

Castleman Disease Of Thorax

AAHLUWALIA, K SAGGAR, P SANDHU, V KALIA

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Introduction

Castleman disease is an unusual condition of unknown etiology and pathogenesis, originally described by Castleman in 1954 (1). Later, in 1972, Keller et al described two distinct histologic variants- hyaline vascular type and plasma cell type. We report two cases of thoracic Castleman disease; one with localized and the other with disseminated form of Castleman disease. On histology, both cases were of hyaline vascular type.

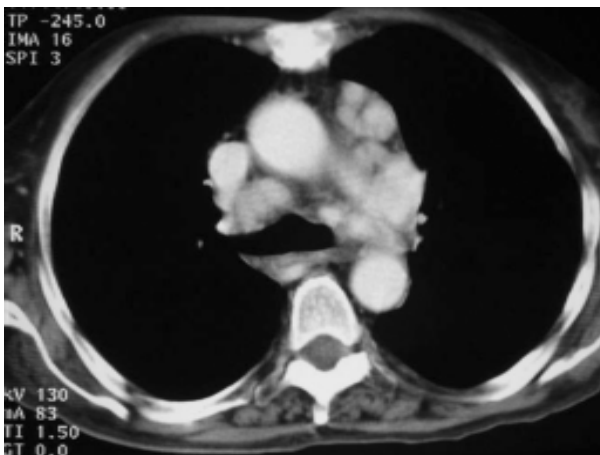


Figure 1. Spiral CECT of the chest shows multiple discrete homogeneously enhancing lymph nodes in the precardinal region and AP window.

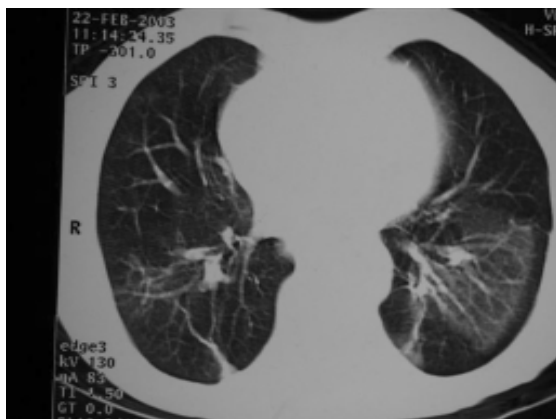


Figure 2. Axial CT section in the same patient shows a patch of ground glass haze in left lower lobe.

Case 1

A sixty year old woman presented with breathlessness of one month duration associated with fever. She also had anorexia, weight loss and generalized weakness. She was a known case of type 2 diabetes mellitus. General physical examination was unremarkable except for cervical lymphadenopathy. Chest radiograph revealed mediastinal widening. Spiral CT of the chest revealed multiple discrete lymphnodes in the mediastinum, which showed marked homogeneous enhancement following intravenous contrast administration (Figure 1). A patch of ground glass haze was seen in left lower lobe (Figure 2). Subsequently, cervical lymph node biopsy revealed Castleman disease of hyaline vascular type.

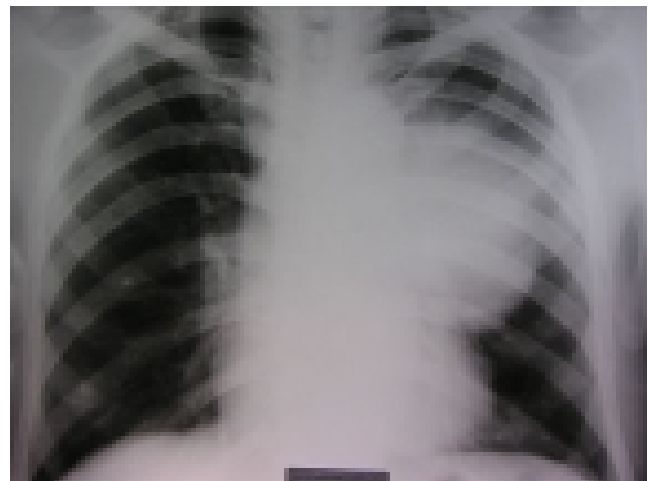


Figure 3. Chest radiograph (PA view) showing a well marginated mediastinal mass projecting on the left side

Case 2

A fifty-year-old man presented with complaint of urinary retention. Routine chest radiograph showed widening of mediastinum with a sharply marginated mass projecting on the left side (Figure 3). Subsequently, CT scan revealed a large lobulated mass in anterior mediastinum abutting the chest wall. On CECT, it showed intense nodular

From the Department of Radiodiagnosis, Dayanand Medical College & Hospital, Ludhiana-141001 (Punjab)

Request for Reprints: Dr Archana Ahluwalia, 125-D, Kitchlu Nagar, Ludhiana-141001 Punjab

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enhancement (Figure 4). No discrete mediastinal lymphadenopathy was seen. CT guided biopsy of the lesion confirmed Castleman disease of hyaline vascular type.

