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Natasha Fonseca, MD

Thomas Jefferson University Hospitals, Natasha.Fonseka@jefferson.edu

Ewa Ruel, MD

*Thomas Jefferson University Hospitals*Follow this and additional works at: <http://jdc.jefferson.edu/tmf> Part of the [Medicine and Health Sciences Commons](#)[Let us know how access to this document benefits you](#)

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A BENIGN CAUSE OF WIDENED MEDIASTINUM: A CASE OF MEDIASTINAL LIPOMATOSIS

Natasha Fonseka, MD, Ewa Ruel, MD

Introduction

We report a patient with a widened mediastinum secondary to a rare and benign condition known as mediastinal lipomatosis (ML). ML is caused by accumulation of adipose tissue within the mediastinum. Case reports associate ML with obesity, diabetes, Cushing's syndrome, steroid use while other cases remain idiopathic.¹⁻⁴ We report a patient with Glioblastoma Multiforma (GBM) on chronic steroids, who was found to have ML on imaging despite lack of obesity or Cushing's syndrome. This case provides significant educational benefit in approaching a patient with a widened mediastinum.

Case Presentation

A 59-year-old male with a history of recurrent GBM status post stereotactic radiation and ongoing chemotherapy presented to the hospital with lethargy, altered mental status and expressive aphasia consistent with ongoing progression of his malignancy. The patient was not obese (weight 72.1 kg; height 6'1", body-mass index 21), non-diabetic, and had been on a daily dose of 2-8mg of dexamethasone intermittently (equivalent to prednisone 25-50mg daily) for approximately a year and half for GBM.

On admission, the patient was in no acute distress and denied abrupt or sharp onset of thoracic or abdominal pain. Physical

exam revealed a well healed scalp scar from previous GBM resection, expressive aphasia, and chronic right sided weakness. Despite chronic steroid use, there were no physical manifestations of Cushing's syndrome such as round face, fat pad, or purple stria. He had equal blood pressures in both arms and intact pulses bilaterally. The patient's fasting glucose was within normal limits.

A plain chest radiograph demonstrated widening of the mediastinum, which was new compared to a study 6 months earlier (Figure 1). A follow up chest CT with contrast showed anterior-superior mediastinum lipomatosis (Figure 2). The pericardium and other surrounding structures were not involved. Mediastinum and hila were within normal limits on chest CT performed 7 months prior. The diagnosis of ML was made by CT. Since the patient remained asymptomatic, no further workup was required. Although steroids have been associated with ML, Neuro-Oncology deemed the benefits of steroids outweighed the risks. Therefore, the patient continued steroid treatment and was discharged home with Neuro-Oncology follow up.

Discussion

Mediastinal Lipomatosis is a benign cause of mediastinal widening secondary to mature adipose deposition. Although our patient remained asymptomatic with ML, symptoms

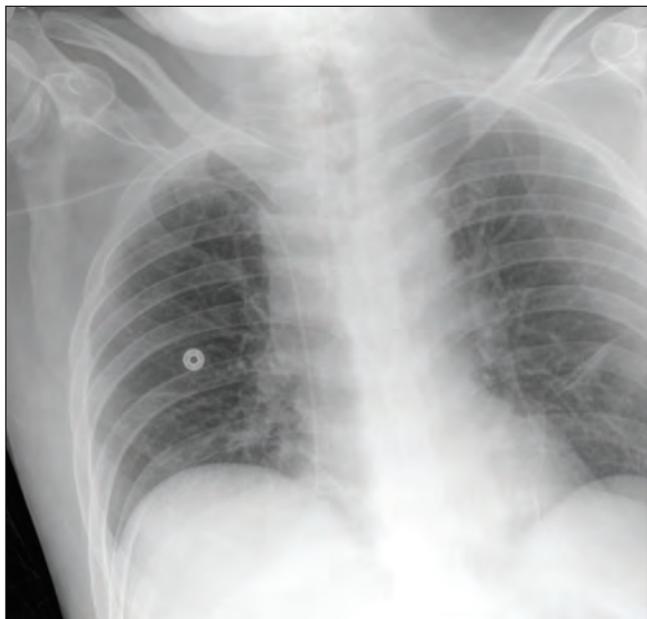


Figure 1. CXR: Apparent widening of the superior mediastinum. Mild atelectasis in the left midlung is noted.

