Case Report

Massive Pleural Effusion as A Presenting Feature of SLE : A Diagnostic Challenge

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Abstract :

Large volume pleural effusions as a presenting feature of Systemic Lupus Erythematosus (SLE) may present a diagnostic and therapeutic challenge. Exudative and transudativeaetiologies are both possible as well as co-existing infectious causes. Multiple aetiologies may also be found contributing to the effusion in the same patient. The optimal therapeutic modality is not clearly established. We hereby present a case report of a 9 year old male child, illustrating these dilemmas and briefly discuss the learning points therein.

Keywords :

Systemic Lupus Erythematosus, Pleural effusion

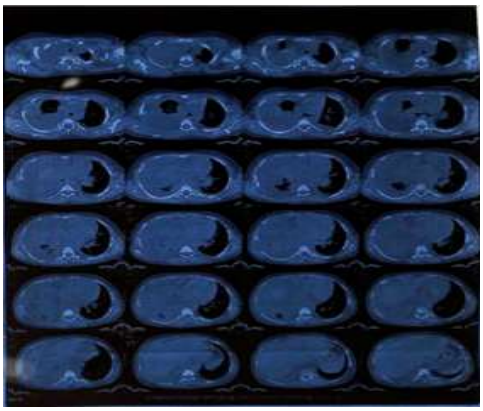
Introduction :

Serositis is a common feature of SLE. However, large volume effusion occurring as a manifestation of active SLE is rare. This finding can lead to a diagnostic dilemma as it may represent an inflammatory exudative effusion due to serositis, a coexisting infection which may be pyogenic or tubercular or have a transudativeaetiology due to complications of SLE like constrictive pericarditis or heart failure due to hypertension from renal involvement[1,2]. Hereby we present a 9 years old with SLE with such a diagnostic and therapeutic conundrum.

Case Description :

A 9 year old male child presented with fever of 1 month duration and swelling of the entire body for 5 days. He had a history of multiple skin lesions that started over the malar area with sparing of the nasolabial fold for the past 6 weeks which progressed to the abdomen and extremities. He was admitted elsewhere, and lupus was suspected, but he was also found to have a right sided consolidation and effusion, hence received antibiotics before transfer to our institute.

At admission the positive findings included a malar rash with erythematous plaques over the trunk andlimbs, pallor andanasarca. There were clinical findings suggestive of a right sided consolidation, pleural effusion and hepatomegaly. Investigation established a diagnosis of SLE, with massive right sided pleural effusion and patchy consolidation, along with Grade III A nephritis.



HRCT showing right sided Massive pleuaral effusion

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Investigations :

Hb	7.1	Sr ANA	185
TC	15200 (N90,L6)	Anti ds DNA	450
plt	404000	C3	17.3
CRP	44.3	C4	4.5
procal	0.15	DCT	3+
U pr:cr	3.04	Pl fl ANA	**1:640**
ALT	67	Retic ct	1.5
AST	56	alb	2.3

ECHO - Mild pericardial effusion
APLA - Negative

Immune suppression was commenced with hydroxy chloroquine and oral corticosteroids at 1.5mg/kg daily. Though consolidation changes were present on the CT scan, preliminary investigations did not suggest bacterial infection, possibly due to prior treatment with antibiotics. All cultures were negative (blood, pleural fluid). The factors contributing to pleural effusion included serositis, infection and hypo-albuminemia. In view of the lymphocyte predominant effusion an extensive tuberculosis workup was done but was negative. As the respiratory compromise needed HFNC support and there was non-resolution of effusion with a week of antibiotic therapy, an intercostal drainwas inserted, draining 700 ml on day 1 with persistent drainage (150-200ml) on a daily basis. With these findings persistent pleural effusion was thought to be due to serositis, and steroid therapy was escalated to pulse methyl prednisone followed by 2 mg/kg/day of IV methyl-prednisolone, due to concerns about oral absorption in view of presumed gut oedema. With these measures there was a gradual reduction in pleural fluid drainage to less than 20ml / 24 hours over a period of 8 days after which the drain was removed.

A renal biopsy revealed class 3 lupus nephritis. Hypertension was present requiring multiple antihypertensive medications (enalapril, amlodipine, prazosin). The child was discharged after 26 days in hospital, with oral corticosteroid therapy, hydroxychloroquine and mycophenolatemofetil.

Discussion :

Pleuro-pulmonary involvement occurs commonly in childhood onset SLE (cSLE) with 7-75% of patients being affected as per case series[3]. Pleural involvement is the most common pulmonary manifestation of SLE (30 % of patients with cSLE). Pleural effusion may also be the sole presentation of SLE in a small number of cases (5%)[4]. In some autopsy series, pleural effusions have been found in 93% of cases.

Aetiology :

The majority of patients with cSLE may develop pleural effusion as part of serositis, resulting from pleural inflammation, and this is often bilateral and exudative in nature[1]. However we should evaluate for the presence of life threatening causes of pleural effusion such as infection, CCF, AKI, pulmonary embolism and malignancy. An acute cardiac diagnosis leading to CCF due to myocarditis, pericarditis, valvular insuficiency or endocarditis may be present in up to 17.8% of cSLE and may also lead to pleural effusion[5]. Renal involvement leading to AKI causing fluid overload may be a cause.

Effusion may accompany pulmonary embolism and malignancy as well. Apart from the above, anti TNFa medications and antihypertensives like hydralazine may cause drug induced pleuritis.

Pathogenesis :

Pleural inflammation leading to increased vascular permeability and decreased absorption of pleural fluid contributes to effusion. Local autoimmune complex depositions, production of pro inflammatory cytokines, compliment activation and direct binding of anti dsDNA antibodies to the mesothelium have also been postulated as cause[6,7].

Clinical Presentation :

Fever, cough, dyspnoea, pleuritic chest pain.

Laboratory Features :

Appearance of effusion - yellow or serosanguinous. Elevated levels of protein and LDH with normal glucose levels[1]. Cell count 200-15000/cmm with either neutrophilic or lymphocytic predominance. Lupus erythematosus (LE) cells may be found in the pleural fluid. A higher proportion of pleural ANA positivity is found in lupus pleuritis. It is more sensitive than specific. A titer of = 1:160 may differentiate lupus pleuritis from other causes[8]. Along with this criterion, homogeneous pleural fluid ANA staining patterns and pleural fluid to serum ANA titre ratios =1 may also point to the diagnosis. C3, C4 levels are both found to be low in lupus pleuritis.

Treatment :

NSAIDS and a short course of oral steroid or, in resistant cases, pulse methyl prednisolone. ICD may be rarely required. In chronic lupus pleuritis, not responding to medical therapy, local therapy may be resorted to. These options, with very limited pediatric experience include intrapleural steroid injections, pleurodesis with talc or tetracycline and as a last resort pleurectomy.[9]

Conclusion :

Large pleural effusions with SLE may be occasionally present and cause diagnostic and therapeutic dilemmas and need to be differentiated from infective causes. Pulse methylprednisolone may be helpful to resolve the serositis once infection is ruled out.

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