CASE REPORT

Posterior mediastinal Castelman's disease presented as hypervascular lipomatous tumor

Michaelides M[1], Nikolaidou A[2], Konstantinou D[1], Balis GC[2], Tsitouridis I[1]

[1] Department of Diagnostic and Interventional Radiology, Papageorgiou General Hospital, Thessaloniki, Greece.
[2] Department of Pathology, Theagenio Cancer Hospital, Thessaloniki, Greece.

Abstract
Castelman's disease (CD) is a rare lymphoproliferative disorder most often found in the chest. Herein we describe the imaging findings of a histologically proven case of CD involving the posterior mediastinum in a 41-year-old asymptomatic man presented as a well-defined hypervascular lipomatous tumor. To our knowledge less than 10 cases of posterior mediastinal CD have been reported so far and this is the first case of CD mimicking mediastinal lipomatous tumor. Hippokratia 2011; 15 (4): 361-362

Key words: Castelman's disease, posterior mediastinum, lipomatous tumor, CT, MRI

Corresponding author: M Michaelides, Department of Radiology, Papageorgiou General Hospital, Thessaloniki Ring Road, Nea Eukarpia, 56403, Thessaloniki, Greece. Tel: 00306972701104, e-mail: mihalismihailidis@gmail.com

Castelman's disease (CD) is a rare benign lymphoproliferative disorder usually located in the thorax, most frequently occurring as an asymptomatic mediastinal or hilar mass[1]. Here we present a case of CD located in the posterior mediastinum, presented as a well-defined hypervascular lipomatous tumor in a 41-year-old asymptomatic man. Less than 10 cases of posterior mediastinal CD have been reported[2-5].

Case report

A 41-year-old asymptomatic man with no significant medical history was referred to our department for evaluation of a well-defined mass of the posterior mediastinum (Figure 1). CT of the thorax demonstrated a well-defined tumor located in the left costovertebral sulcus with fatty and strongly enhancing components. There was no pleural effusion or other abnormal findings from the mediastinum and lung parenchyma (Figure 2). On MRI, the central nodule of the lesion demonstrated intermediate to high signal intensity on HASTE T2WI with intense contrast enhancement on post-contrast FLASH T1WI, while the peripheral lipomatous component demonstrated high signal intensity on HASTE T2WI and low signal intensity on post-contrast FLASH T1WI due to fat suppression (Figure 3).

Two weeks later, the patient was subjected to left lateral thoracotomy. At surgery, a lipomatous hypervascular mass was found in the posterior mediastinum occupying the intercostal space. The tumor was covered by the parietal pleura and was supplied by branches of the intercostal arteries. It was firmly attached to the chest wall and was carefully completely removed. The resected mass was a well-defined, ovoid tumor, measuring 7.5X4.5X2.5cm. On cut, the mass had an outer layer of yellow-tan color and elastic consistency, while the core of the mass, was grey-tan colored and elastic, resembling an enlarged lymph node. Multiple paraffin sections confirmed the presence of a lymph node in the center of the mass that generally preserved its architectural structure. Other findings included follicles with involuted germinal

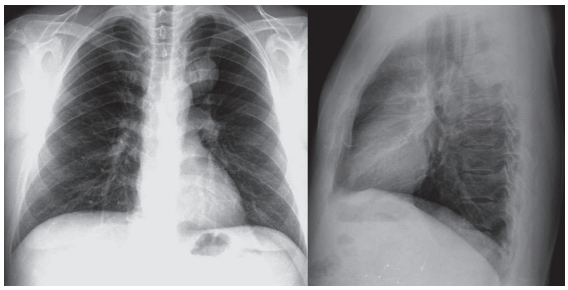

Figure 1: Chest X-ray demonstrating a well-defined left sided mass of the posterior mediastinum.

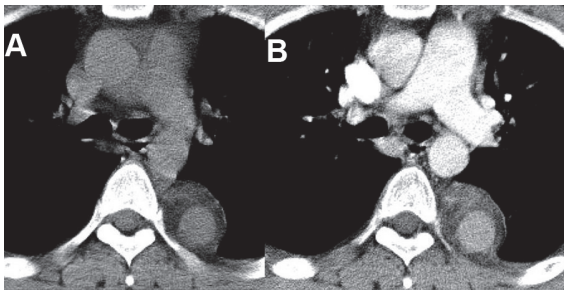

Figure 2: CT before (A) and after (B) administration of contrast medium demonstrating a well-defined left sided fatty mass of the posterior mediastinum measuring 51X32 mm, occupying the intercostal space. Within the mass, a round component measuring 24mm demonstrates intense contrast enhancement (from 43 HU to 104 HU). The lipomatous elements of the lesion also enhance (from -27 HU to 36 HU).

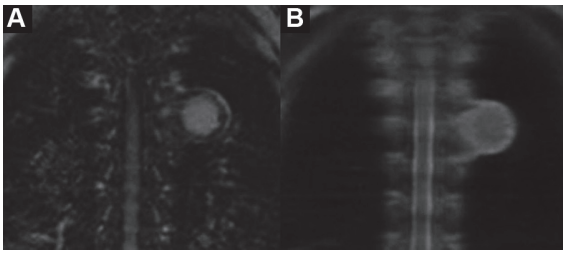

Figure 3: MRI of the lesion. A. Post-contrast FLASH T1WI in coronal plane. B. HASTE T2WI in coronal plane. The central nodule of the lesion demonstrates intense contrast enhancement and is surrounded by fat-suppressed lipomatous tissue.