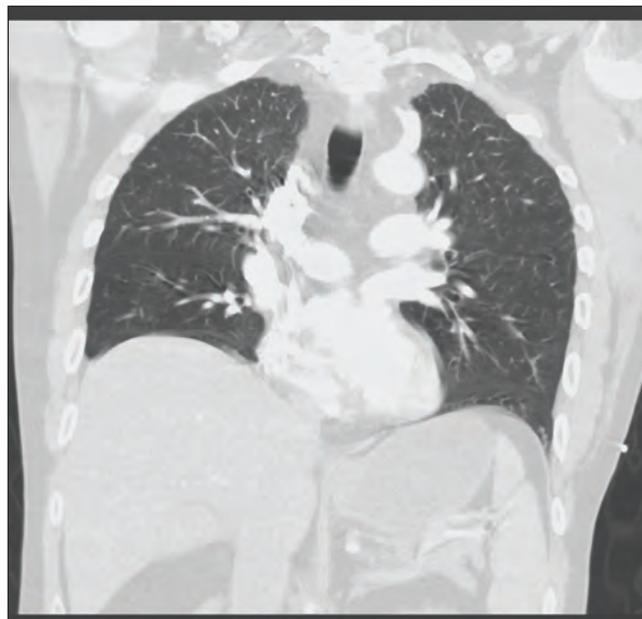


Figure 2. CT scan with contrast: Anterior-superior mediastinum lipomatosis.

may include dyspnea, cough, and chest pain.³⁻⁵ Physical exam findings range from a benign exam to decreased breath sounds, obesity, or associated signs of Cushing's syndrome.^{3,4} Unlike most case reports reviewed, our patient did not have physical exam signs of obesity or steroid excess. The patient, however, was on chronic steroids, which may be associated with ML.^{3,4} Though the time course is unknown for development of fat accumulation, this patient appears to have developed ML within 6-7 months.

In most case reports, ML is identified by an incidental mediastinal widening on chest radiographs. Although ML is considered benign, it does have certain clinical implications. Kashikar et al report a patient presenting with progressive shortness of breath who was ultimately found to have segmental atelectasis of the lung related to ML.¹ In the case presented by Peek et al, ML caused right hemidiaphragm paralysis secondary to phrenic nerve compression and mimicked cardiomegaly on CXR.⁶ ML can also cause low voltages on electrocardiograms.² A rare complication of ML is superior vena cava compression, which can cause difficulty with central venous catheterization.⁷ In one case, a primary mediastinal large B-cell lymphoma was found within mediastinal lipomatosis.⁸ 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.⁹⁻¹⁰

Treatment

Treatment of ML depends on the degree to which it affects the patient. ML is usually asymptomatic and no treatment is necessary. Often, the patient is obese, and weight loss is strongly recommended.^{2,16} Tapering steroids may improve symptoms and reverse radiological findings.⁵ In the case presented by Nguyen et al, surgical resection was required to relieve symptoms of dyspnea caused by ML.⁴

Differential

ML is an uncommon cause of mediastinal widening and acute causes must be excluded first. Emergencies include aortic dissection, esophageal rupture, trauma, hemorrhage or mediastinitis.¹² Mediastinal widening on CXR can be followed up with immediate CT, MRI and Echocardiogram. Critical warning signs can be elicited on history and physical exam. 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 BP mmHg difference in blood pressure between arms, and possible neurological abnormalities.^{11, 12} Other critical causes of mediastinal widening include anterior mediastinal masses such as lymphoma and thymoma, which can be distinguished from benign lipomatosis by imaging.^{13, 14}

Imaging

The study of choice for diagnosis of ML is CT or, less often, MRI. 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. Interestingly, CT may also show incidental heart involvement with a characteristic dumbbell shape within the interatrial septum. However, of those studied with interatrial septum lipomatosis, no patient had evidence of electrocardiogram or other cardiac abnormalities, again emphasizing the benign nature of this diagnosis.¹³⁻¹⁵

Conclusion

In contrast to medical emergencies and malignancy, mediastinal lipomatosis is a rare but benign cause of mediastinal widening on CXR. Symptoms caused by mediastinal lipomatosis include dyspnea, cough and arrhythmias, but most people are asymptomatic. The diagnosis is made by CT once acute conditions have been ruled out. Since the prevalence of obesity and use of steroids is increasing, it is important to understand the presentation, diagnosis, and treatment of mediastinal lipomatosis as we will likely see more cases of ML in the future.

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