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Case report

Primary systemic amyloidosis: A rare cause for pleural effusion

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A B S T R A C T

Pleural effusion is a common problem dealt by most of the practicing clinicians. Some causes for pleural effusion are less often considered as a differential diagnosis owing to its rarity. Here we report a case of renal amyloidosis on alternate day haemodialysis for about two months time presenting with left sided pleural effusion. On evaluation this turned out to be a case of amyloidosis on thoracoscopic pleural biopsy suggesting the possibility of Primary systemic amyloidosis.

Case report

A forty two year old lady was referred to us from the department of nephrology with dyspnoea and left sided pleuritic chest pain of one week duration. She was managed in the nephrology ward as a case of synpneumonic effusion with antibiotics and other supportive measures. She was an established case of renal amyloidosis and was being worked up as a prospective transplant case. She was detected to have hypertension in the past and also gave history of preeclampsia and frequent foetal loss. She was thoroughly worked up for the evidence of collagen vascular disease which were negative. She had proteinuria, hypothyroidism and hypertension. Her renal biopsy revealed mild mesangial cellularity, Congo red positive eosinophil material in the glomeruli, tubular atrophy, interstitial inflammation and thickened vessel wall and this was reported as consistent with renal amyloidosis.

There was evidence of massive left side pleural effusion with mediastinal shift on digital Chest X-ray Fig. 1. Thoracocentesis was done and it initially drained straw coloured fluid which later on turned to hemorrhagic fluid on subsequent aspirations. She did not have oliguria, haematuria. There was history of loss of appetite, exertional dyspnoea and orthopnoea for last two weeks. Two of her maternal uncles had chronic kidney disease, details of which were not known. Her renal functions gradually worsened over a period of eight years and she was diagnosed as stage V Chronic Kidney disease. She was initiated on haemodialysis since last two weeks and an alternate day schedule was planned. Pleural fluids study showed low ADA, exudative effusion with a total count of 300 cells/mm^3 with 79% lymphocytes.

The pleural fluid cytology did not reveal any malignant cells. She was subjected to repeated thoracocentesis, alternate day haemodialysis and blood transfusions on top of broad spectrum antibiotics. Her Mantoux was non reactive. She was also on weekly doses of erythropoietin, antihypertensives, oral soda bicarbonate, diuretics and iron supplements. She was on thyroxine replacement following thyroidectomy for last fifteen years. Blood investigations during admission showed Hb-8.4 gm%, TC-6000 cells/mm^3, DC-P76L24, ESR-90 mm/h. Her renal function values were S.Creatinine-13.1, Urea-155 mg%. Serum Potassium was 4.1 meq/L. Her total protein was 5.9 gm% with albumin 3 gm%. Her serum electrophoresis did not reveal any abnormal paraprotein. Her skeletal survey also was normal.

She had worsening dyspnoea while she was in the nephrology ward and was shifted to our side and was subjected to a thoracoscopy. Under conscious sedation and local anaesthesia, single port Thoracoscopy was performed by the Jacobe's technique. Pleural space was entered after draining around one litre of hemorrhagic fluid under controlled suction. Lung collapsed partially and few adhesions could be seen. Parietal Pleura revealed sheets of yellow granular nodules Fig. 2. Biopsy was taken and haemostasis attained. Histopathology of the nodule showed evidence of Congo red positive abundant amyloid material Fig. 3. Intercostal tube drainage was put. Her CT chest taken prior to Thoracoscopy showed massive pleural effusion on the left with minimal effusion in the right side shown by white bold arrows in Fig. 4.

Her post thoracoscopic chest X-ray PA showed evidence of cardiomegaly and bilateral costophrenic blunting supporting the pre procedural CT evidence of bilateral effusion Fig. 5.

In view of her gross cardiomegaly a cardiology consultation was taken and the echocardiogram done subsequently did not reveal any evidence of pericardial effusion or cardiac amyloidosis. Hence she was managed conservatively with intercostal tube drainage, diuretics, hydrolazine, cilnidipine, weekly erythropoietin, paren

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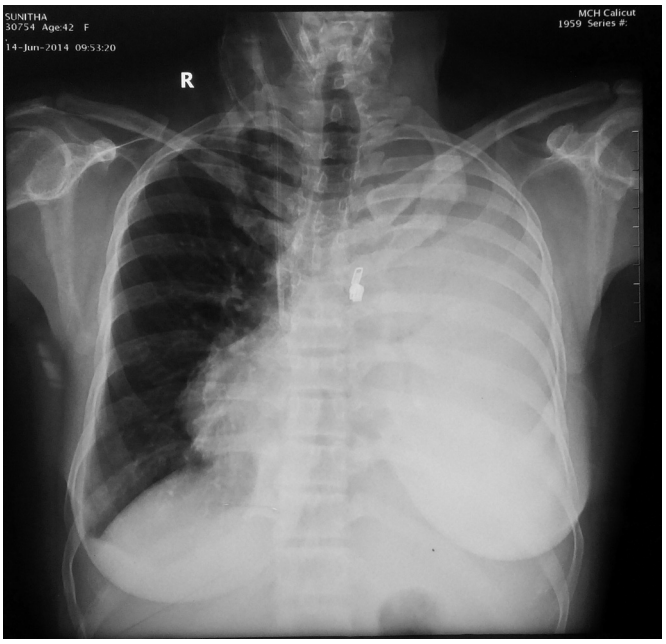

Fig. 1. Chest X-ray showing left massive effusion.

teral iron supplementation, alternate day haemodialysis, and oral sodabicarbonate. She continued to have drainage which made pleurodesis not possible and during the course of her stay in the pulmonary medicine unit she started developing more effusion on the right side Fig. 6.

