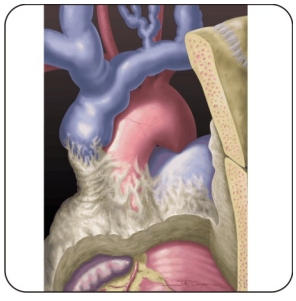
CASE REPORT PROF-1364

SUPERIOR VENA CAVA OBSTRUCTION;
PRESENTATION OF IDIOPATHIC MEDIASTINAL FIBROSIS.



DR. ILTIFAT SULTAN
Associate Prof. Chest Diseases
Dept. of Chest Diseases
Punjab Medical College, Faisalabad

SUMMARY... iltifatsultan@hotmail.com Idiopathic mediastinal fibrosis is a disease of unknown etiology. The cause is probably an abnormal fibro-proliferative response within the mediastinum. It can result in clinical syndromes due to invasion and compression of mediastinal structures.

A case of young male with idiopathic mediastinal fibrosis and superior vena caval obstruction is described.

INTRODUCTION

Idiopathic mediastinal fibrosis is also known as fibrosing mediastinitis is a chronic inflammatory process caused by proliferation of acellular collegen and fibrous tissue within the mediastinum. This can result in clinical syndromes due to invasion and compression of various structures of the mediastinum, the superior vena cava is often involved, but it can also involve pulmonary arteries, veins and tracheobronchial tree.

Radiology, CT, MRI features may be helpful in diagnosis, accurate diagnosis rests on histological confirmation. Medical therapy is disappointing and surgery has limitations.

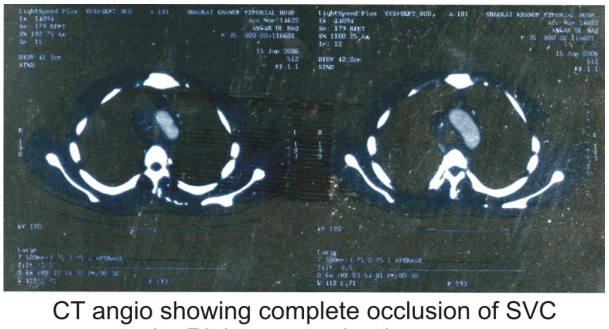
CASE PRESENTATION

35 years old male teacher, non smoker presented with puffiness of face, worse after getting up in the morning for the last one year. He also complained of exceptional dyspnea and a dull chest discomfort for the same length of time. Productive cough with scanty sputum for three months and heaviness of head. No H/o fever and heamoptysis. No H/odysphagia nausea, vomiting. No H/o hoarseness, dependent edema or palpitations. Nothing was significant in past medical history. Patient had anti TB therapy for one year in recent past, but no history of drug intake. Personal and family history were non contributory. The patient was never exposed to birds or animals. He had suffered from conjunctivitis. Jugular veins were distended and non pulsatile. Distended veins were also visible on anterior aspect of neck and anterior chest with flow away from mediastinum. Respiratory system examination was unremarkable.

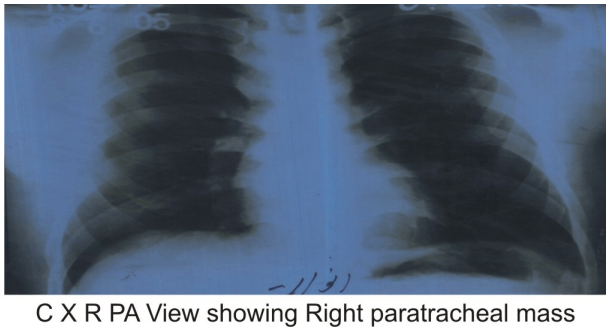
INVESTIGATIONS

Full blood count, renal, liver and thyroid function tests

were normal. ECG, Echocardiography and Carotid Doppler studies on both sides of the neck were normal. X Ray chest revealed right paratracheal mass. Sputum for AFB, cytology, and fungal culture were negative, C-T angiogram through chest and upper abdomen showed right paratracheal mass most probably infiltrating the superior vena cava (SVC) with more than 99% occlusion, reconstitutes distally just above the atrium, inferior vana cava (IVC) was patent. Multiple collaterals draining along the anterior mediastinum and through azygous system were seen. The periclavicular and upper abdominal para-aortic recesses were clean.



CT angio showing complete occlusion of SVC by Right paratracheal mass



C X R PA View showing Right paratracheal mass

No focal hepatic or splenic abnormality was seen. Biliary tract, gall bladder, pancreas, adrenals and kidneys were normal. Anterior mediastinotomy with biopsy of mass showed dense hyalinized keloid type collegan fibers with Ch. Inflammation. There were interspersed clusters of Chronic inflammatory cells. No malignant lesion seen.

The patient was given steroids for 6 months tapering the doses with no clinical benefit. Because of distressing symptoms related to superior vena cava (SVC) obstruction, he was offered surgical treatment, but the patient refused.

DISCUSSION

Idiopathic mediastinal fibrosis represents a Ch. Inflammatory process, resulting in the deposition of dense fibrous tissue containing an infiltrate of predominantly plasma cells with some lymphocytes, polymorphs and fibroblasts typically located in the superior mediastinum near the bifurcation of trachea as well as in the pulmonary hila and often extending in adjacent tissues.

There is no gender predilection and it affects all ages. Asymptomatic patient usually presents with an abnormal chest X-Ray specifically showing hilar and or mediastinal mass in the right paratracheal region. Other patients present with various combinations of symptoms including cough, dyspnea, chest pain, haemoptysis, dysphagia, neck vein distention, chest wall varicosities and headaches all representing this clinical syndrome.

Currently there is no definite treatment available. When etiology is unknown, steroids are used but these does not appear to be beneficial, median sternotomy with graft from superior vena cava (SVC) and Innominate vein to right atrium are recommended.

REFERENCES

1. Light AM. **Idiopathic filnosis of mediastinum a discussion of three cases and review of literature** J Clin Patrol 1978; 31:78-88.

2. Kalweit G Huwerg, Strauber, et al, **Mediastinal Compression syndromes due to Idiopathic fibrosing mediastinitis.** Report of three cases and review of literature Thorac cardiovasc surg 1996; 44:105-9.

3. Kinugasas, Tachibana, Kawakami M, et al, **Idiopathic mediastinal fibrosis report of a case,** surg Today 1998;28:235-8.

4. Schowenger dt CG, Suycmotor, Miam FB, **granulomatous and Fibrosis mediastinitis a review and analysis of 180 cases** J. Thorac cardiovasc surg 1969;57:365-79.

5. Yacoub MH, Thompson VC, **Chronic Idiopathic pulmonary hilar Fibrosis a clinicopathological entity;** Thorax 1971:2:365-75.

6. Grocia JM, Ramirez R, Bacos J, **Technique for reconstruction of SVC in fibrosing mediastinitis** J Thorac cardiovasc surg 1973;65:547-51.