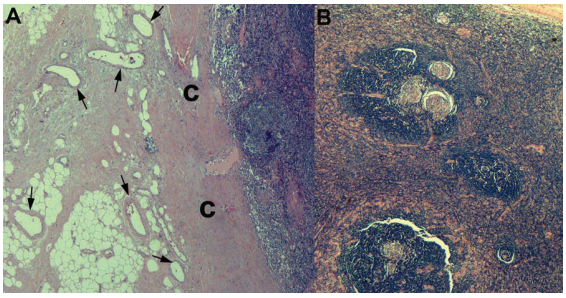

Figure 4: Photomicrographs of the lesion (Hematoxylin-Eosin). A. The adipose tissue surrounding the lymph node contains multiple hyperaemic vessels of variable size (arrows). A thick capsule (C) separates the adipose tissue from the lymph node. B. Section of the lymph node, showing follicles with involuted germinal centers and the "onion skin"-like arrangement of mantle zone lymphocytes.

centers, hyperplastic vessels with hyalinized stroma and prominent, concentric, "onion skin"-like arrangement of lymphocytes. The peri-follicular space presented numerous mature lymphocytes, hyperplastic venules and diffuse plasma cells. These findings were characteristic of hyaline vascular type of CD. Sections taken from the tumour's yellow-tan area revealed adipose tissue with hyperaemic vessels of variable size and perivascular, chronic inflammatory cell infiltrates (Fig. 4).

The patient was discharged on the post-operative day 10. One year of follow-up revealed excellent clinical condition and no signs of recurrence.

Discussion

CD was first described by Castleman in 1954[6]. Approximately 70% of CD is located in the chest along the tracheobronchial tree in the mediastinum or lung hilus, although can be also found in extrathoracic lymph nodes (neck, axilla, pelvis, retroperitoneum) and in muscles. The etiology is unknown. Patients are usually asymptomatic or have nonspecific symptoms such as coughing, dyspnea, chest pain and back pain[1,4].

Histologically CD is divided into 3 types· the hyaline vascular, the plasma cell and the mixed type[1,5]. The hyaline vascular type is the commonest (91%) and is characterized by hyaline vascular follicles with interfollicular lymphocytes and capillary proliferations. Plasma-cell type (9%) is characterized by large follicles with intervening sheets of plasma cells. CD is also divided into unicentric and multicentric form[3]. Unicentric form, more often of hyaline vascular type, is usually an asymptomatic and benign disease confined within the chest and treatable by surgical resection. On the other hand multicentric form, usually of plasma-cell type, is an aggressive extrathoracic disease associated with more complicated systemic manifestations not amenable to surgical treatment[3-5].

Preoperative diagnosis is difficult because of the absence of specific symptoms and imaging findings. Needle biopsy has disappointing results and increased risk due to the hypervascularity. Thus, final diagnosis is usually established only after surgical removal of the lesion[4,5].

CD of the posterior mediastinum with chest wall involvement is very rare and so far less than 10 cases have been reported. The majority of these cases were symptomatic, of hyaline vascular type and with no reported recurrence or malignancy after surgical excision[1-5]. Other atypical locations of thoracic CD include pericardium, lung, pleura, axilla and supraclavicular fossa[3].

In our case differential diagnosis included mediastinal lipomatous tumors with enhancing components such as liposarcoma, atypical lipoma, atypical neurogenic tumor, mediastinal myelolipoma, mediastinal angiomyolipoma, mediastinal angiolipoma and mature teratoma. To our knowledge, this is the first case of CD simulating posterior mediastinal hypervascular lipomatous tumor. We cannot explain the presence of hypervascular adipose tissue surrounding a lymph node with CD. We believe that this was an abnormal response to chronic inflammation resulting in overgrowth of the normal mediastinal adipose tissue. The presence of chronic inflammatory infiltrates within the fat is in favor of that.

References

1. Ueda H, Kouso H, Tsukamoto S. Castleman's disease of the chest wall. J Thorac Oncol. 2009; 4: 426-428.
2. Kim KH, Kim SD, Kim SH, Park JY. Giant Lymph Node Hyperplasia (Castleman's Disease) as a Rare Cause of Back Pain. J Korean Neurosurg Soc. 2008; 43: 169-171.
3. Ko SF, Wan YL, Ng SH, Lin JW, Hsieh MJ, Fang FM, et al. Imaging features of atypical thoracic Castleman disease. Clin Imaging. 2004; 28: 280-285.4. Erdogan A, Eser I, Ozbilim G. Posterior mediastinal localization of Castleman's disease: report of a case. Surg Today. 2004; 34: 772-723.
5. Rena O, Casadio C, Maggi G. Castleman's disease: unusual intrathoracic localization. Eur J Cardiothorac Surg. 2001; 19: 519-521.
6. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymph node hyperplasia resembling thymoma. Cancer. 1956; 9: 822-830.