Recurrent Massive Pleural Effusion with Neurosarcoïdosis: A Rare Presentation of Sarcoidosis

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Abstract

49 year old female, a known case of thalassemia minor with hypothyroidism on treatment, presented with left sided massive pleural effusion. Pleural tapping revealed exudative effusion and she was started on four drug AKT. She had recurrent pleural effusions and required repeated aspirations. Thoracoscopic pleural biopsy revealed non caseating granulomas. She continued to deteriorate after 8 months of AKT and was readmitted with severe vomiting, ataxia, diplopia, weakness and weight loss. Investigations revealed miliary pattern on CXR and multiple granulomatous lesions in the brain on MRI. She responded to treatment with high dose corticosteroids. We are presenting this case to highlight sarcoïdosis as one of the causes of large pleural effusion.

Abbreviations: CXR - Chest X Ray; AKT - Anti Koch’s Treatment; ICD - Inter costal drain; MT - Mantoux Test; ACE - Angiotensin Convertase Enzyme; CNS - Central Nervous System

Case

A 49 year old female presented with complaints of breathlessness on exertion, dry cough and low grade fever since 15 days. She was a known case of thalassemia minor and hypothyroidism on Tab. Eltroxin. She was HIV seronegative. There is no past history of any other significant illness.

On general examination she was tachypnoeic with a respiratory rate of 32/min. On respiratory system examination, breath sounds were reduced on the left side, with a dull note on percussion. CXR was suggestive of a large left sided effusion. She was diagnosed to have a left sided massive pleural effusion and pleural aspiration was performed. Pleural fluid showed pH 7.34, proteins 4 g%, WBC 1200, 80% lymphocytes, 20% neutrophils and normal ADA levels of 35IU/l suggestive of an exudative effusion. Pleural fluid smear and culture for AFB were negative. Pleural fluid cytology was negative for malignant cells. Gram stain and culture also were negative.

Haematological investigations showed mild anaemia with Hb 8.6%, rest were within normal limits.

MT was negative. Sputum AFB was Negative. Patient was started on AKT, with HRZE. She received daily antitubercular treatment. Patient continued to have refilling pleural effusion even after 3 months of AKT for which she underwent therapeutic pleural aspiration thrice. Repeat pleural fluid examination did not yield any conclusive diagnosis however possibility of lymphoproliferative disorder was raised. She was then referred to higher centre for left sided thoracoscopic pleural biopsy. During the procedure she had sudden cardiopulmonary arrest, was resuscitated and put on ventilator, was on the ventilator for one week after which she recovered. Left sided ICD insertion was done for refilling pleural effusion. Pleural biopsy microscopy sections on H&E stain showed moderate lympho-histiocytic infiltrate with several ill defined granulomas and occasional one with Langhans giant cells (Figs. 1 and 2). Z-N stained sections do not show acid fast bacilli. She was continued on AKT.

She presented again after a period of four months, this time with severe weakness, persistent loss of weight, decreased vision of the right side, diplopia and ataxia. Her chest X-ray revealed bilateral upper, mid and lower zone reticulo-nodular opacities. She was put on second line AKT i.e Kanamycin, Cycloserine, Ethionamide, PAS, Pyrazinamide and Ethambutol with no significant improvement. HRCT- Thorax revealed multiple tiny nodules in both lung fields, fibrosis with volume loss in left Lower lobe with pleural thickening.

MRI- Brain revealed multiple, tiny ring to disc enhancing lesions with minimal perifocal edema in bilateral cerebral,
Sarcoidosis is a multisystemic, granulomatous disorder of unknown etiology. It commonly affects young & middle aged adults, more common in females. It was first described by Hutchison, in the year 1877. Lungs are the most commonly affected organ. It usually presents with multiorgan involvement, the organs frequently involved are skin, CNS, heart, kidneys and eyes. The presence of bilateral large effusions in sarcoidosis is unusual. The reported prevalence of pleural involvement in sarcoidosis varies from 0 to 5% with unilateral, small effusions usually. The incidence of pleural effusion with histologically proven sarcoidosis is 0.16%. Cardiac sarcoidosis occurs in 3 to 5 percent of patients. Patients manifest with bundle branch block, tachyarrhythmia, bradyarrhythmia, congestive heart failure, pericarditis or cardiomyopathy, ventricular aneurysm, angina and papillary muscle dysfunction.

Initially sarcoidosis was thought to be uncommon in India. The first documented case was in 1956 which was followed by several case reports. The first large series of 75 cases was published by Dr S K Gupta in 1982. Until December 2000, 647 published cases have been recorded in India. Dr Sharma S K and Dr Mohan A have exhaustively reviewed the various uncommon presentations of sarcoidosis in India. The authors have reported pleural involvement, optic neuritis, cardiac involvement including complete heart block, Congestive cardiac failure, supraventricular tachyarrhythmias, and neurosarcoidosis and skin involvement, among others.

Neurosarcoidosis occurs in less than 5 percent of patients. Manifestations include involvement of cranial nerves (especially the facial and optic nerves), encephalopathy, seizures, cerebellar signs (such as ataxia or tremor), hearing loss, peripheral neuropathy and space-occupying masses. Cerebrospinal fluid may show elevated protein levels, decreased glucose levels and pleocytosis.

Ocular disease in sarcoidosis is much more common and affects approximately one quarter of patients with systemic sarcoidosis. Ocular manifestations include uveitis, chorioretinitis, cataracts, blindness, lacrimal gland swelling and inflammation, and retinal periphlebitis. Pulmonary involvement is by far the commonest and occurs in about 90 per cent of the patients. The diagnosis is based on a compatible clinical and/or radiological picture, histopathological evidence of non-caseating granulomas in tissue biopsy specimens and exclusion of other diseases capable of producing similar clinical or histopathological appearance.

The goals of treatment for sarcoidosis include resolving inflammatory lesions that are interfering with organ function, preventing pulmonary fibrosis and diminishing symptoms. The commonly used drugs are steroids, Chloroquine, Methotrexate, Azathioprine, Cyclophosphamide. Heart Lung Transplantation or single or both lung transplantation has been tried in severe cases. Our patient was treated with oral steroids, 0.5mg/kg/day for 4 weeks and then tapered to maintenance dosage over a period of 1 year as per the ATS guidelines.

Sarcoidosis is not usually thought of in the differential diagnosis of massive bilateral pleural effusion. However as our case report shows it can be responsible for even bilateral or unilateral massive pleural effusion in a small percentage of cases. Therefore, alternative diagnosis of sarcoidosis should also be considered in cases of massive pleural effusion specially in patients who do not respond or deteriorate in spite of anti tuberculosis treatment.

References


Fig. 3: MRI- Brain T1 weighted image revealed multiple, tiny ring to disc enhancing lesions with minimal perifocal edema in bilateral cerebral, cerebellar hemispheres and pons – Suggestive of multiple granulomas.