

Case Report

Unicentric Castleman disease located in the anterior mediastinum misdiagnosed as invasive thymoma: a case report

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ABSTRACT. Castleman disease is a rare lymphoproliferative disorder of unknown etiology. The localized form, which usually presents as a slow-growing mass, is most commonly located in the mediastinum. Invasion of the vena anonyma by a mass has rarely been reported. We herein describe a case of initially misdiagnosed invasive thymoma in a 72-year-old woman, but postoperatively proven to have anterior mediastinal Castleman disease with invasion of the vena anonyma.

Key words: Castleman disease; Invasive thymoma; Vena anonyma; Anterior mediastinum

INTRODUCTION

Castleman disease (CD) is a rare form of localized lymph node hyperplasia of uncertain etiology. There are two types of unicentric CD: the more common hyaline vascular type and the plasma cell type. The mediastinum is the most common site of involvement. It is often difficult to distinguish CD from thymic tumors. Especially when a tumor has invaded the vena anonyma, it is easy to misdiagnose CD as invasive thymoma. Surgical resection is the main treatment strategy for unicentric mediastinal CD (Herrada et al., 1998), and is typically performed via standard thoracotomy (Hountis et al., 2008).

CASE PRESENTATION

A 72-year-old Chinese woman presented to the Department of Respiratory Medicine at our hospital with a 2-month history of coughing and chest congestion. She denied any history of fever, weight loss, or fatigue, and her family history revealed no malignancies. There was no evidence of infection with human immunodeficiency virus. Her physical examination findings were essentially normal. Routine blood laboratory results were simply suggestive of mild anemia. A spiral computed tomography (CT) scan of the chest indicated the presence of a mass in the anterior mediastinum (Figure 1A and B).

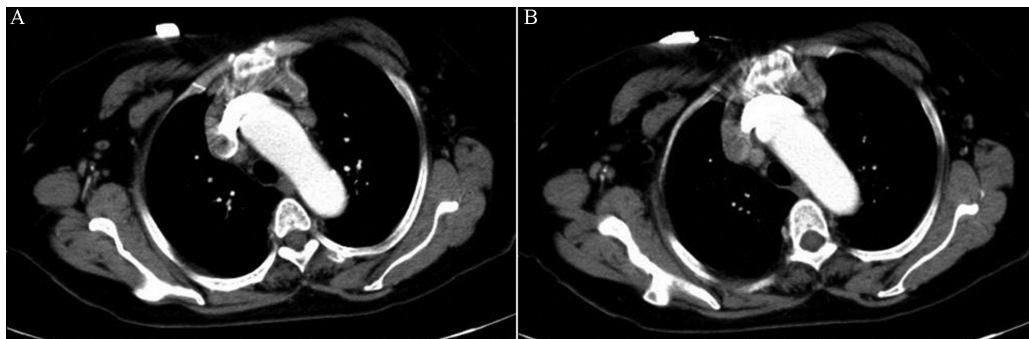


Figure 1. Preoperative computed tomography scans showing a mass in the anterior mediastinum, with close proximity to the left vena anonyma. **A.** Tumor close to the vein wall and subpanels; **B.** Most of the vein wall surrounded by tumor.

Surgical resection of the mass was performed through a sternal thoracotomy under double-lung anesthesia. The tumor was approximately 2.0 x 4.5 x 1.5 cm in size and closely affixed to both sides of the mediastinal pleura and left vena anonyma. The tumor was carefully separated from the surrounding tissues, and then completely dissected along with the bilateral mediastinal pleura. After the vena anonyma located adjacent to the mass was securely ligated, the whole mass with a portion of the venous wall was resected, and 5-0 prolene sutures (Somerville, NJ, USA) were used to suture the venous side in a continuous pattern. The intraoperative bleeding volume was about 80 mL. The patient's postoperative course was uneventful. Pathologic examination of the surgical specimen indicated a case of plasma cell CD, which was unexpected (Figure 2). The patient recovered well and was discharged 8 days after surgery. She remained asymptomatic throughout the subsequent follow-up visits. Her initial coughing and congestion did not recur. A 6-month follow-up CT examination revealed no evidence of relapse.

