Case Series

Spectrum of uncommon enhancing anterior mediastinal masses

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Abstract

We present to you three abnormally enhancing mediastinal masses in the form of Castelman's disease, Ectopic parathyroid adenoma and a pseudoaneurysm. Castelman's disease is a rare, benign, lymphoproliferative disorder. Radiologically, it can present as either unicentric (local) or multicentric(systemic). Though parathyroid adenoma is a common cause for hyperparathroidism, 10% of the cases are found in ectopic parathyroid gland. Hence the prompt identification and diagnosis can be a challenge. Pseudoaneurysm is a condition in which there is a defect in vessel wall causing leakage from the artery. It is an emergency condition which when untreated may cause serious complications such as rupture and can have a high mortality rate.

Keyword: anterior mediastinal.

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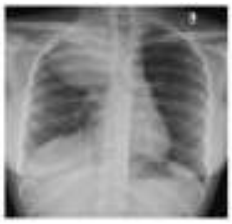
INTRODUCTION

An array of anterior mediastinal masses are diagnosed using computed tomography (CT) as the modality of choice. In such cases enhancement of the masses can be a important factor in narrowing down the diagnosis. . Contrast enhancement is an important factor in diagnosing lesions of vascular origin, such as parathyroid adenomas and Castleman's disease. In addition to contrast enhancement, the exact location and morphology of the mass, along with clinical features such as patient age, gender, signs, symptoms, and laboratory values, can usually lead to a short list of possible etiology. We present to you three abnormally enhancing mediastinal masses in the form of Castelman's disease, Ectopic parathyroid adenoma and a pseudoaneurysm. Castelman's disease is a rare, benign, lymphoproliferative disorder. Radiologically, it can present as either unicentric (local) or multicentric (systemic). Though parathyroid adenoma is a common cause for hyperparathroidism, 10% of the cases are found in ectopic parathyroid gland. Hence the prompt identification and diagnosis can be a challenge. Pseudoaneurysm is a condition in which there is a defect in vessel wall causing leakage from the artery. It is an emergency condition which when untreated may cause serious complications such as rupture and can have a high mortality rate.

CASE SERIES

Castelman's disease/ Angiofollicular lymphnode hyperplasia (AFH) is a rare, benign, lymphoproliferative disorder. It classified radiologically as (a) Unicentric (localized) or (b) Multicentric (multifocal). It can be further classified histologically as (i) Hyaline vascular which is the most common form of CD and occurs in unicentric cases, (ii) Plasma cell - A less enhancing variant which is associated with multicentric type, (iii) HHV-8 VIRUS associated castleman's disease is seen in multisystemic conditions in conjuncture with Kaposi's sarcoma and primary effusion lymphoma. Rarer histological forms include Mixed and Plasmablastic subtypes. Unicentric CD is not life threatening and surgical invention is usually sufficient. Recurrence rate is

very low in such cases. However multicentric variant has systemic involvement and can progress to lymphoma if untreated. It is often associated with Kaposis sarcoma and AIDS. Multicentric CD may require chemotherapy and radiotherapy along with surgical intervention. Its prognosis is relatively poor[1]. Our patient presented with history of cough and non-specific chest pain.



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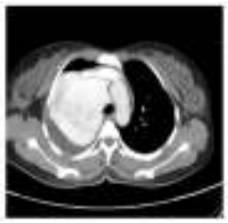

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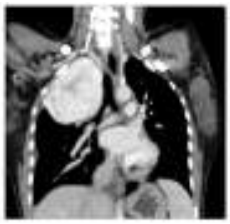

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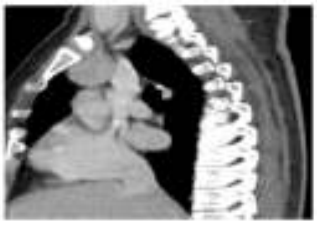

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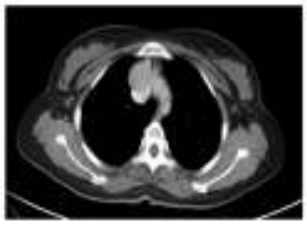

