monitoring.

Discussion :

The rate of mitochondrial genetic mutation is very high. The mitochondrial Deoxyribonucleic acid (mtDNA) can exist in a cell as mixture of mutant and normal mtDNA (Heteroplasmy). In mitochondrial cytopathies cellular DNA also affected, hence all kinds of inheritance patterns are present and there is varied presentation involving almost all organs like the central nervous system (CNS), the peripheral nervous system (PNS), the heart, kidneys, muscles, eyes, ears, pancreas, liver and others.[1]

Leigh syndrome is one of the important mitochondrial diseases.

Most clinical presentations of Mitochondrial

Short Chain Enoyl-CoA Hydrstase1 deficiency are like Leigh syndrome which occurs in the neonatal or early childhood period. Clinical features of ECHS1 Deficiency are seizures, poor sucking, recurrent vomiting, lethargy, hypotonia, developmental delay and regression, episodes of lactic acidosis leading to respiratory and renal derangement, optic atrophy, sensory neural hearing loss, growth failure, dystonia, ataxia, hypertrophic cardiomyopathy and so on.

This child had several causes for high anion gap metabolic acidosis including hypovolemia, septic shock, and liver failure. Hence the child was initially treated for acute causes including fluid resuscitation, appropriate antibiotics, supportive treatment for liver failure, FFP transfusion. In spite of treatment of these causes lactic acidosis persisted, hence on the background of global developmental delay with metabolic worsening due to intercurrent infection, disproportionate acidosis with high lactate in relation to acute causes and persistence of lactic acidosis after resolution of acute causes an inborn error of metabolism (IEM), specially mitochondrial disease was suspected and investigated. Blood CSF lactate ratio,[2] abnormal VEP and MRI was suggestive of Leighs disease. The diagnosis of mitochondrial diseasewas confirmed through genetic testing.

ECHS1 Deficiency is rare disease; the exact prevalence and incidence is unknown.

There are no consensus clinical criteria for ECHS1 Deficiency, so the diagnosis is mainly based on genetic studies.[3]

Management is according to clinical presentations which may require a multidisciplinary approach.

The recommended surveillance for ECHS1 deficiency are – comprehensive neurological and developmental assessment, growth monitoring, echo cardiography, ophthalmologic and hearing evaluation.

Genetic counselingis necessary for future pregnancy as it is an autosomal recessive disorder.

Conclusion :

The presence of metabolic abnormalities like lactic acidosis disproportionate to or persistent after resolution of ongoing illness should lead to a suspicion of underlying metabolic disorders like IEM or mitochondrial disease.

We should look for other findings and supportive evidence and do relevant investigations corroborating with clinical findings to get the actual diagnosis.

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Case Report

An Aggressive Thyroid Carcinoma with Poor Prognosis

Supriya Dutta[1], Soumit Dey[2], Pranamita Ray[3], Chhanda Datta[4]

Abstract :

Anaplastic thyroid carcinoma (ATC) with rhabdoid features is extremely rare and only few cases have been reported in literature. We report a case of ATC with rhabdoid features in a 50-year-old male, who presented with a rapidly enlarging neck swelling. Ultrasound was reported as TIRADS 5 and fine needle aspiration of the thyroid swelling revealed it as poorly differentiated carcinoma (BETHESDA VI). The patient underwent total thyroidectomy. Pathology report showed rhabdoid phenotype of thyroid an aplastic carcinoma. This entity is highly aggressive and associated with a poor prognosis.

Keywords :

Thyroid cancer, Anaplastic carcinoma.

Introduction :

Anaplastic thyroid carcinoma (ACT) is highly aggressive uncommon tumour amongst all human malignant diseases. Papillary and follicular thyroid carcinoma account for 80-90% thyroid cancers whereas anaplastic thyroid cancer accounts for less than 5%.[1] It is the 3rd most common thyroid cancer.[2] Anaplastic carcinoma of thyroid develops in patients who have long standing cases of nodular goitres or incompletely treated papillary or follicular cancers.[3] Few anaplastic carcinomas present with metastasis in cervical lymph-node lung, bone, liver or brain.[4] Anaplastic thyroid carcinoma (ATC) with rhabdoid features is extremely rare[5] and only few cases have been reported in literature. The rhabdoid phenotype is a pathological presentation associated with aggressive nature not only in thyroid gland but also in other organs.[6]

Case Report :

A 50 years old male presented with complaints of rapidly increasing swelling in-front of neck over a period of 3 months associated with difficulty in deglutition. Patient had pressure symptoms. There was no family history of thyroid disease and no history of radiation exposure to the neck region.

Ultrasound examination was reported as TIRADS 5. A fine needle aspiration of the thyroid mass was performed. Cytology revealed it as poorly differentiated carcinoma (BETHESDA VI). Patient underwent total thyroidectomy. Specimen was sent to department of pathology for histopathology examination. Unfortunately, post surgery, the patient was lost to follow up.

