Mediastinal Lipomatosis with Dyslipidemia: Cause of Dyspnea

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Abstract
We report a case who presented to us with dyspnea and was found to have ML in the absence of steroid use, diabetes, Cushing syndrome and obesity. This case provides an important differential diagnosis in a patient presenting with dyspnea widened mediastinum and cardiomegaly. This is probably among very few cases of ML with dyslipidemia.

Introduction
Mediastinal lipomatosis is a rare benign condition which causes mediastinal widening. It is characterized by deposition of adipose tissue within the mediastinum causing distortion of mediastinal silhouette. Usually it is secondary to the steroid intake, diabetes mellitus, Cushing’s syndrome and exogenous obesity rarely it can be idiopathic also. It is more common in middle aged males and is associated with alcohol abuse. CT and MRI can decisively diagnose this benign condition, which also help in deciding treatment options. We hereby report a case of mediastinal lipomatosis, who presented to us with complaint of dyspnea.

Case Report
A 60-year-old female, came to our OPD with the complaint of dyspnea for last 3 months. It was gradual in onset and generally on exertion. She was not in acute distress. There was no chest pain or coughing. There was no past history of diabetes, hypertension, steroid intake, tuberculosis or chest pain on exertion. On clinical examination, she was overweight with weight 64 kgs (BMI was 25 kg/m²). Waist-hip ratio was 0.9. Xanthelasmas were present over the upper eyelids (Figure 1). There was no lymphadenopathy. Her resting pulse rate was 94/min, regular, blood pressure was 130/80 mm of Hg and respiratory rate was 20/min. Chest was clear, without any added sounds. Cardiovascular system was normal clinically. Her abdomen was soft and pendulous, with no organomegaly. CNS examination was normal. Musculoskeletal system was also normal. Both breasts were normal on palpation.

On investigations, her hemoglobin was 11.5 gm%, total leucocyte count was 6300/cmm: neutrophils 73%, lymphocytes 24% and eosinophils 3%, without any abnormal cells. Her liver and renal function tests were within normal range. Her fasting blood sugar was 80 mg/ml and post prandial blood sugar was 118 mg/ml. HBsAg and HIV was negative. TSH level was 2.8 mU/L. Serum cholesterol was 280 mg% and serum triglyceride was 210 mg%. USG of abdomen showed fatty liver. Spirometry revealed mild restriction. ECG and 2D Echo were normal. Chest X-ray showed mediastinal widening and cardiomegaly (Figure 2).

Fig. 1: Upper eyelid showing xanthelasma

Fig. 2: Chest x-ray showing mediastinal widening and cardiomegaly
To evaluate the causes of mediastinal widening, CT thorax was done which revealed mediastinal widening due to presence of fat. Fat deposition was also noted in epicardium and pericardium giving impression of cardiomegaly (Figure 3). There were no lymph nodes. Histopathology was not required as CT scan is definitive for diagnosing fat.

Finally the diagnosis of mediastinal lipomatosis with dyslipidemia was made. Patient was put on statins and was advised for diet control along with suitable regular exercises. After losing 4 kgs of weight, patient’s dyspnea improved. She was lost to further follow-up.

Discussion

Mediastinal lipomatosis is a rare benign condition characterized by deposition of mature adipose tissue within the mediastinum, distorting the mediastinal silhouette. Conditions causing mediastinal lipomatosis are Cushing’s disease, steroid use, alcoholism, obesity, Diabetes mellitus. Other causes are hyperuricaemia, hypothyroidism, hypertension, hyperlipidemia, thyroid cancer. It can be seen with or without obesity as seen in this patient who was only overweight, but had dyslipidemia.

Patients may remain asymptomatic or may present with cough, chest pain or dyspnea. Dyspnea in our case was due to extensive ML causing narrowing of trachea. Physical findings may be normal, or have decreased breath sounds, or signs of associated condition can be present. Like in our case, xanthelasma were seen over upper eyelid towards inner canthus.

In majority of the cases, ML was an incidental finding as mediastinal widening on chest radiograph. ML can present with certain clinical features. Kashikar et al report a case presenting with progressive shortness of breath who was ultimately found to have segmental atelectasis of the lung related to ML. A rare complication of ML is superior vena cava compression, which can cause difficulty with central venous catheterization. A more significant complication is laryngeal compression secondary to excess adipose tissue in the mediastinum leading to airway compromise and right ventricular outflow tract obstruction.

The study of choice for diagnosis of ML is CT Scan or, less often, MRI. CT Scan is definitive in diagnosing lipomatosis as density may not be as pronounced as other masses like Lymphoma, Thymoma since it is composed of fat. Initial CXR will show a widened mediastinum with increased lucency. A follow up CT reveals a collection consistent with fat (attenuation of 50 to 100 Hounsfield units). The adipose tissue can extend from the superior mediastinum to the diaphragm and may involve the heart and lungs.

ML is an uncommon cause of mediastinal widening so firstly all the acute causes must be excluded. It includes trauma, aortic dissection, esophageal rupture and mediastinitis. If there is mediastinal widening on CXR, it should always be followed up with immediate CT, MRI and echocardiogram. Critical warning signs can be elicited by taking proper history and on physical examination. Notably, aortic dissection often manifests as sudden onset of sharp, tearing ripping pain in the chest or back with maximal intensity at onset leading to absent pulses, >20 mmHg difference in blood pressure between the two arms. Other critical causes of mediastinal widening include anterior mediastinal masses such as lymphoma and thymoma.

In our case, patient was not in acute distress.

Management

Management of ML depends on whether the patient is symptomatic or not and the conditions associated with it. ML is usually asymptomatic and no treatment is necessary. If the patient is obese, then the weight loss is strongly recommended. Tapering the doses of steroids may improve symptoms and can reverse radiological findings. In the case presented by Nguyen et al, surgical resection was done to relieve symptoms of dyspnea caused by ML. Treat the associated disease if it is the cause of ML.

Conclusion

Mediastinal lipomatosis is a rare but benign cause of mediastinal widening on CXR in contrast to medical emergencies and malignancies. Symptoms caused by mediastinal lipomatosis include dyspnea, cough, chest pain and arrhythmias, but majority of the people are asymptomatic. The diagnosis is made by CT scan once acute conditions have been ruled out. Since the prevalence of obesity is increasing, it is important to understand the presentation, diagnosis, and to recognize this entity as a cause of mediastinal widening, as by doing so, we can avoid many unnecessary investigations and invasive procedures.

References