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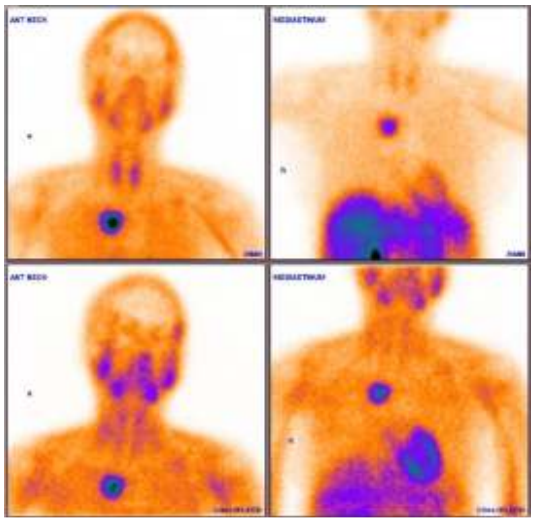

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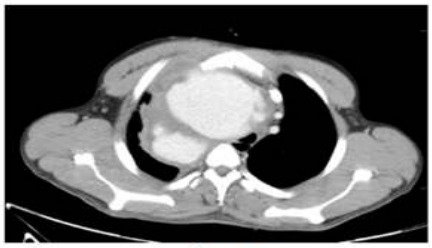

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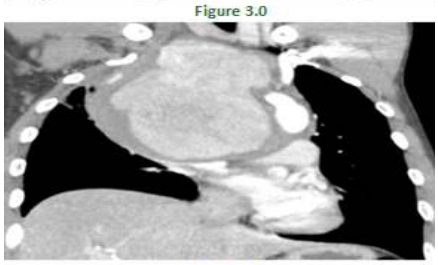

Figure 3.1

Legend

Figure 1.0: Chest PA view; **Fig 1.1:** Axial section; **Figure 1.2:** Arterial phase (Coronal Section) showing an intensely enhancing lesion
Figure 2.0: Sag view showing an enhancing lesion antero-superior to the aortic arch; **Figure 2.1:** Axial section;
Figure 2.2: Parathyroid scintigraphy showing abnormal uptake in the mediastinum
Figure 3.0: Axial section showing the enhancing mass surrounded by a thin rim of thrombus.
Figure 3.1: Coronal section of the pseudoaneurysm involving the innominate artery.

His chest X-ray showed large, opacity in the anterior mediastinum. She underwent CT-Thorax scan which showed a relatively well-defined, intensly enhancing soft tissue density lesion which was seen predominantly involving the anterior mediastinum. The mass lesion was seen to have a broad base contact with the mediastinum. There was no evidence of calcification noted. No evidence of cavity/fluid level was noted. In view of the above findings Caslemans disease was given as the primary diagnosis. This was confirmed by a biopsy done later which revealed it to be of the Hyaline Vascular category. Castleman's disease is mainly caused by the proliferation of the lymphoid tissues. Unicentric/Localized variant does not have usually have any underlying conditions and may be diagnosed coincidentally.Unicentric is more common in children and young adults while multicentric castlemans is seen to affect adults in their 5th and 6th decades. Multicentric Castleman's disease is of systemic etiology and is seen to cause excessive secretion of cytokines, more specifically Interleukin -6. It usually presents with systemic involvement such as organomegaly, fever, weight loss and autoimmune manifestations. HHV-8 associated Castleman's disease is seen in conjuncture with Kaposi's sarcoma and to a lesser degree with Primary effusion lymphoma which is also caused by HHV-8. Multicentric castleman's disease in rare cases can show symptoms overlapping with POEMS syndrome and may also progress to lymphoma[1]. The most common site for Castlemans disease is thorax (70%). Castleman's disease may not be visible on abdominal radiographs unless they are massive or calcified. Sonologically it may present as a hypoechoic mass with hyperechoic foci within and exhibits peripheral vascularity along with a feeding artery

penetrating the hilum and in plain CT it typically shows a characteristic "Arborizing" calcification. In MRI, the lesion shows hypointensity on T1 and hyperintensity on T2. The arborizing calcifications may be seen as low signal[2]. Histologically, the hyaline-vascular type, is characterized by germinal follicles with hyalinized vessels surrounded by concentric layers of small lymphocytes with proliferative interfollicular vascular stroma while the plasma cell type is characterized by a paucity of follicular hyalinized vessels in the germinal follicles. The mixed type, as its name suggests exhibits both hyaline-vascular and plasma cell features. As mentioned before unicentric castleman's disease may be asymptomatic and its treatment involves complete excision of the lesion with no reported case of recurrence. Multicentric variant however, requires a more complex treatment approach. A combination of chemotherapy, radiotherapy, antiviral therapy, targeted therapy and surgical resection is involved[2].