Macroscopic examination showed total thyroidectomy specimen measuring 10x6x5cm involving both the lobes and isthmus; Right lobe measuring 5x4x3cm and left lobe measuring 6x5x4cm. Cut surface of left lobe showed circumscribed lesion with grey white and fleshy area measuring 5x4cm. Cut section of right lobe showed a whitish lesion with nodules and cystic spaces within it measuring 4x3cm and the lesion eroded the thyroid capsule. Isthmus also showed a whitish nodular lesion (Fig-1).

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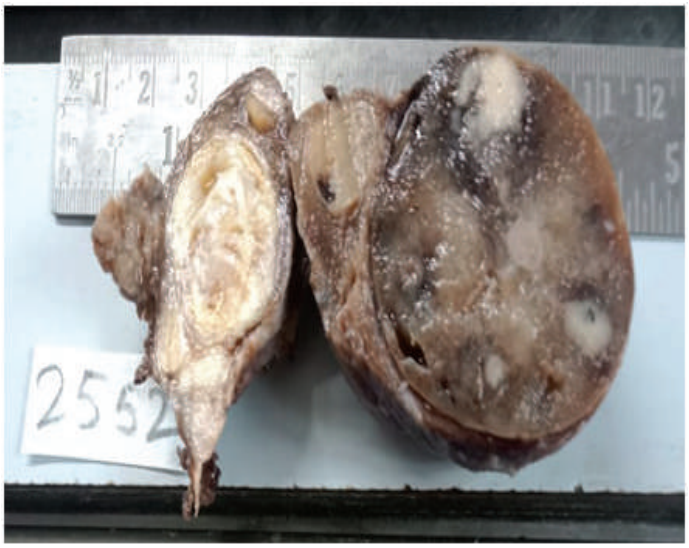
Microscopic examination showed a hypercellular neoplasm composed of large pleomorphic cells arranged haphazardly having round to oval hyperchromatic eccentric nuclei, and abundant eosinophilic cytoplasm (Fig-4). Tumour cells infiltrated in to the adjacent residual normal thyroid tissue (Fig-2). Extrathyroidal extension to skeletal muscle and perineural invasion were present (Fig-3 & 5). A diagnosis of undifferentiated carcinoma of thyroid was made based on histopathological examination. Immunohistochemical staining showed immunoreactivity for TTF1 was positive for normal thyroid tissue but negative for tumour cells. Immunostains for Melan A, ALK and CD45 were negative for tumour cells. The large pleomorphic cells with abundant cytoplasm and eccentric nuclei showed immunoreactivity for Desmin.

A diagnosis of anaplastic carcinoma of thyroid with rhabdoid differentiation was made based on histopathological and immunohistochemical examination.



← **Figure 1 : Gross specimen of total thyroidectomy showing both the lobes and isthmus were involved by the lesion.**

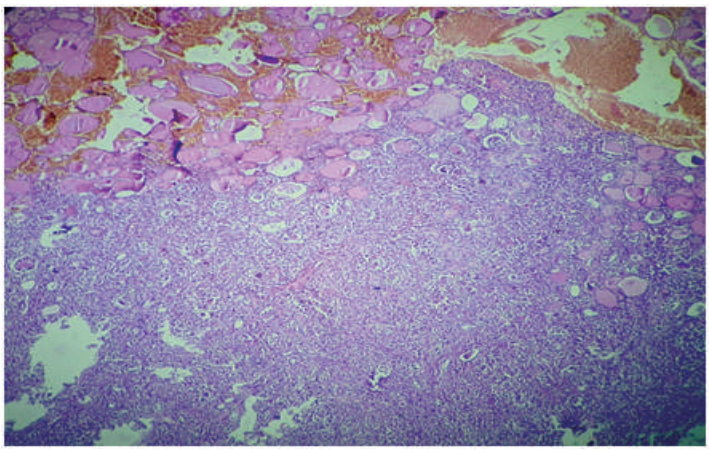


Figure 2 : Microscopic Examination (H & E) (X40) shows hypercellular area infiltrating the adjacent normal thyroid tissue.

