Sclerosing mediastinitis, also known as fibrosing mediastinitis, is a rare condition characterized by proliferation of fibrous tissue usually dense in the mediastinum. Despite this condition is benign, it is associated to a higher morbidity due to its obstructive nature, and less commonly, to mortality. It is idiopathic in most cases, also being possibly related to tuberculosis, histoplasmosis, and other granulomatous diseases, such as sarcoidosis, fungal and autoimmune diseases, radiotherapy and retroperitoneal fibrosis. Affected patients are usually young and present with signs and symptoms due to obstruction of vital mediastinal structures, such as esophagus, airways and large vessels. The present study reports the case of a 19-year-old girl with chest pain, exertional dyspnea and sleeping in squatting position with no clinical evidence of granulomatous disease. After extensive workup her CT scan chest revealed a dense mediastinal mass with calcifications that was approached by CT-Guided biopsy. Histopathological study demonstrated an etiologically undefined chronic inflammatory pattern compatible with sclerosing/fibrosing mediastinitis.

Case Report
A 19-year-old previously healthy girl presented with complaints of exertional dyspnea for 8 months, low grade fever and sleeping in squatting position for 6 months, retrosternal chest discomfort for 1½months. She visited local doctors multiple times during these 8 months but condition remained static. Fever subsided temporarily but tended to recur. A day came when she landed in private hospital with worsening dyspnea, from where she was referred to PIMS cardiology with suspicion of pericardial effusion. In November 2011 she presented to cardiology OPD and was admitted with diagnosis of pleura-pericardial effusion. Fluid aspirated from both cavities was exudative and lymphocytic with no growth. She was discharged on ATT and steroids. She visited OPD again on 16/12/11 without any improvement. She was referred to pulmonology with massive left sided pleural effusion that was detected on CXR. Diagnostic pleural aspiration showed transudative lymphocytic picture followed by pleural biopsy. During the procedure, patient collapsed (hypotension). She was shifted to CCU where urgent echocardiography revealed moderate to severe pericardial effusion with early tamponade effect. Pericardiocentesis of about 2 L fluids was performed. In January 2012 she was shifted to pulmonology department for evaluation of non-responding pleuropericardial effusion despite on ATT. She had facial puffiness, increased neck circumference and signs of left sided pleural effusion. H/P report of pleural biopsy showed marked infiltration by polymorphous lymphoid cells and histiocytes without any evidence of granulomas formation (suggestive of chronic inflammation).

No definitive diagnosis was established till January 2012 so pleural biopsy was repeated, H/P showed atypical infiltrate comprising of round to oval cells with hyper chromatic nuclei and scanty cytoplasm. Foci of overlying reactive mesothelial cells. Immunohistochemistry was advised due to suspicion of non-Hodgkin’s lymphoma that came to be negative. αFP and βhCG levels were normal. CECT chest showed a homogenous density mass in anterior mediastinum encasing left main stem bronchus and left sided pulmonary vessels (Fig 1). CT guided trucut biopsy was done that revealed “abundant pauci-cellular densely sclerotic hyalinized collagenous tissue infiltrating and obliterating adipose tissue. Scattered aggregates of mature looking small lymphoid cells were seen in between the fibrous tissue. No atypical lymphoid cells were seen. Fungal Stain and Immunohistochemistry were negative. Highly suggestive of Sclerosing Mediastinitis (idiopathic mediastinal fibrosis)”. She was started on 30 mg of prednisolone, low dose warfarin and 40 mg of frusemide. Chest intubation was done, left hemothorax was drained followed by pleurodesis. Pericardiocentesis was also repeated. Thoracic surgeon opinion was taken but, in view of diffuse and bilateral involvement, conservative management was advised. Patient was discharged and advised follow up after two weeks and planned to repeat CECT chest after 6 months to asses for the development of collaterals.
and interstitial pattern are also observed. MRI is useful in the determination of the extent of disease and in the preoperative assessment, usually demonstrating an image with intermediate signal intensity on T1-weighted sequences and heterogeneous signal intensity on T2-weighted sequences due to fibrotic nature and presence of calcifications.

The course of the disease is unpredictable, sometimes with spontaneous remission, and other times with exacerbations of symptoms. Approximately 30% of patients die from complications resulting from compression and fibrosis. Bilateral or carinal involvement represents the worst prognosis. Biopsy of the affected lymph nodes or mediastinal tissue is needed for definitive diagnosis. Treatment includes the use of corticosteroids, surgical approach for excision of tissue and local management of obstructive complications by using stents.

Despite , it is uncommon, sclerosing mediastinitis should be considered in almost all cases of mediastinal widening at chest radiograph, primarily as its relation to granulomatous diseases such as tuberculosis that is endemic in our country. The investigation must include CECT chest and workup for granulomatous diseases. Biopsy with histopathological study confirms the diagnosis. Regular follow-up must be conducted with these patients for monitoring of development of possible complications of the disease.

References