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Clinicoradiological Findings In A Patient With Fibrosing Mediastinitis And Anti-Phospholipid Antibody Syndrome

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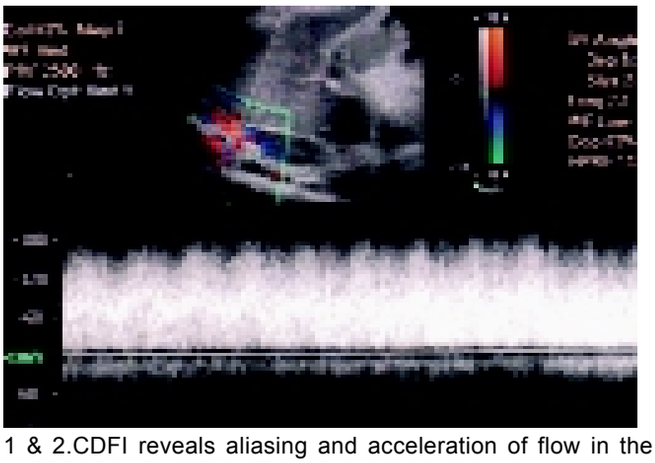
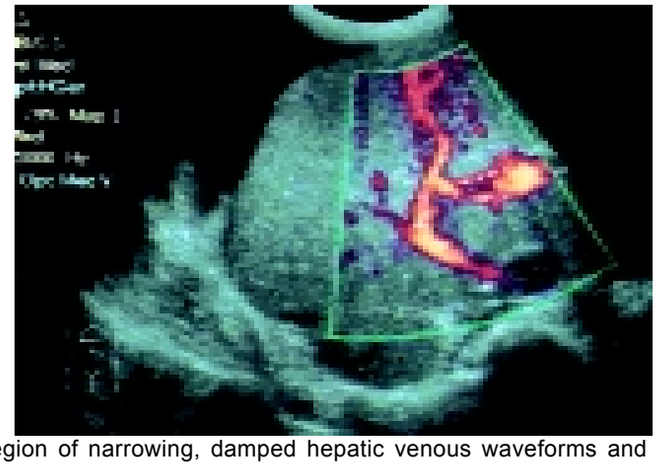
INTRODUCTION

Fibrosing mediastinitis is a chronic inflammatory process caused by proliferation of acellular collagen and fibrous tissue within the mediastinum. Most cases are idiopathic, although many cases are attributed to an abnormal immunologic response to various infections like Histoplasma capsulatum, Mycobacterium tuberculosis etc. Recently, the term idiopathic fibroinflammatory lesion of the mediastinum has been suggested to replace the present term fibrosing mediastinitis. Although, fibrosing mediastinitis has been associated with a multiplicity of clinical syndromes and diseases, the occurrence of fibrosing mediastinitis and superior venacaval obstruction together has not been described in the setting of anti-phospholipid syndrome before. A case of hypercoagulability disorder (anti-phospholipid syndrome) associated with fibrosing mediastinits and superior cava vein obstruction is described herein in which imaging played a pivotal role in the detection of various complications of the disease.

CASE REPORT

A 35 year old female presented to the emergency department with progressively increasing abdominal distension, facial swelling since a month and recent onset breathlessness. A chest radiograph showed non-specific mediastinal widening with bilateral pleural effusions. Ultrasound of the abdomen revealed gross ascites and hepatomegaly with disproportionate enlargement of left lobe and the caudate. Doppler flow imaging showed narrowing of the intrahepatic IVC by a non-occlusive hyperechogenic thrombus leading to aliasing and flow acceleration adjacent to the thrombus . The normal modulations of the venacava and hepatic waveforms were completely suppressed.(Fig.1) Multiple abnormal intrahepatic collaterals were also seen.(Fig.2) These findings were highly suggestive of Budd-chiari syndrome. The patient subsequently developed acute renal failure and was scanned again to rule out renal vein thrombosis. Ultrasound confirmed extension of the thrombus into bilateral renal veins. However this could still not completely account for some of the patients other symptoms like facial puffiness and bilateral pleural effusions. Therefore CT of the chest and abdomen was performed and revealed diffusely increased attenuation of mediastinal fat in absence of any well defined mass / abscess / collection.(Fig. 3) The superior venacava was not seen and instead a prominent azygous vein and





1 & 2.CDFI reveals aliasing and acceleration of flow in the region of narrowing, damped hepatic venous waveforms and multiple intrahepatic collaterals.