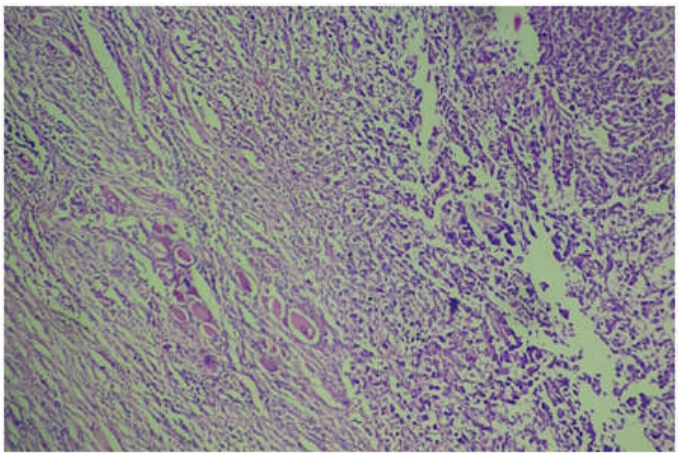
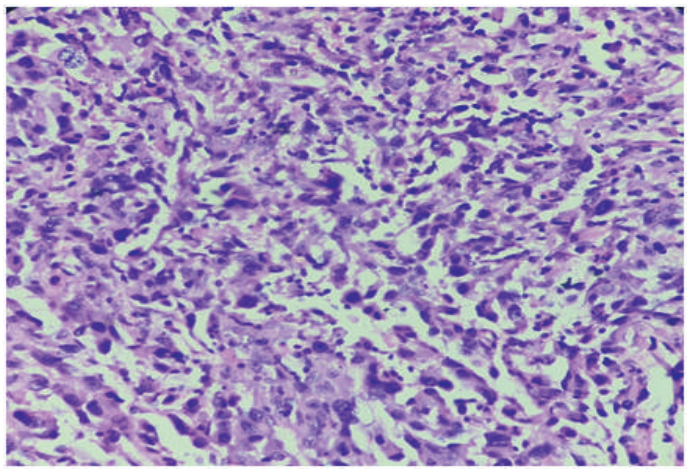


Figure 3 : Microscopic Examination (H & E) (X100) shows extrathyroidal extension to skeletal muscle.

Figure 4 : Microscopic Examination (H & E) (X400) shows large pleomorphic cells arranged haphazardly having round to oval hyperchromatic eccentric nuclei, and abundant eosinophilic cytoplasm.



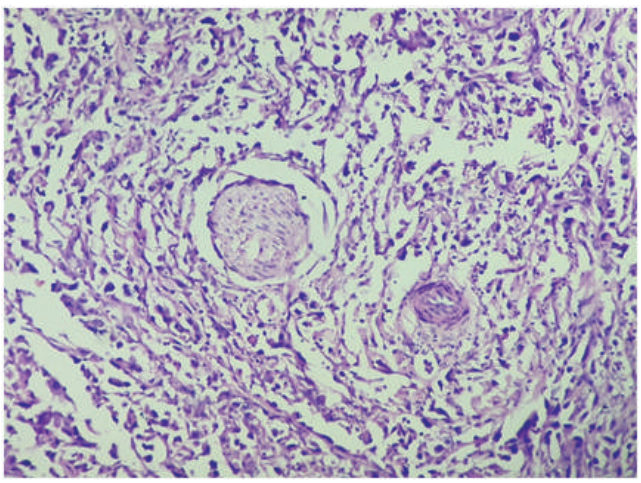
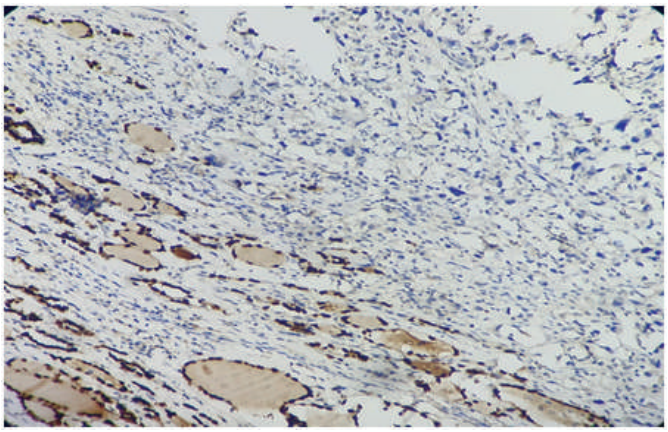
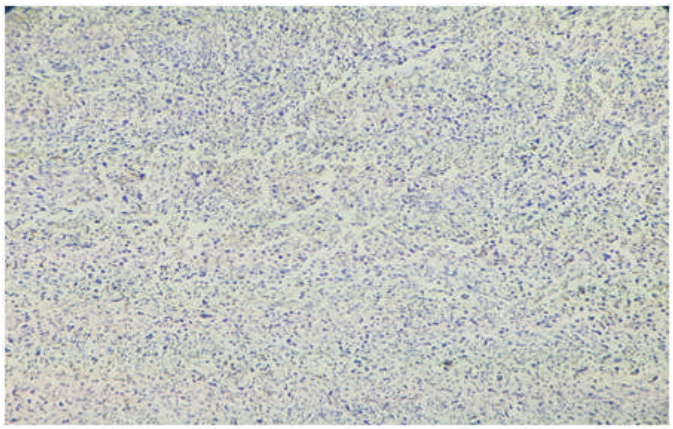


