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Unusual presentation of mediastinal mass which grew slowly over 5 years and became aggressive which transformed into high grade lymphoma and presented as hypercalcemia

ABSTRACT

A 62-year-old male patient, without any comorbid conditions, presented with an asymptomatic, slow-growing mediastinal mass since the past 5 years that became aggressive and transformed into a high-grade lymphoma, with metastasis and presented as hypercalcemia.

Key words: Mediastinal mass, PET Scan, Hypercalcemia, Diffuse Large B cell Lymphoma (DLBCL)

INTRODUCTION

The mediastinum is an important segment of the thorax that contains vital intrathoracic structures such as the heart and great vessels, trachea and main bronchi, oesophagus, thymus, venous and lymphatic structures, and nerve tissue.

Mediastinal tumours can be caused by lymphoma (Hodgkin's disease and non-Hodgkin's), Thymoma (a tumour of the thymus), Thyroid mass (generally a benign growth, but can sometimes be cancerous), Oesophageal mass/ malignancy and Neurogenic tumours. The most common cause of posterior mediastinal tumours are nerve sheath neoplasms, ganglion cell neoplasms, and paraganglionic cell neoplasms. Approximately 70% of mediastinal neoplasms are benign like Lymphadenopathy (Tuberculous), Neuroeteric cyst and Aortic aneurysm. Thymomas are the most common mediastinal tumours. They represent one third of all anterior mediastinal tumours (1). Regardless of the diagnosis, all large mediastinal masses may cause compression or invasion of vital structures, like vascular structure, heart and pericardium causes hemodynamic decompensation, oesophagus involvement causes dysphagia and larger airways compression resulting in respiratory insufficiency. Almost 40% of people who have mediastinal tumours experience no symptoms. Most of the growths are often discovered on a chest X-ray which is performed for another reason. The tests most commonly used to diagnose and evaluate a mediastinal tumour are Computed tomography (CT scan), Ultrasound, 2-D echo emanation, Needle biopsy or aspiration under CT guidance, Bronchoscopy or Mediastinoscopy with biopsy.[1]

CASE REPORT

A 62-year-old gentleman, non-smoker and non-alcoholic, presented to the hospital with cough (dry), dyspnea on exertion, and diminished appetite since 2 months. He complains of cough which is not associated with expectoration and occurs mainly on lying down on the left side. No H/O hemoptysis. He also felt a mild and dull aching chest pain for the past 2–3 months. He complained of 8.5 kg weight loss over the past 2–3 months. He gives no history of any comorbid conditions such as diabetes mellitus, hypertension, ischemic heart disease, or thyroid dysfunction. There is no history of tuberculosis or bronchial asthma.

The patient was apparently alright 5 years back when he developed a mild dry cough, which did not respond to routine treatment. Investigations revealed a mediastinal mass on his X-ray chest [Figure 1]. Computed Tomography (CT) scan of the chest with contrast revealed 9 cms X 9.1 cms and extending 10 cms in cephalocaudal direction. He deferred CT scan-guided biopsy at that time (2015). Follow-up CT scan chest was performed after 1 year and there was a marginal increase in the size of the mediastinal mass. The patient did not agree to biopsy or removal by video-assisted thoracoscopy since he was only mildly symptomatic at that time.

Now after 5 years, examination revealed mild puffiness of the face, pulse 98/min, regular in rhythm, blood pressure 140/90 mm of Hg, raised and fixed JVP, and dilated veins over

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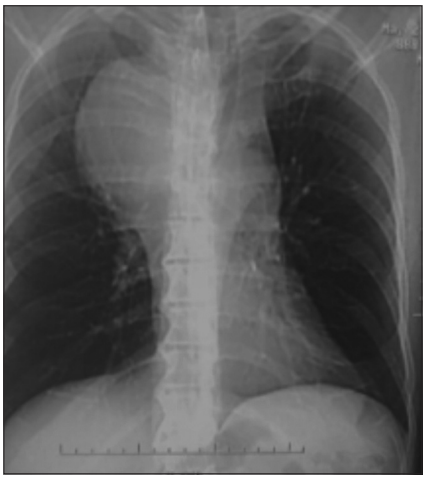

Figure 1: X-ray 21-05-2015

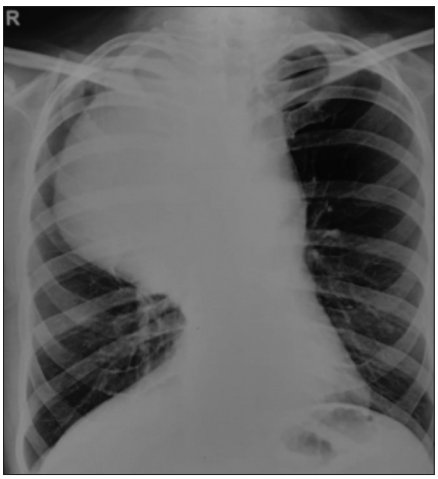

Figure 2: X-ray 20-03-2000

[Figure 3] the chest with blood flow from above downward (superior vena cava syndrome). Systemic examination revealed diminished air entry on the right side of the chest, and no other significant abnormality. Abdominal system examination revealed mild splenomegaly. No lymphadenopathy or hepatomegaly was detected. Cardiovascular examination did not reveal any murmur or foreign/added sound.

