Complete resection of unicentric Castleman disease in the superior mediastinum: A case report

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ABSTRACT

INTRODUCTION: Castleman disease (CD) is a rare benign lymphoproliferative disorder characterized by benign lymph node hyperplasia in a single site (unicentric CD [UCD]) or in multiple sites (multicentric CD [MCD]). Patients with UCD are usually asymptomatic; however, those with MCD usually develop fever, weight loss, and peripheral lymphadenopathy.

CASE PRESENTATION: We describe a case of surgically resected UCD in the superior mediastinum in which the involved lymph node was surrounded by important vessels and trachea, necessitating a median sternotomy for complete resection of the tumor.

DISCUSSION: Preoperative diagnosis of UCD in the thorax is very difficult, and surgical resection or excision is necessary. Complete resection is recommended for UCD because it is curative and has a 5-year survival rate of 100%.

CONCLUSION: UCD should be included in the differential diagnosis of asymptomatic mediastinal tumors. Surgical resection or excision is preferred to ensure an accurate diagnosis and appropriate treatment if malignant disease cannot otherwise be ruled out.

1. Introduction

Castleman disease (CD) is a rare benign lymphoproliferative disorder in which benign lymph node hyperplasia occurs in a single site (unicentric CD [UCD]) or in multiple sites (multicentric CD [MCD]). MCD was recently found to be associated with human herpes virus 8 (HHV-8) and human immunodeficiency virus (HIV) infections; however, its causes remain unclear. Patients with UCD are usually asymptomatic, and the diagnosis is achieved by imaging examination such as chest computed tomography (CT). Surgical resection is the optimal treatment for UCD. We herein present a rare case of complete surgical resection of UCD in the superior mediastinum.

2. Presentation of case

An abnormality was found incidentally on chest radiographs of a 77-year-old woman over a 2-year period and she was referred to our hospital for further examination and treatment. She had no remarkable abnormalities on physical examination, in particular, no peripheral lymphadenopathy, and no pertinent family history. Standard laboratory examination results were normal. The concentrations of most tumor markers were normal, including cytokeratin 19 fragment, pro-gastrin-releasing peptide, soluble interleukin-2 receptor, alpha-fetoprotein, and carcinoembryonic antigen; only the concentration of human chorionic gonadotropin was slightly increased at 1.8 mIU/mL (reference range, 0.0–0.7 mIU/mL). Chest CT scan revealed a smooth tumor in the superior mediastinum with relatively strong enhancement by contrast medium. The tumor was closely adjacent to other structures, including the aortic arch, brachiocephalic artery, left carotid artery, and trachea, but it was not invading any other tissues (Fig. 1). No hilar lymph nodes were enlarged. The preoperative differential diagnoses were thought thymoma, malignant lymphoma, teratoma and benign lymph node enlargement such as that associated with CD.

The patient was advised to undergo surgical resection for diagnosis and treatment and the mediastinal tumor was resected via median sternotomy. It was located adjacent to the aortic arch, brachiocephalic artery, left carotid artery, and trachea and behind the left brachiocephalic vein (Fig. 2A). Because intraoperative cytology examination indicated the possibility of lymphoma, the tumor was completely resected en bloc after separation from the surrounding tissues. It was a tan-pink color and measured 6.0 × 1.5 × 1.5 cm (Fig. 2B). The patient's postoperative course was uneventful, and she was discharged on the postoperative day 17.

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Histopathologic examination of the resected mediastinal tumor revealed hyaline-vascular-type CD [Fig. 3].

3. Discussion

CD, a rare benign lymphoproliferative disorder of unknown cause, was first described in 1954 [1]. Benign lymph node hyperplasia can occur in a single site (UCD) or in multiple sites (MCD) and located in the chest in approximately 70% of cases, in the neck in 15%, and in the abdomen and pelvis in 15% [2].

UCD characteristically develops in the third and fourth decades of life [3] and is usually asymptomatic with a benign course [2,4]. Imaging examinations such as CT, magnetic resonance imaging, and positron emission tomography/CT are usually performed; however, it is difficult to achieve a definitive diagnosis by imaging. In many cases, an accurate diagnosis is only made after resection of the tumor [5]. Mediastinal CD can mimic thymoma, malignant lymphoma, teratoma, and metastatic lymphadenopathy [2,3]. In the present case, we were unable to rule out the possibility of malignant disease and therefore performed complete surgical resection for both accurate diagnosis and curative treatment.

Complete surgical resection is the recommended treatment for UCD because it is curative and has a 5-year survival rate of 100% [6]. Resection under video-assisted thoracoscopic surgery has recently been reported [7]. In the present case, because the involved lymph node was surrounded by important vessels and the trachea, we performed a median sternotomy to enable complete resection of the tumor. It is important to select a surgical approach that is appropriate to the site of the lesion.

The clinical course of MCD is very different from that of UCD in many respects. MCD commonly presents in the sixth decade of life; however, individuals with HIV infection tend to develop it at a younger age [3]. MCD is usually associated with an immunodeficient states such as those associated with HIV or human herpes virus 8 infection [3,4,8]. Patients with MCD characteristically develop various symptoms, such as fever and weight loss. Treatment generally involves chemotherapy, antiviral medication, or antiproliferative regimens such as anti-interleukin 6 receptor or IL-6 therapy [2]. However, despite these treatments, 70% of patients with MCD die of infective complications and 30% of cancer [4]. The median survival following a diagnosis of MCD reportedly ranges from 14 to 30 months [8].

Fig. 1. Chest CT scan image showing a lesion (yellow arrows) in the superior mediastinum that is relatively strongly enhanced by contrast medium.
Fig. 2. (A) The tumor is located adjacent to the aortic arch, brachiocephalic artery, left carotid artery, and trachea. (B, upper) The tumor is a smooth, tan-pink, 6.0 × 1.5 × 1.5 cm mass. (B, lower) No necrosis or abscess was found inside the tumor.

Fig. 3. Photomicrographs showing lymphoid tissue with follicular hyperplasia. The follicles contain germinal centers are surrounded by follicular small lymphocytes. The germinal centers have multiple capillaries and hyalinization substances (yellow arrow). (A) Low-power view (hematoxylin-eosin, ×10). (B) High-power view (hematoxylin-eosin, ×20).

4. Conclusion

UCD should be included in the differential diagnosis of asymptomatic mediastinal tumors. Preoperative diagnosis of UCD is very difficult. Complete surgical resection is preferred for accurate diagnosis or treatment or both if malignant disease cannot otherwise be ruled out.

Conflict of interest

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Ethical approval

Not requested.
Author contribution

A.H. acquired the data and wrote the article. E.K. and Y.F. coordinated and critically revised the study. All read and approved the final manuscript.

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