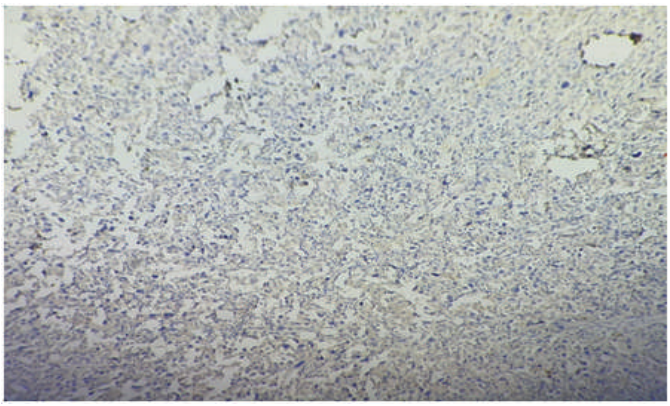
Figure 5 : Microscopic Examination (H & E) (X100) shows perineural invasion by the tumour cells.



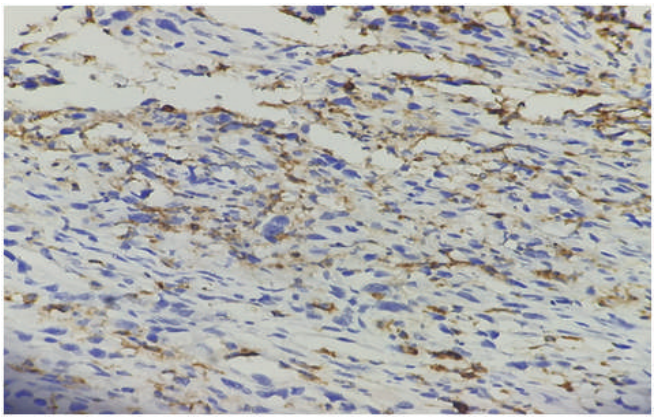
← Figure 6 : TTF1

Figure 7 : Melan A →



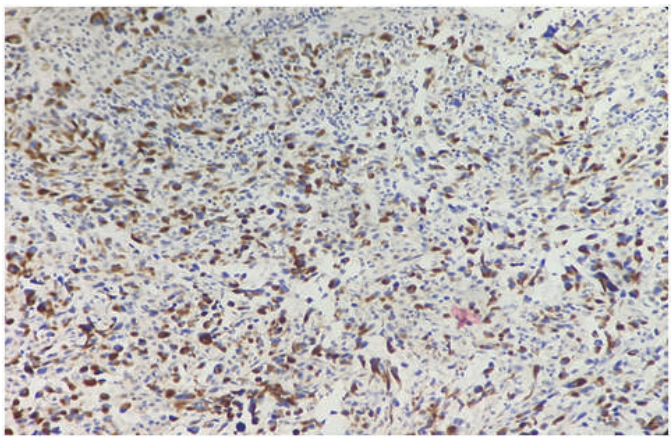


← Figure 8 : ALK



← Figure 9 : CD45

Figure 10 : Desmin →



Discussion :

Anaplastic carcinoma is an aggressive form thyroid gland cancer. It usually arises by dedifferentiation of a pre-existing differentiated carcinoma. The major risk factor in anaplastic thyroid carcinoma cases have previous history of benign or malignant thyroid disease.

ATC with rhabdoid features is extremely rare[5] and only few cases have been reported in literature. A recent literature review including 12 cases of rhabdoid tumours of the thyroid has shown that the rhabdoid phenotype is reported in 5 cases of follicular carcinoma, 3 cases of papillary carcinoma and 4 cases of anaplastic carcinoma.[7]

Clinically, most of patients present with a rapidly enlarging neck swelling with local compressive symptoms, like our patient.[8]

This variant appears macroscopically as a solid, irregular mass, with a whitish or greyish colour.[7] Microscopically, features include the presence of large pleomorphic cells with abundant cytoplasm and characteristic cytoplasmic eosinophilic inclusions and eccentric nuclei.[5]

The differential diagnosis of such tumour includes high grade lymphoma, malignant melanoma etc. These diagnosis are eliminated by the morphological aspect and the immuno-histochemical study.

This entity is highly aggressive and associated with a poor prognosis due to lack of response to radio and chemotherapy. It is associated with local recurrences and metastases with a median survival of 6 months.[5]

Conclusion :

Thyroid tumour with rhabdoid phenotype is a rare tumour with difficult diagnosis. This entity is highly aggressive with a poor prognosis. Clinical, Histopathological and immuno-histochemical examination are helpful to confirm the diagnosis. Surgery was performed in most of the cases and the benefit of adjuvant therapy was not clear.

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