ECTOPIC PARATHYROID ADENOMA

Parathyroid adenomas are benign tumours of the parathyroid glands and are the most common cause of primary hyperparathyroidism. Up to 5% of parathyroid adenomas can occur in ectopic locations. In such cases radiological imaging plays an essential role in localizing the exact location of the gland. Common ectopic locations include mediastinum, retropharynx, carotid sheath and intra-thyroid. Our patient had complaints of recurrent renal calculi since childhood and was clinically diagnosed to have primary hypercalcemia. Patient initially had an ultrasonography scan before admission which did not show the ectopic parathyroid due to its retrosternal position. Post admission she underwent a 99mTc MIBI Dual Phase Parathyroid scintigraphy which was done at 20 mins and 2.5 hours delayed. It did not show any area of abnormal tracer uptake in the region of thyroid gland. However there was an area of abnormal tracer uptake noted in the thoracic region. She then underwent a computed tomography scan which showed a well defined soft tissue density seen anterior to the SVC. It showed an intense enhancement on contrast administration. **I**n view of the clinical features and uptake in the nm scan, ectopic parathyroid adenoma was given as the radiological diagnosis.

RESULT

Primary hyperparathyroidism is the third most common endocrine disorder in adults. About 85% of the primary hyperparathyroidism is due to adenoma[3]. The success of surgery in primary hyperthyroidism is based on clear diagnosis and preoperative localization methods. The currently used diagnostic techniques, which are non-invasive, include Tc - 99m MIBI scanning, SPECT, ultrasonography, CT imaging, and MRI[4,5,6].

PSEUDOANEURYSM

Pseudoaneurysms are a rare but serious condition which can present as enhancing anterior mediastinal mass. They can be rapidly progressive and hence often life-threatening. Usually resulting of a surgery or a traumatism. They most often occurs after trauma, cardiac procedures or infections. In contrast to patients with aneurysm, those with false aneurysm most commonly die of hemorrhage. Chest radiographs with an abnormal cardiac silhouette and rapidly expanding size are subtle signs, which are often overlooked. Noninvasive tests such as color-flow Doppler echocardiography, 2-dimensional echocardiography, cineangiographic computed tomography, and transesophageal echocardiography allow relatively easy recognition of these apparently rare lesions with increasing frequency. A 33 year old male came with complaints of acute onset chest pain and redness of the chest wall. He had a history of blunt trauma to the chest four months ago. Patient's chest X-ray showed suspicious splaying of the aortic arch and widening of the mediastinum. In view of this, he underwent computed tomography which showed a large partially thrombosed lobulated pseudoaneurysm arising from the innominate artery. It was found to be displacing the mediastinum and adjacent vessels laterally. A thin rim of thrombus was seen completely surrounding the pseudoaneurysm. The right subclavian artery and right common carotid artery were arising from the pseudoaneurysm with the origin of both arteries being narrowed. The patient was immediately taken up for surgery and surgical ligation was done. Post operatively there were no complications and the patient was then discharged.

DISCUSSION

Pseudoaneurysm is a breach in the vessel wall which is contained by the adventitia or surrounding perivascular soft tissue. There is a direct communication between the vessel lumen and the pseudoaneurysm, however unlike a true aneurysm it does not have all arterial wall layers. Therefore, the risk of rupture is higher than that of a true aneurysm of comparable size due to poor support of the aneurysm wall and hence pseudoaneurysms are considered a surgical emergency. Pseudoaneurysms are most commonly seen as a complication of arterial catheterization, biopsy or surgery, but can also present with due to fibromuscular dysplasia (dissection), mycotic aneurysm or myocardial infarction. However, pseudoaneurysm of the innominate artery is a rare complication of closed chest trauma. It has various clinical presentations—from superior vena cava

syndrome to a chance finding of mediastinal widening on chest X-ray[7].

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