

Interactive CardioVascular and Thoracic Surgery

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Interact CardioVasc Thorac Surg 2011;13:346-347; originally published online Jun 1, 2011;

DOI: 10.1510/icvts.2011.272229

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Case report - Thoracic oncologic Mediastinal Castleman's disease mimicking thoracic paravertebral schwannoma

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Received 19 March 2011; received in revised form 6 May 2011; accepted 16 May 2011

Abstract

A 51-year-old female underwent resection of a solid lesion in the posterior mediastinum, preoperatively interpreted at imaging as thoracic schwannoma, requiring double sequential surgical procedure to be resected. The histologic examination of the resected mass diagnosed a hyaline-vascular Castleman's disease.

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Keywords: Computed tomography; Giant lymph node hyperplasia; Mediastinal neoplasms; Neurilemmoma; Thoracic surgery

1. Introduction

Castleman's disease (CD) is a rare lymph proliferative disorder of unknown aetiology that often involves the thorax [1]. Since Castleman's first description, three sub-types have been described with characteristic histological features, localisation, clinical presentation and prognosis: the hyaline-vascular subtype is generally localised, benign, asymptomatic, resectable with good prognosis and low recurrence rate; the plasma cell type and mixed type are more aggressive diseases with multifocal involvement of nodal stations and organs. This disorder is often misdiagnosed or undiagnosed without histological confirmation, even though an incorrect surgical approach can lead to intraoperative complications, such as massive bleeding [2]. We report a case of hyaline-vascular symptomatic CD of the posterior mediastinum, with the radiological appearance of a thoracic paravertebral schwannoma.

2. Clinical summary

A 51-year-old female presented for a physical examination for persistent cough. A chest radiograph revealed a left paravertebral nodular opacity. Computed tomography (CT) of the thorax performed with intravenous administration of iodine contrast revealed a multilobulated solid lesion in the posterior mediastinum, partly involving the lateral foramen between T-6 and T-7 (Fig. 1). The images were suggestive of thoracic schwannoma and the patient underwent gross-total resection of the tumour through a combined posterior

and left trans-thoracic approach. The postoperative course was free of major complications, but the patient experienced important thoracotomy-related pain, not responsive to NSAID. At histological examination, the nodal tissue showed follicular hyperplasia, focally associated with atrophic germinal centres penetrated by hyalinised capillaries, resulting in the so-called 'lollipop follicles'. Some foci of lamination of the mantle cell layers could also be seen. According to these microscopic findings and the clinical picture, a diagnosis of hyaline-vascular CD was then rendered (Fig. 2). At six-month follow-up patient is alive, painless and without evidence of recurrence of disease.

3. Discussion

CD is a rare lymphoproliferative disorder, first described in 1956 by Castleman et al. [3]. It is usually benign, idiopathic and the median age of presentation ranges from adolescence into the seventh decade [4]. It is rarely associated to autoimmune phenomena or within POEMS syndrome or in conjunction with a non-Hodgkin lymphoma especially in HIV positive patients [1]. Clinically CD presents both as benign localised resectable disease or as an aggressive multicentric variant associated with systemic symptoms and frequent recurrence; in the latter, symptoms tend to mimic a lymphoma (fatigue, fever, night sweats, weight loss).

Histologically three forms have been described: the hyaline-vascular type is the more frequent (85%) and often asymptomatic, the other sub-types are less frequent (15%) and more aggressive and symptomatic. About 70% of cases occurs in the chest [5], usually (90%) involving the mediastinum. Radiological evaluation generally shows a mediastinal or hilar mass, homogeneously enhanced after contrast,

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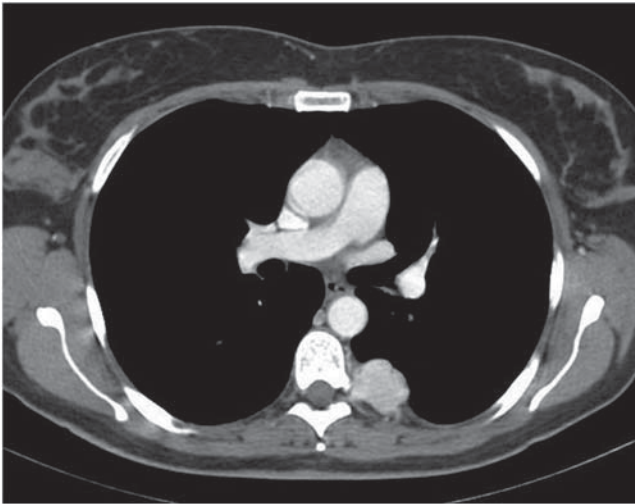


Fig. 1. CT-scan of the chest showed a multilobulated solid lesion of 33×20×30 mm in the posterior mediastinum, partly involving the lateral foramen. The lesion appeared without calcification and with homogeneous contrast enhancement. CT, computed tomography.

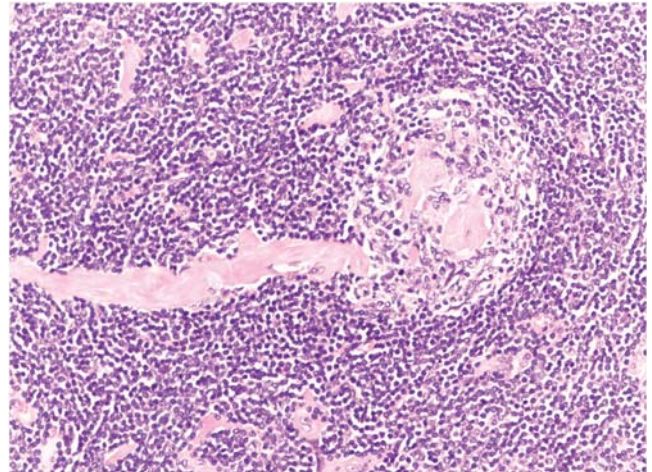


Fig. 2. The tissue showed follicular hyperplasia, focally associated with atrophic germinal centers penetrated by hyalinised capillaries, resulting in the so-called 'lollipop follicles'. Some foci of lamination of the mantle cell layers could also be seen (so-called 'onion skin' aspect) (H&E, ×100).

resembling thymoma or lung cancer [6]. At CT-scan three different patterns of the hyaline-vascular type have been described: a solitary, non-invasive mass, an infiltrative mass with associated lymphadenopathy, or lymphadenopathy without mass [7]. The treatment of choice is surgical for the solitary lesions, recurrence can occur with marginal subtotal resection.

Although CD can rarely arise primarily in the posterior mediastinum [2, 6], it has never been reported to mimic schwannomas [8]. In our report, the mass, located in the upper costo-vertebral space, was interpreted as a thoracic benign nerve sheath tumour; percutaneous needle biopsy was not recommended and the surgical approach was primarily neurosurgical. We did not experience intraoperative massive bleeding, as described in literature [2], but the presence of strong adhesions contraindicated a video-assisted thoracic surgery (VATS) approach subsequent to the posterior dissection of the spinal roots. We performed complete resection of the mass through left lateral thoracotomy, thus causing major postoperative pain.

Because a variety of unusual lesions have radiological features similar to thoracic benign nerve sheath tumour [9], in case of a spinal dumbbell-shaped or paravertebral tumour a diagnosis other than schwannoma or neurofibroma should

be considered, including CD. In both cases surgical approach is curative, so every effort must be made for the procedure to be radical.

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