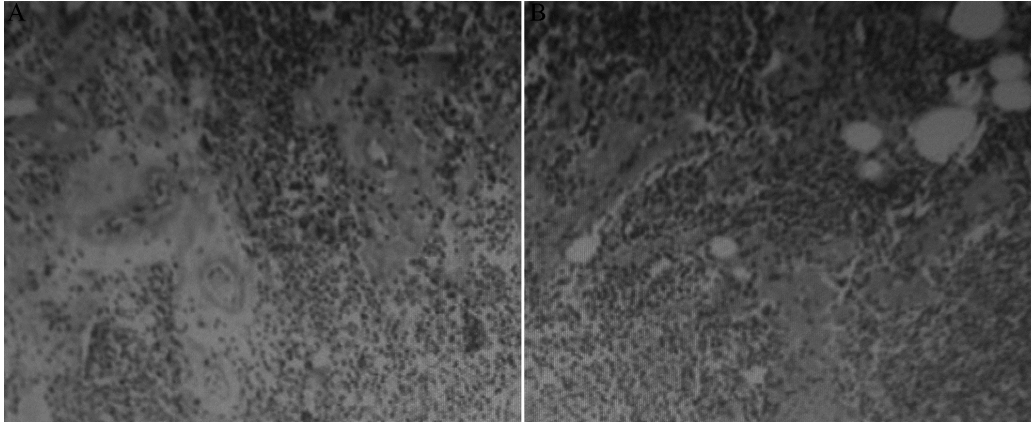


Figure 2. Histopathological findings of the resected tumor. **A. B.** Same meaning that histopathological findings of the resected tumor.

DISCUSSION

In 1956, Castleman et al. described a group of patients with benign mediastinal lymphoid masses that they determined to originate from mediastinal lymph node hyperplasia. The patients' condition was termed CD. Previously, CD was often classified into unicentric and multicentric types, but recent reports have been more inclined to define CD by its histopathogenic type as follows: hyaline-vascular CD, plasma cell CD, human herpes virus-8 (HHV-8)-associated CD (also known as plasmablastic CD), and multicentric CD. Hyaline-vascular CD accounts for approximately 70% of all cases and affects men and women equally (Cronin and Warnke, 2009).

Although the exact cause of CD is unknown, the plasma cell type is likely associated with infection and inflammation. Available data suggest that interleukin-6 (IL-6) receptor polymorphism may be a contributing factor in CD (Stone et al., 2013). Moreover, immune dysregulation has also been implied as the originating factor of CD. A recent report indicates that approximately 25% of cases are clinically confirmed to be associated with HHV-8 infection (Dossier et al., 2013). Furthermore, excessive production of IL-6 in the germinal centers of multicentric CD has been observed to be associated with HHV-8 infection and to possibly play an important role in the pathophysiology of the disease (Lee et al., 2008; van Rhee et al., 2010). In addition, some CD cases are thought to be at risk for B-cell malignancy, whereas some of them, a few multicentric CD masses might transform into malignant lymphoma. However, in fact, most CD patients do not develop malignant tumors during follow-up.

Multicentric CD commonly presents as multifocal lymphadenopathy and hepatosplenomegaly (Dham and Peterson, 2007). Patients usually develop fever, night sweats, fatigue, weight loss, and other symptoms in plasma cell unicentric and multicentric CD, whereas anemia, elevated erythrocyte sedimentation rate, thrombocytopenia, abnormal liver function, hypoalbuminemia, and other laboratory parameter changes are also possible (Dham and Peterson, 2007).

Unicentric CD is often localized in the mediastinum. It typically appears on CT scans as a well-defined mass of soft tissue density with little calcification. Therefore, it is important to differentiate mediastinal CD from other mediastinal tumors, especially thymomas. When a mediastinal CD mass is observable as closely adherent to the peripheral vascular tissue on CT,

it is easily misdiagnosed as an invasive thymoma. However, systemic symptoms are usually associated with CD, which may help in the diagnosis. Additionally, other imaging modalities, such as positron emission tomography, might also be useful for disease identification and staging (Madan et al., 2012).

Unicentric CD often requires surgical resection. While video-assisted thoracoscopic surgical resection has become an effective and reliable option for excising mediastinal masses (Nq and Yim, 2010), the tumor in our case was in close proximity to the left vena anonyma with no apparent boundaries between the mass and the vein; therefore, thoracoscopic surgical resection could not be performed. Thus, we believed that the surgery could be performed more safely through sternal thoracotomy. To prevent venous bleeding, part of the vena anonyma wall was removed after complete tumor resection. Several reports also indicate that prior to surgical resection, preoperative embolization of hypervascular mediastinal CD might help minimize intraoperative bleeding (Robert et al., 2008). The majority of CD patients achieve long-term survival and low recurrence rates. Our patient was followed-up for 6 months without recurrence, and her pathological findings indicated a case of plasma cell-type focal CD. If patients present with systemic symptoms, these symptoms often disappear quickly after lymphadenectomy. In our case, the patient's cough and fever quickly dissolved, but her anemia recovered more gradually over a 4-week period.

CONCLUSIONS

CD is very rarely located in the anterior mediastinum close to large blood vessels. Many of such cases have been misdiagnosed as invasive thymoma. The possibility of CD should be considered if preoperative CT of the anterior mediastinal parenchyma cannot exclude a lymph node mass. Furthermore, even when intraoperative findings reveal a mass in close proximity to the vessels without apparent gaps, the tumor is unlikely to progress into the interior of the vessels, owing to its non-invasive nature. Thus, it can be completely resected via careful surgical separation or removal of part of the vessel wall.

Conflicts of interest

The authors declare no conflict of interest.

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