CHINESE MEDICAL SCIENCES JOURNAL

CASE REPORT

Unicentric Castleman's Disease with Cardiovascular Involvement

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Key words: mediastinal tumor; unicentric Castleman's disease; surgery; cardiovascular involvement

Chin Med Sci J 2017; 32(3):198-200. DOI: 10.24920/J1001-9294.2017.030

CASTLEMAN'S disease (CD), a rare lymphoproliferative disorder of unknown etiology, was first described in 1956 as a benign mass in the mediastinum. Although CD can present anywhere in the body, 70% of the cases are in the chest along the tracheobronchial tree or hilum of the lung in the middle mediastinum; however, they can also occur in the anterior or posterior compartments. CD is classified as unicentric (UCD) or multicentric (MCD) based on the anatomical distribution, and histologically as hyaline-vascular, plasma cell, or mixed subtypes.¹ Although MCD is less common than UCD, it can be rapidly progressive and often fatal.² Systemic symptoms are rare with UCD, which has a good prognosis. UCD is typically of the hyaline-vascular type while most reported MCD cases have been the plasma cell type.³ UCD with cardiovascular involvement has been rarely reported in literature. We presented, from our center, two cases of UCD

of the hyaline-vascular type with cardiovascular involvement, and reviewed the literature with special emphasis on diagnosis and treatment.

CASE DESCRIPTION

Case 1

A 24-year-old male was transferred to our hospital with refractory pericardial and bilateral pleural effusion. Echocardiography revealed excellent left ventricular function. A whole-body PET/CT scan (Fig. 1) demonstrated an isolated mediastinal mass with high fluorodeoxyglucose uptake. Contrast-enhanced CT and magnetic resonance imaging revealed a solitary mediastinal vascularized mass in the aortopulmonary window. Due to the tumor's anatomical location and adhesions to surrounding structures, as well as the frozen section diagnosis, a complete resection necessitating transection of the ascending aorta was undertaken with cardiopulmonary bypass. A firm tumor measured 70 mm × 50 mm × 40 mm was removed and confirmed as UCD of the hyaline vascular type. The postoperative course was uneventful, and the patient was discharged from the hospital on the postoperative day 10. The

complete resection was confirmed by CT. The patient's systemic symptoms resolved quickly postoperatively. Thirty days after surgery, to address the pericardial involvement, a total of 40 Gy of radiation therapy was then delivered to the mediastinum over a 30-day period. There had been no evidence of progression or recurrence within 68 months of follow-up.

Case 2

A 23-year-old male with an asymptomatic mediastinal mass was admitted to our hospital because of the inconclusive diagnosis from a CT-guided fine-needle biopsy and an open biopsy *via* thoracotomy. CT scan demonstrated a solitary 60 mm × 50 mm × 40 mm mass with intralesion calcification, located anterior to the innominate veins and the origin of the superior vena cava (Fig. 2).

Figure 1. A whole-body PET/CT demonstrated an isolated mediastinal mass with high fluorodeoxyglucose uptake.

Figure 2. Computed tomography scan demonstrated a solitary 60 mm × 50 mm × 40 mm mass (arrow) with intralesion calcification, which was located anterior to the innominate veins and the origin of the superior vena cava.

An incomplete excision of the mass was performed because frozen section excluded malignancy and the hypervascular lesion had significant adhesions to the superior vena cava. The final histological findings were typical for the hyaline vascular form of UCD. The postoperative course was uneventful. Because of the infiltrative nature of the mass and the involvement of the superior vena cava, the patient was subsequently referred to radiation oncology, and received a total of 50 Gy of mediastinal radiation therapy over a period of 30 days. During 24 months of follow-up, no local progression, recurrence, or other problems were observed.

DISCUSSION

Since the first report in 1954, CD has been reported in approximately 1000 patients.⁴ The challenge with CD is establishing the diagnosis preoperatively because of the lack of disease-specific symptoms and signs. In most cases, the diagnosis of CD is dependent on postoperative histological examination, requiring either removal or biopsy of the lesion for definitive diagnosis. Preoperative imaging findings in cases of CD are nonspecific and frequently associated with a local mass and regional or multiple lymphadenopathy. On CT scan, the lesions may present as a homogeneous or heterogeneous soft tissue mass, and contrast enhancement depending on the injection rate and the volume of contrast media. Because CT reveals better the mass's borders, nature of the calcifications, and vascularity, it is important for clinical evaluation, management planning, and follow-up.

We advise that in cases of mediastinal masses displaying characteristics such as calcification or vascular enhancement, with or without systemic or local symptoms, the possibility of the diagnosis of CD should be seriously considered. Differential diagnoses include substernal goiter, thymus tumors, parathyroid tumors, lymphoma, germ cell tumors, schwannoma, or paraganglioma.

Open biopsy *via* thoracotomy or sternotomy is usually necessary for mediastinal CD because needle biopsy may yield inadequate tissue architecture and low diagnostic accuracy, while thoracoscopic biopsy is too dangerous for this highly vascularized tumor with marked adhesions.

Treatment for CD differs between unicentric and multicentric disease. Both of our UCD patients were treated and cured with surgical resection. A systematic review of 404 published cases showed that surgery is the gold standard for management of UCD.¹ However, if complete resection of the lesion is difficult or hazardous because of the vascular nature of the tumor and proximal critical structures,

incomplete resection may still be helpful because the recurrence rate after subtotal resection is low.⁵ Extended surgery is unnecessary because CD is a benign disease. For unresectable unicentric disease, radiotherapy is a safe option if it is based on the proximity of critical structures. However, although radiation may be effective, it is not curative and the results vary.⁶ Long-term follow-up is required to detect the development of malignancy.

The diagnosis of CD is ultimately established by histopathology, requiring either removal or biopsy of the lesion for a definitive diagnosis. The hyaline-vascular type of UCD has a good prognosis after complete surgical excision.

Conflict of Interest Statement

The authors have no conflict of interest to disclose.

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