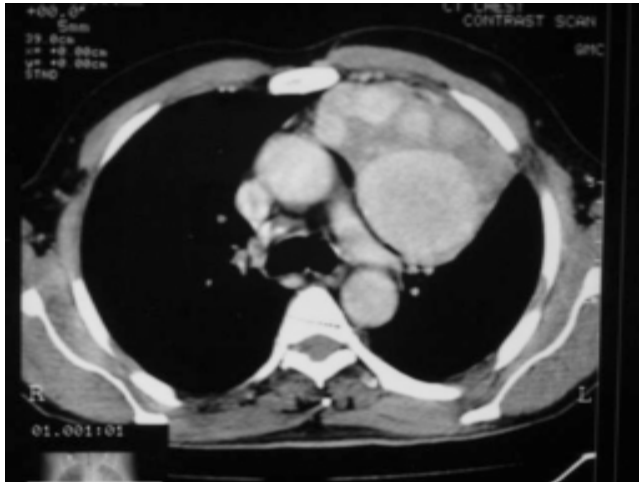


Figure 4. Spiral CECT of chest scan reveals a large lobulated mass in anterior mediastinum abutting the chest wall with intense nodular enhancement

Discussion

Castleman disease, also known as angiofollicular mediastinal lymph node hyperplasia or giant lymph node hyperplasia, is an uncommon benign lymphoproliferative disorder characterized by hyperplasia of lymphoid follicles.(1,2). Histologically, it is of two types- hyaline vascular and plasma cell. Hyperplasia of lymphoid follicles with germinal centre formation and presence of numerous capillaries with hyalinized walls characterize the hyaline vascular type. This type accounts for 90% of the cases and usually manifests as solitary perihilar or mediastinal mass in asymptomatic patients (2).

The plasma cell type is characterized by relatively few capillaries and the presence of mature plasma cells between the hyperplastic germinal centres. This form is usually associated with clinical manifestations including fever, fatigue, anemia, polyclonal hypergammaglobulinemia and bone marrow plasmacytosis (2,3).

Clinically, Castleman disease of thorax is classified into two major subgroups- localized disease and disseminated disease.

The patients are classified as having localized Castleman disease when only one mediastinal compartment is involved on CT or MR imaging and there is no clinical or radiological evidence of additional disease in an extra thoracic site. The involvement of more than one mediastinal compartment or evidence of disease in an extra thoracic location is classified as disseminated Castleman disease.

On imaging, three morphologic patterns are observed in localized Castleman disease: a solitary well circumscribed mediastinal mass, a dominant infiltrative mass with associated lymphadenopathy or lymphadenopathy without

a dominant mass. Disseminated Castleman disease manifests with diffuse mediastinal lymphadenopathy that involves multiple mediastinal compartments. Focal calcification can be seen in 5-10% cases. The enlarged lymph nodes typically show marked homogeneous enhancement following intravenous contrast administration (4,5). However, the enhancement is less intense in plasma cell type than in hyaline vascular type of disease. In addition to lymphadenopathy in disseminated thoracic Castleman disease, there may be associated lung parenchymal findings in the form of lymphocytic interstitial pneumonia which presents with centrilobular nodules, interlobular septal and bronchovascular interstitial thickening, ground glass attenuation and air space consolidation (2).

On T1W MR images, the lesions are hyperintense compared with skeletal muscles. These become markedly hyperintense on T2W images and show strong enhancement following I/V administration of Gadolinium. The presence of flow voids suggestive of feeding vessels around the mass provide an important clue to the hypervascular nature of these lesions (6,7).

Angiography is reported to demonstrate an intense tumor blush and enlarged feeding vessels arising from bronchial, internal mammary or intercostal arteries (6).

Pre-operative diagnosis of Castleman disease is difficult, as fine needle aspiration cytology is usually non-diagnostic. Definitive diagnosis requires a large tissue sample using mediastinoscopy or biopsy. Complete surgical resection is the mainstay of therapy in localized Castleman disease. The patients with disseminated Castleman disease of either histologic subtype have a poor prognosis. These patients are usually managed with a combination of radiation therapy, corticosteroids and chemotherapy (4).

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