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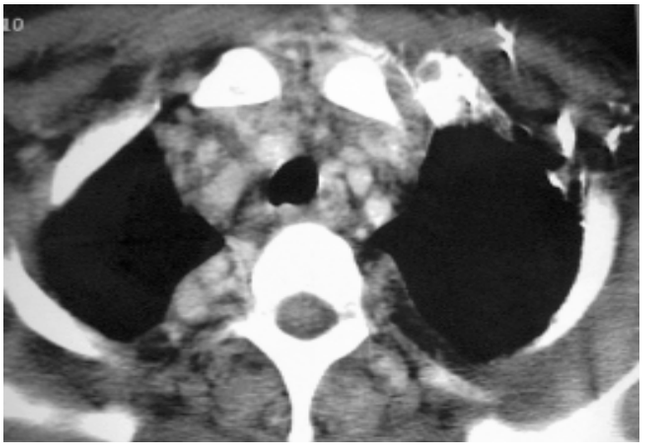
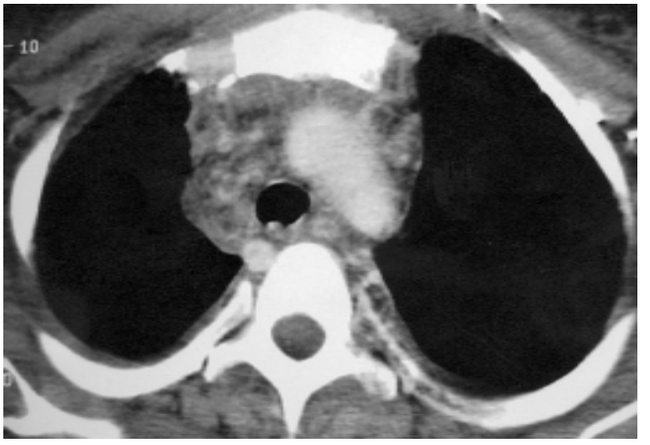
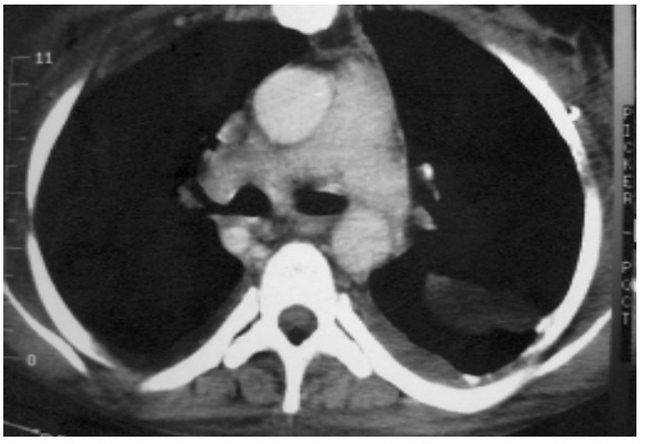
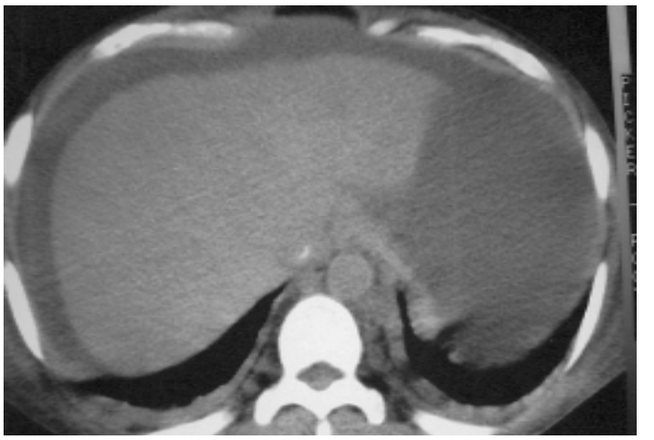
multiple venous collaterals were seen within the mediastinum. Bilateral pleural effusions and pleural calcification were also confirmed. (Fig. 4 & 5) Scans through the upper abdomen showed a calcific focus in the intrahepatic inferior venacava likely representing a calcified thrombus or web. (Fig.6) The patient also underwent upper GI endoscopy which revealed mid - esophageal varices and features of florid portal gastropathy .