Hence she was presumed to have primary systemic amyloidosis with possibly bilateral pleural deposits on the basis of thoracoscopic biopsy on the left side and renal amyloid diagnosed on renal biopsy. She was transferred back to the parent nephrology unit for a combination therapy of Bortezomib, Lenalidomide and dexamethasone along with hemodialysis. Other agents tried with varying

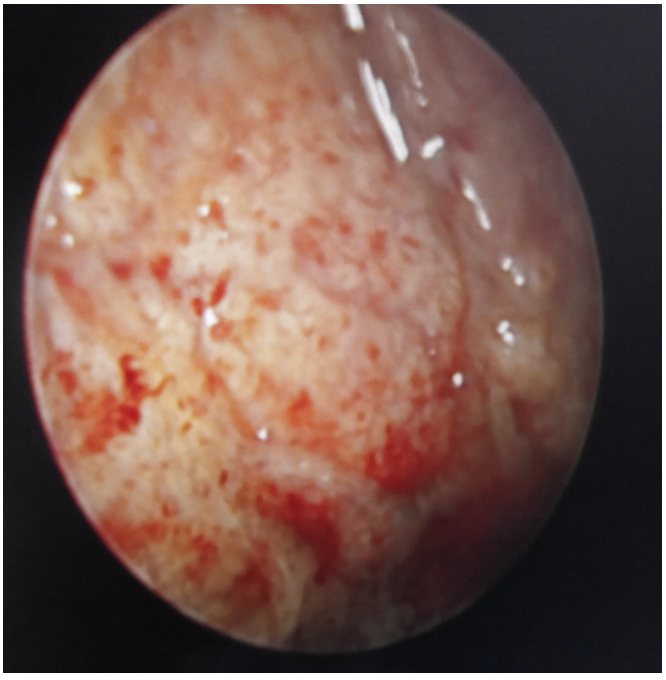

Fig. 2. Thoracoscopic view of amyloid deposits.

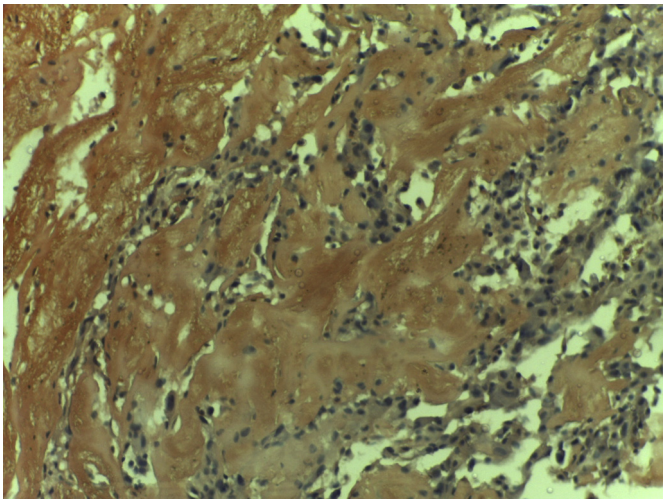

Fig. 3. Congo red staining positive material.

success in stem cell transplant ineligible primary systemic amyloidosis patients includes antimetabolites like cyclophosphamide, vincristine, adriamycin or proteasome inhibitors and immunomodulatory agents which includes bortezomib, melphalan,colchicine, thalidomide and pomalidomide.

Discussion

Amyloidosis is a rare disorder with a potential for multisystem involvement, characterized by extracellular deposition of different proteins as insoluble beta pleated sheets resulting in disruption of the function of the organ affected. Amyloidosis had been classified over the years based on the site of deposition and presence or absence of other diseases [1]. The term “Generalized” or “Systemic” had been used to describe deposition in multiple anatomic sites and “localized” used to describe deposition in one anatomic site. The term “secondary” used to describe patients with coexistent

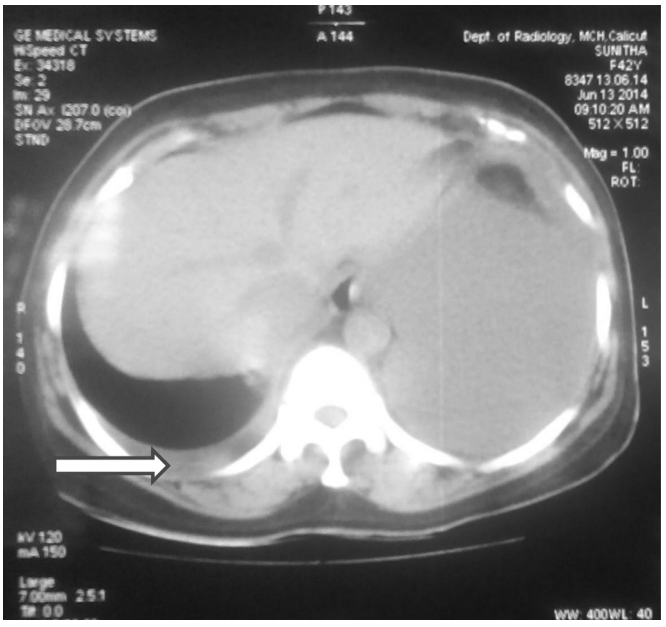

Fig. 4. CT chest showing (L) sided massive effusion and right sided minimal effusion (Bold arrow).