Investigations

Hb: 13 g/dL, Total leucocyte count: 6.83 X 10^9/L, platelet count: 291 X 10^9/L, RBS: 124 mg/dL, Se. Creatinine: 2.92 mg/dL which reduced to 1.5 mg/dL after treatment with intravenous normal saline and treatment of hypercalcemia. Blood Urea Nitrogen: 38 mg/dL, Na: 139 mEq/L, K: 3.6 mEq/L, Cl: 95 mEq/L, INR: 1.08. Se. Calcium: 15.2 mg/dL after 3 days treatment reduced to 9.9 mg/dL, Se. PO4: 3.6 mg/dL, HCO3: 28 mEq/L, ALT: 11.1 U/L, ALP: 60 U/L, total bilirubin: 0.46 md/dL, total protein: 6.77 g/dL, albumin: 4.0 g/dL, globulin: 2.77 g/dL, and urine routine: Protein:

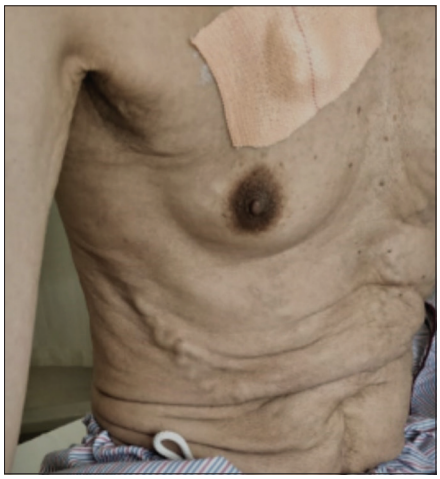

Figure 3: Dilated vein over chest wall / Biopsy mark

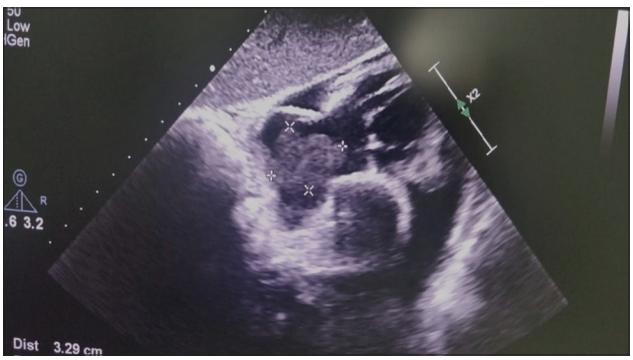

Figure 4: 2-D Echo showing Mass in the right atrium

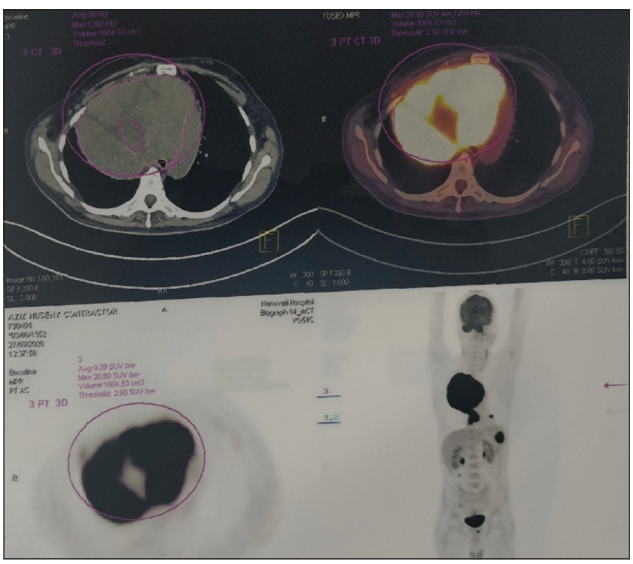

Figure 5: PET Scan showing mediastinal malignancy with multiple mets

trace, pus cells: 4-6/hpf, rest normal, triple "H" Negative, T3: 1.01 ng/dL, T4: 9.40 ng/dL, TSH: 3.8 mIU/L, Se. PTH: 8.49 ng/L (low), Se. Vit. D: 43.8 ng/mL (normal), and uric

acid: 10.7 mg/dL which reduced to 6.8 mg/dL after 2 days treatment.

