Development of Castleman Disease in the Paravertebral Space Mimicking a Neurogenic Tumor

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Castleman disease is a relatively rare disease, characterized by well-circumscribed benign lymph-node hyperplasia. The disease may develop anywhere in the lymphatic system, but is most commonly reported as unicentric Castleman disease in the mediastinum along the tracheobronchial tree. It is usually asymptomatic and detected on plain chest radiography as an incidental finding. We report an incidentally detected case of Castleman disease in the paravertebral space that was preoperatively diagnosed as a neurogenic tumor and treated by complete surgical resection.

Key words: 1. Castleman disease
2. Mediastinal neoplasm
3. Neurogenic tumor

Case report

A 19-year-old male patient was incidentally found to have an abnormal shadow in the left thorax on chest radiography. He did not complain of any symptoms, such as coughing, dyspnea, chest discomfort, or neurologic symptoms. His previous medical and family history were unremarkable. No unusual findings were observed on a physical examination. His hematologic and serologic profiles, including inflammatory markers, were within the normal ranges. Plain chest radiography showed a well-defined mass in the paravertebral region (Fig. 1). Chest computed tomography (CT) revealed a 3.4-cm mass with a lobulated contour between the left ninth and tenth intercostal spaces. However, the CT images did not show foramen dilatation or central calcification. Furthermore, there was no radiologic evidence of invasion of the

Fig. 1. Preoperative plain chest radiography. A lobulated contour is seen surrounding a mass measuring roughly 3.4 cm (along the arrows).
adjacent vasculature, soft tissue, or bony tissue; these findings were highly suggestive of a neurogenic tumor (Fig. 2).

We planned to excise the mass via video-assisted thoracoscopic surgery (VATS) in the left thorax. The lesion was a hypervascular mass located grossly in the paravertebral space and showed cystic changes or partial necrosis (Fig. 3A). It was a tumor measuring 4.0×3.0×2.0 cm located laterally to the sympathetic trunk, but at some distance from the sympathetic nerve (Fig. 3B). It abutted the intercostal nerve and surrounding tissue and seemed to originate from the intercostal nerve (Fig. 3C). There were no definitive feeding vessels surrounding the mass. Thus, based on its macroscopic appearance, we assumed the mass to be a neurogenic tumor originating from the intercostal nerve. The tumor was completely removed, including excision of the affected intercostal nerve and periosteum adjacent to the rib because of severe adhesions to the mass. Frozen-section biopsy was performed intraoperatively, and the tumor was determined to be a lymphoproliferative lesion. Subsequently, the permanent histologic examination revealed atypical lymphoid hyperplasia, consistent with what is observed in hyaline-vascular–type Castleman disease (CD). Histologically, the tumor was composed of lymphoid follicles with hyalinized vessels surrounded by circumferentially arranged layers of lymphocytes in an onion-skin pattern (Fig. 4). The patient’s postoperative hospital course was uneventful, and he was discharged without complications on the second postoperative day.

Informed consent of the patient was waived by Institutional Review Board.

**Discussion**

CD is a rare and usually benign lymphoproliferative disorder of uncertain etiology, first described by Castleman et al. [1] in 1954. It is also known as
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Fig. 4. Histologic features demonstrating lymphoid follicles with hyalinized vessels surrounded by numerous lymphocytes in an onion-skin pattern (H&E staining, ×200).

'angiomatous lymphoid hamartoma,' 'angiofollicular lymph node hyperplasia,' 'benign giant lymphoma,' and 'giant lymph node hyperplasia' [2,3].