3, 4 ,5. Contrast material-enhanced CT scan (mediastinal window) shows increased attenuation of mediastinal fat; non-visualisation of the superior vena cava, enhancement of multiple mediastinal collateral veins and prominent azygous vein; Also note bilateral pleural effusions and pleural calcification on the left.



6. Non-contrast CT confirms the presence of calcified thrombus in intrahepatic IVC

The patients history and clinical profile were reviewed again and it was found that she had been hospitalized in the past for investigation of multiple recurrent mid trimester abortions. She had been diagnosed as having anti-phospholipid syndrome; however the patient had not been compliant with her anti-coagulant medications. During her current hospitalization, the patient refused endovascular management for the treatment of SVC and IVC obstructions and treatment was initiated with oral anticoagulation, corticosteroids and diuretics. She subsequently had a downhill clinical course and succumbed to her disease.

DISCUSSION

Patients with fibrosing mediastinitis are usually young and present with signs and symptoms of obstruction or compression of the superior vena cava, pulmonary veins or arteries, central airways, or esophagus. The most common presenting complaints include cough, dyspnea, recurrent pulmonary infection, hemoptysis, and pleuritic chest pain. The radiographic findings of fibrosing mediastinitis in an individual patient depend on the mediastinal structures involved. Although chest radiographs of patients with fibrosing mediastinitis usually appear abnormal, the findings can be quite subtle and the extent of mediastinal involvement is frequently underestimated on the basis of these findings (1)

Patients with superior vena cava obstruction may have bilateral widening of the superior mediastinum due to engorged collateral veins . Involvement of the central airways can result in segmental or lobar atelectasis or recurrent pneumonia in the affected portions of lung. Less commonly, actual narrowing of the affected airways is seen. The area of narrowing usually occurs at the level of the carina and in the majority of cases involves both main bronchi. Pulmonary arterial obstruction is typically unilateral and can result in an appreciable diminution in size and quantity of vessels and localized regions of

oligemia. Pulmonary venous obstruction manifests radiographically with peribronchial cuffing, septal thickening, and localized edema. Both pulmonary arterial or venous obstruction can also cause pulmonary infarcts ; pleural effusions are uncommon in this disease.(2) There are two types of fibrosing mediastinitis: focal and diffuse . The focal type usually manifests on computed tomographic (CT) as a localized, calcified mass in the paratracheal or subcarinal regions or in the pulmonary hila. The diffuse type manifests as a diffusely infiltrating, often noncalcified lesion that affects multiple mediastinal compartments. The first type, is more common and is probably caused by a fibroinflammatory response to H capsulatum or M.Tuberculosis antigens in a genetically susceptible population. This type usually results in relatively focal mediastinal or hilar fibrosis with extensive calcification. The second, and much less common, type of the disease occurs idiopathically and results in diffuse, multicompartmental fibrosis that does not usually calcify (3).

On MR imaging, fibrosing mediastinitis manifests as a heterogeneous, infiltrative mass of intermediate signal intensity on T1; it has a more variable appearance on T2-weighted images with regions of both increased and markedly decreased signal intensity seen in the same lesion.

Esophageal varices (downhill type) can be seen in a few cases of superior vena cava obstruction (4) As our patient was asymptomatic for esophageal varices, endoscopy was done in our case to document the presence and extent of varices and plan future management.

The fluorine-18 fluorodeoxyglucose positron emission tomographic findings of fibrosing mediastinitis have been reported. Although the lesions are mostly hypometabolic, focal areas of increased metabolic activity can be seen (5).

To conclude, we have tried to describe the clinicoradiological findings in a patient with anti-phospholipid syndrome and fibrosing mediastinitis. While fibrosing mediastinitis has been reported to be the most common benign cause of the superior vena cava syndrome(6) and Budd-Chiari syndrome due to IVC or hepatic venous thrombosis is a known manifestation and/or complication of anti-phospholipid syndrome, the occurrence of fibrosing mediastinitis and SVC obstruction in a hypercoagulable condition like anti-phospholipid syndrome has not been reported before.

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