Se. Protein Electrophoresis

No evidence of multiple myeloma, ECG is showing sinus tachycardia with a short QT interval (0.30 seconds) dated 25 March 2020, X-ray chest: Mediastinal mass, more on the right side, pulmonary function test: Mild obstruction with moderate restrictive disorder, and ABG: Mild hypoxia without retention of CO_2 [Figure 4]. 2-D Echo Examination: Large mass in RA (extending from SVC to RA). Normal left ventricular size and contractility. No pulmonary hypertension.

USG of Abdomen

A USG abdomen and pelvis are carried out on the March 9, 2020, a large 3.2 cm × 3.1 cm × 2.9 cm sized solid heterogeneous lesion was noted at the upper pole of splenic parenchyma with hypoechoic areas within -? Necrotic areas with peripheral vascularity were noted. Findings suggestive of indeterminate aetiology which may be neoplastic in origin. CT scan of chest, abdomen, and pelvis on September 25, 2015: CT findings are remarkable for a mass lesion in the middle mediastinum extending into the anterior mediastinum. It measures about 9.0 cm × 0.1 cm and has a cephalocaudal extent of 10.0 cms. This study is negative for significant mediastinal adenopathy, lung parenchymal/pleural abnormality, and interstitial lung disease or centrilobular opacities. Repeat CT scan chest (March 2020) revealed an increase in size from ~1.3 L of fluid (CT scan-2015) increased to 3.5 L.

PET Scan [Figure 5]

Profile reveals a large solid metabolically active mass lesion with a high retention index in the anterior mediastinum extending into the middle mediastinum, contiguous with a metabolically active lesion which is seen to extend into the right atrium of the heart. This is suggestive of a neoplastic process. A metabolically active nodular lesion with high retention index in the pericardium, overlying the right ventricle of the heart too, is of similar etiology/metastasis. Metabolically active focal splenic lesion is of similar etiology or could represent metastasis. Tiny metabolically active soft-tissue nodules lying closely opposed to the right psoas muscle at the level of L4, closely opposed to the right external iliac vessels, as well as the left levator ani sling too are of similar etiology.

Histopathology of Biopsy

Tru-cut biopsy from anterior mediastinal mass revealed a microscopic structure that showed atypical large lymphoid cells arranged in diffused sheets. Brisk mitotic activity was seen. Immunohistochemistry (IHC) revealed that the atypical lymphoid cells are diffusely positive for CD20, CD10, Bcl2, and focally positive for Bcl 6. These features are consistent with diffuse large B-cell lymphoma.

The patient's investigations revealed a mediastinal mass with superior vena cava syndrome. A renal study revealed increased creatine with hypercalcemia. Hypercalcemia was treated with rehydration with saline, steroids, and calcitonin. PET scan was suggestive of malignancy with metastasis. Lung biopsy was suggestive of large B-cell high-grade lymphoma. He was referred to a hematologist for chemotherapy and further management.

DISCUSSION

Our patient had a very slow-growing lymphoma in the anterior mediastinum.[2] It grew over 5 years and suddenly transformed into a high-grade lymphoma and developed multiple metastases in the right atrium, superior vena cava, pericardium, and spleen. Thus, for a benign-looking mass in the chest, histopathological examination is a must for diagnosis. Although there are more than 60 types of NHL, diffuse large B-cell lymphoma (DLBCL) is the most common type, making up about 30% of all lymphomas.[3] DLBCL is fatal if left untreated, but with timely and appropriate treatment, approximately two-thirds of all patients can be cured. The tumor grows so slowly that patients can live for many years mostly without symptoms, although some may experience pain from an enlarged lymph gland. After 5–10 years, low-grade disorders begin to progress rapidly to become aggressive or high-grade and produce more severe symptoms. Lymphoma can develop when mutations (genetic changes) cause lymphocytes to divide abnormally or to stay alive when they should not. More mutations can happen over time. They might cause abnormal lymphocytes to grow rapidly, like the cells in a high-grade lymphoma.

Possible treatments include:

1. R-CHOP chemotherapy, more intensive chemotherapy regimens, followed by an autologous stem cell transplant, if the lymphoma responds well to chemotherapy.
2. An allogenic stem cell transplant, if patient is fit enough and needs more than one course of chemotherapy to achieve remission.[4] Stem cell transplants are intensive forms of treatment and are not suitable for everyone.[5]

If the transformed lymphoma is restricted to a single site, it might be treated with radiotherapy (usually given after a course of chemotherapy).

CONCLUSION

Early diagnosis is a major prognostic factor for patients with DLBCL, adequate radiological imaging, as well as prompt biopsy and histopathological diagnosis, are vital for management.[2,3]

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