The etiology of CD is still unclear. The disease was later classified clinically as either unicentric or multicentric, and histologically into 3 subtypes: hyaline-vascular, plasma cell–predominant, or mixed [4]. Unicentric CD manifests as local signs and symptoms related to lymphadenopathy, and hence responds well to surgical excision. In contrast, patients with multicentric CD frequently have systemic symptoms. The multicentric type requires systemic chemotherapy [4]. However, patients rarely have nonspecific symptoms, such as dyspnea, cough, recurrent infection, and lymphadenopathy [5]. The hyaline-vascular type is most frequent, accounting for approximately 90% of unicentric CD, whereas the plasma cell–predominant type is seen in both unicentric and multicentric forms of CD. It comprises approximately 10% of unicentric cases, and is aggressive and symptomatic [4,5]. The hyaline-vascular type usually presents and develops as a solitary non-invasive mass [5,6]. CD generally affects the chest, along the tracheobronchial tree in the mediastinum or lung hilus, but it may occur wherever the lymphatic system is present, such as in the pelvis, neck, retroperitoneum, and muscle [3-5]. However, CD in the paravertebral space is extremely rare. There is no sex- or race-based predominance, and most patients are diagnosed with the disease before the age of 30 years [6]. Most patients are asymptomatic, and the disease is detected incidentally on plain chest radiography, as in the present case. The preoperative diagnosis is difficult because of the lack of symptoms or the presence of nonspecific symptoms and radiologic features. CD may mimic other more common mediastinal lesions, such as thymoma, lymphoma, and neurological tumors [7]. It has no characteristic imaging findings, but manifests as a hypervascular lesion that may show intense enhancement on a contrast CT scan, unlike neurogenic tumors. Calcification and central necrosis are less common findings [7]. The radiologic findings in this case were different from those observed in other cases, as hypervascularity and calcification were not detected in the present case. Consequently, there was no evidence of CD. It is, therefore, difficult to diagnose CD through radiology alone, and the final diagnosis requires a histologic examination after surgical removal of the lesion. Surgical resection is the mainstay of treatment for unicentric CD, and has been performed via thoracotomy. While VATS resection has become an effective and reliable option for excising mediastinal masses, only a few reports have described VATS resection in cases of CD, because CD tumors are hypervascular and often have dense adhesions to the surrounding tissue [8]. Thus, in patients with CD, the tumor must be approached with great care, especially in the mediastinum, given its close proximity to vital structures. However, preoperative embolization can be attempted prior to surgical resection to minimize intraoperative bleeding from hypervascular mediastinal CD tumors [8].

In this case, the tumor developed in the paravertebral space, which is an unusual location. The preoperative diagnosis was neurogenic tumor. Furthermore, the tumor abutted the intercostal nerve, and was considered to be a neurogenic tumor originating from the intercostal nerve. We performed complete resection via VATS, and the permanent pathologic findings subsequently confirmed CD. In conclusion, we present a rare case of CD in the paravertebral space.

Conflict of interest

No potential conflict of interest relevant to this article was reported.
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References

1. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymphnode hyperplasia resembling thymoma. Cancer 1956;9:822-30.
2. Erdogan A, Eser I, Ozbilim G. Posterior mediastinal localization of Castleman’s disease: report of a case. Surg Today 2004;34:772-3.
3. Yoon HS, Jang GG, Kang JS, Kim H. The Castlemen’s disease in mediastinum: a case report. Korean J Thorac Cardiovasc Surg 2000;33:265-7.
4. Madan R, Chen JH, Trotman-Dickenson B, Jacobson F, Hunsaker A. The spectrum of Castleman’s disease: mimics, radiologic pathologic correlation and role of imaging in patient management. Eur J Radiol 2012;81:123-31.
5. Rena O, Casadio C, Maggi G. Castleman’s disease: unusual intrathoracic localization. Eur J Cardiothorac Surg 2001;19:519-21.
6. Paci M, Valli R, Tenconi S, Sgarbi G. Mediastinal Castleman’s disease mimicking thoracic paravertebral schwannoma. Interact Cardiovasc Thorac Surg 2011;13:346-7.
7. Alavi A, Asadi Gharabaghi M. Unicentric Castleman’s disease: an uncommon cause of posterior mediastinal mass. BMJ Case Rep 2013;2013:bcr2012008522.
8. Shetty S, Brenes RA, Panait L, Sanchez JA. Video assisted thoracoscopic resection of a posterior mediastinal Castleman’s tumor. J Cardiothorac Surg 2011;6:113.