Castleman’s disease (CD), also known as angiofollicular lymph node hyperplasia, is an uncommon, lymphoproliferative disorder of unknown etiology, mostly involving the mediastinum. Parenchymal lung involvement of the disease is extremely rare. Intrapulmonary CD has been reported in seven cases in the English literature. We describe an asymptomatic 28-year-old woman with lesion in the chest X-ray. Computed tomography (CT) of the chest confirmed a 5.5 × 5 cm well-defined, lobulated mass in the hilum of the right upper lobe. She underwent surgical resection for diagnosis and treatment. Pathologic examination showed hyaline vascular type (Castleman’s disease) lymph node hyperplasia. CD rarely arises from the intrapulmonary lymph nodes. In these patients, preoperative diagnosis is difficult and invasive attempts may be required.

Keywords: Castleman’s disease, intrapulmonary, resection

Introduction

Castleman’s disease (CD) is an uncommon, lymphoproliferative disorder of unknown etiology. It has two distinct clinical forms: unicentric and multicentric. CD is histologically subdivided into hyaline vascular, plasma cell, and mixed types. Unicentric CD is usually of the hyaline vascular type and multicentric disease of the plasma cell or mixed type. Localized CD requires surgery, and complete excision allows full recovery. Corticosteroid and anti-neoplastic chemotherapy are suitable for the multicentric disease. The majority of the unicentric cases are seen in the thorax, especially in the mediastinum. Intrapulmonary CD is exceedingly rare.1–3

Herein, we report an asymptomatic case of unicentric, intrapulmonary CD, who underwent curative surgery.

Case Report

A 28-year-old woman with lesion in the chest X-ray presented to the thoracic surgery department. The patient was asymptomatic with no past medical history. Hematological and biochemical parameters of the patient were within normal limits. HIV-serology was normal. Chest X-ray showed a giant regular lesion in the right hilum. Computed tomography (CT) of the chest demonstrated a 5.5 × 5 cm well-defined, lobulated mass in the hilum of the right upper lobe (Fig. 1A and 1B). For the diagnosis, fiberoptic bronchoscopy was performed under the local anesthesia. Bronchoscopic examination revealed slightly bronchial compression at the medial of the upper lobe bronchus. Endobronchial lesion was not detected. Thoracotomy was performed. Approximately a 5 × 6 × 4 cm regular, stiff mass was detected encircling the proximal
right upper lobe bronchus. Branches of the upper lobe pulmonary artery were seen in the mass. Limited resection and biopsy was not achieved because of the hard adhesions and hypervascularity of the tumor. Right upper lobectomy with mediastinal lymph node dissection was performed since pulmonary malignancy could not be ruled out. The patients was discharged on the 6th postoperative day without complication. Pathologic examination showed hyaline follicles with interfollicular capillary proliferation. On immunohistochemical staining, the hyperplastic cells were positive for CD3, CD20 and negative for Cyclin D1, HHV8. Results of the histological evaluation revealed hyaline vascular type (Castleman’s disease) lymph node hyperplasia (Fig. 2). Histology of resected lymph node was inflammation.

**Comments**

Castleman’s disease (CD), first reported in 1954 by Castleman and Towne, is an uncommon lymphoproliferative disorder of unknown etiology that can occur wherever lymph nodes are present. CD is clinically classified into unicentric (localized) and multicentric (systemic) and then further subdivided histologically into the hyaline vascular (90% of cases), plasma cell (8%–9% of cases), and mixed types (1%–2% of cases). The localized type is a rare form and usually detected incidentally in asymptomatic cases as in our case. Intrapulmonary development of CD is very rare and only 7 cases have been reported in the English Literature (Table 1). Including the present case, 6 patients were treated by lobectomy,1,3,4 2 patients were treated by tumor excision. Excessive bleeding was reported in the last 2 cases with limited resection.2,5 In the diagnosed cases, preoperative arterial embolization may reduce the intraoperative bleeding.4 Limited resection may be tried in the appropriate cases. In our case, mass was located in the central of the upper lobe, and the branches of upper lobe pulmonary artery were invaded by the mass. Thus, we preferred the anatomically resection.

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**Fig. 1** Chest X ray: Mass lesion located in the right hiler region (A). Computed tomography scan of the chest show a well-defined, lobulated, soft tissue mass in the central in the right upper lobe. Upper lobe pulmonary artery brunches were covered by lesion (B).

**Fig. 2** Histopathologic examination showing the characteristic pale concentric (expanded) mantle zone known as “onion skinning” and germinal centers fed by prominent radially penetrating sclerotic vessels, lollipop-like appearance. In addition, lymphoid tissue seen next to the alveol (black arrow) in the small picture (HE × 200 and 20).
Since clinical and radiological findings are not specific, the diagnosis of the CD is difficult. CT shows a solid homogenous soft-tissue mass with clear contrast enhancement. The value of the PET/CT in the preoperative diagnosis is low because it is inadequate to differentiate the malign diseases and benign diseases. Castleman disease can be a cause of false-positive findings by mimicking metastatic adenopathy. The diagnosis is very difficult in these patients, and a thoracotomy is usually needed for the diagnosis and treatment as our case.

CD should be considered in the differential diagnosis of the asymptomatic, relatively well-defined mass in the lung. Localized CD requires surgery, and complete surgical resection allows full recovery in all cases.

**Disclosure Statement**

We have no financial or other interest in the manufacture or distribution of the device.

**References**


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Table 1 Characteristics of patients with intrapulmonary Castleman’s disease

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LUL: left upper lobe; LLL: left lower lobe; RLL: right lower lobe; RUL: right upper lobe; H-V: hyaline vascular type.