Surgical Treatment of Castleman Disease Using Cardiopulmonary Bypass

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Castleman disease is a rare benign lymphoproliferative disorder of unknown etiology. A 29-year-old woman presented with intermittent chest pain for 2 years. Upon examination, computed tomography showed an intensely enhanced solid mass that encased her right pulmonary artery. The tumor was resected safely and completely via standard thoracotomy with cardiopulmonary bypass.

Key Words: Castleman disease • Middle mediastinal tumor • Surgery

INTRODUCTION

Castleman disease (CD) is a rare lymphoproliferative disorder of unknown etiology, which was first described by Dr. Benjamin Castleman in 1956.1 Morphologically CD has been divided into two clinical subtypes: a localized type and a multifocal type. The localized type occurs more commonly than the multifocal type and is usually localized in the mediastinum. It is widely accepted that surgical resection is the mainstay of management for localized mediastinal CD and the surgical treatment is variable, ranging from standard thoracotomy to minimally invasive video-assisted thoracoscopy (VATS).2-6 Because Castleman tumors are highly vascular and may have tight adhesions to the surrounding vital structures, they must be treated with great care and caution, particularly in the mediastinum. We have described a case of localized CD in the middle mediastinum that was successfully treated via open thoracotomy with cardiopulmonary bypass.

CASE REPORT

A 29-year-old female patient presented at our cardiothoracic clinic seeking for a second opinion of a mediastinal mass discovered at a local hospital. She had complained of intermittent chest pains for the previous 2 years, but had no fever or secondary symptoms of malignancy. Additionally, the patient had no significant medical or family history. Her physical examination and laboratory data including complete blood count and comprehensive biochemical parameters were within normal limits. Her chest radiograph showed a widened mediastinum and cardiomegaly; non-contrast enhanced computed tomography (CT) scan of the chest and abdomen revealed a solid mass (7.6 cm in maximal diameter) with central arborizing calcification in the middle mediastinum (Figure 1A). Contrast-enhanced CT demonstrated intense enhancement of the mass, which encased the right pulmonary artery (Figure 1B). Magnetic resonance imaging (MRI) and positron emission tomography scan were not performed. Tumor markers including α-feroproteins, β-human chorionic gonadotropin, lactate dehydrogenase, and carcinoembryonic antigen were normal.
We initially suspected a localized CD and the differential diagnoses included thymoma, intrathoracic goiter, neurogenic tumor, and germ cell tumor. The reason the surgeon decided to surgically remove this lesion was because localized CD have a high cure rate with complete surgical excision. Tumor resection was performed via median sternotomy under general anesthesia. The pericardial cradle was created for adequate exposure of the cardiovascular structures. The tumor mostly occupied the area within the aorto-pulmonary window and was highly vascularized, and could not be radically resected without cardiopulmonary bypass because it encased the right pulmonary artery. The patient was then put on the right femoral artery-bicaval cannulation. The systemic temperature of the patient was cooled down. The aorta was cross-clamped and heart was arrested by antegrade delivery of cool blood cardioplegic solution. The left heart was vented via the right superior pulmonary vein. The aorta was transected at the mid-ascending aortic level for adequate exposure of the tumor, and meticulous dissection along the tumor margin was performed and the tumor was freed from the aorta and pulmonary trunk. However, the right pulmonary artery (RPA) was encased by the tumor mass from its take-off down to the first branching. The RPA was resected with tumor and removed en-bloc, without damage to the surrounding structures. Mediastinal lymph nodes were also dissected out. The RPA was reconstructed with a 14-mm woven prosthesis as an interposition graft and the transected aorta was then reanastomosed. After evacuation of intracardiac air, the heart was reperfused with warm cardioplegia and the aortic clamp was relieved. After recovery of stable hemodynamics, decannulation was performed and systemic heparinization was reversed by protamine. Meticulous hemostasis was done and pericardial drain tubes were set, with the sternal halves wired.

On gross inspection, the resected tumor was a 7.0 × 5.5 × 4.5 cm and weighed 110 g. A segment of large blood vessel measuring 2.0 cm in length was encased by the tumor, and the color of cut surface of the tumor was yellow. Microscopic examination revealed marked interfollicular expansion, proliferative hyaline vessels, and some follicles were scatteredly distributed in the lymphoid tissue. The follicles had a marked expansion of the mantle zone and small inconspicuous germinal centers with branching hyalinization. The follicles were surrounded by concentric sheets of lymphocytes arranged in an onion-skin pattern. Hyalinized vessels were in the interfollicular area and occasionally penetrated lymphoid follicles radically forming a "lollipop" appearance. Histopathological findings of the tumor were consistent with a hyaline-vascular CD.

On the 3rd postoperative day, the patient complained of abdominal pain and fever, and abdominal ultrasonography revealed mild ascites and pleural effusion. Routine blood tests showed only leukocytosis without other abnormalities, and portable chest radiography showed cardiomegaly and a widened mediastinum. Non-contrast chest CT revealed bilateral pleural effusion and a large amount of pericardial effusion. A resternotomy procedure was then performed under the preliminary diagnosis of hematoma. The retained mediastinal hematoma was evacuated during the resternotomy procedure and the culture of the hematoma was negative. The patient otherwise recovered uneventfully and was discharged on the 19th postoperative day. Four months after surgery, the patient was doing well and a follow-up CT of the chest showed no residual tumor and no late-occurring complications (Figure 2).

**DISCUSSION**

Castleman disease is a rare form of lymph node hyperplasia, most commonly presenting as a solitary mediastinal mass, and was first described by Dr. Benjamin Castleman in 1956. Castleman disease is classified in two clinical subtypes: a localized type and a multifocal type, based on the extent of lymph node involvement.
Localized type occurs more frequently than the multifocal subtype, especially in the chest. The histopathologic classification divides CD into 4 types: hyaline-vascular CD, plasma cell CD, human herpes virus-8 (HHV-8) associated CD, and multicentric CD not otherwise specified. Additionally, hyaline-vascular CD accounts for approximately 90% of all cases. However, localized CD is the most common hyaline-vascular type.

Localized CD usually affects young patients less than 30 years old, and occurs equally in both men and women. The mediastinum is the most common location for localized CD, with less common extrathoracic sites being the neck, axilla, abdomen, and pelvis. Localized type is often asymptomatic at presentation and sometimes there are symptoms related to local pressure from the mass, like cough, chest pain, and dyspnea. Hypervascularity of this type of lesion had been reported to present as spontaneous hemothorax. Multifocal type frequently exhibits complicated systemic manifestations.

On CT scans, localized CD is depicted as well-circumscribed masses with soft-tissue attenuation. Calcification is uncommon in these lesions and can include punctate, coarse, or arborizing patterns, as shown in the present patient. On MR imaging, these highly vascular tumors appear solid and heterogeneous T1- and T2-hyperintensity compared with skeletal muscle. Upon PET, CD shows avid fluorodeoxyglucose (FDG) uptake.

As the recurrence of localized CD in the thorax is rare when the resection is complete, surgery is therefore the preferred treatment in these lesions. The surgical approach of localized CD is variable, from open thoracotomy to VATS in the thorax lesions. Recently, most benign mediastinal masses have been managed with VATS, as VATS was shown to be a safer, more effective and, less invasive than open thoracotomy. However, VATS probably should not be performed for resection of a localized mediastinal CD when there is the presence of tight adhesions of the tumor to surrounding vital structures. Tight adhesion and hypervascularity of localized mediastinal CD may cause profuse bleeding during surgery. Iyoda et al. have reported conversion to open thoracotomy when treating a posterior mediastinal CD due to profuse bleeding. Robert et al. advocated preoperative embolization prior to surgical resection to minimize intraoperative bleeding from hypervascular mediastinal masses. In our patient, the VATS approach cannot be used to treat the lesion because of the location of the tumor and encasement of the right pulmonary artery by the tumor. Thereafter, the tumor in our patient was resected safely via standard thoracotomy with cardiopulmonary bypass.

In conclusion, we reported a rare case of localized mediastinal CD that was treated safely and successfully by surgical resection employing thoracotomy with cardiopulmonary bypass. Our case suggests that tumor location, radiographic characteristics, and experience of the surgeons are important factors for choosing either VATS or open thoracotomy for the management of localized mediastinal CD. Although Castleman disease is uncommon, it should be included in the differential diagnosis of mediastinal tumors.

REFERENCES