

# Resection of unicentric interlobar Castleman disease with following adjuvant radiotherapy

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## Abstract

A 22-year-old female patient with rare interlobar unicentric Castleman disease is presented. The tumour was discovered incidentally and thoracoscopic biopsy was planned to rule out malignancy. Due to dense adhesions to the adjacent anatomical structures and diffuse bleeding when mobilizing the tumour, a thoracoscopic approach was converted to thoracotomy. The tumour was removed without lung resection. Adjuvant radiotherapy was used to avoid possible recurrence of the disease. During the follow-up of 6 years, the patient remains free of any symptoms and evidence of recurrence.

## Keywords

Castleman disease, interlobar, surgery, radiotherapy

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## Introduction

Castleman disease (CD), known as a giant lymph node hyperplasia or angiofollicular lymph node hyperplasia, is a rare lymphoproliferative disorder described first in 1956 by Castleman et al.<sup>1</sup> CD has two main forms: unicentric (affects regional lymph nodes mainly in chest or abdomen) and multicentric (disseminated, acting like malignant lymphoma, affecting multiple lymph nodes with potential involvement of visceral organs). Two main histopathological subtypes of CD can be distinguished: hyaline vascular type (unicentric, considered benign) and plasma cell type (multicentric, aggressive). Also, a mixed type form of CD can occur.<sup>2</sup> Clinical significance of CD originates from neoplastic potential that may manifest in the development of various lymphoproliferative malignancies, that is, Hodgkin lymphomas, follicular dendritic cell sarcomas, extranodal B-cell lymphomas and Kaposi sarcomas.<sup>3</sup>

Patients are usually presented with incidental solitary mass in the chest (71%), which can occur in every level of the tracheo-broncho-pulmonary lymphatic chain (so-called nodal CD).<sup>4</sup> Other origins (incl. extranodal involvement) such as thoracic wall, retroperitoneal space, pelvis, neck, subcutaneous tissue, intramuscular, spleen and liver have been reported less frequently.<sup>2</sup> Unicentric CD is generally asymptomatic, whereas multicentric type may present various systemic signs, that is, asthenia, pyrexia, hyperhidrosis,

anaemia, arthralgia, peripheral neuropathy, dysproteinemia and thrombocytopenia.<sup>5</sup> CD can occur in all age groups; peak incidence is between 20 and 40 years.<sup>4,5</sup> No sexual predominance or specific physical examination finding and laboratory test exist.<sup>4</sup> Diagnosis of CD requires thorough morphological investigation, since intraoperative frozen section can be misleading being non-diagnostic or misdiagnosed as lymphoma.<sup>4,6</sup> Radiologic imaging (chest radiography, computed tomography (CT), magnetic resonance imaging (MRI)) shows only localization of usually well-circumscribed tumour with constant or various contrast enhancement—resembling lung cancer.<sup>6</sup>

Surgery is considered standard therapy for the localized form of CD.<sup>7</sup> However, in unresectable cases or suspicion of residual tumours after surgery, (neo-)adjuvant therapies can

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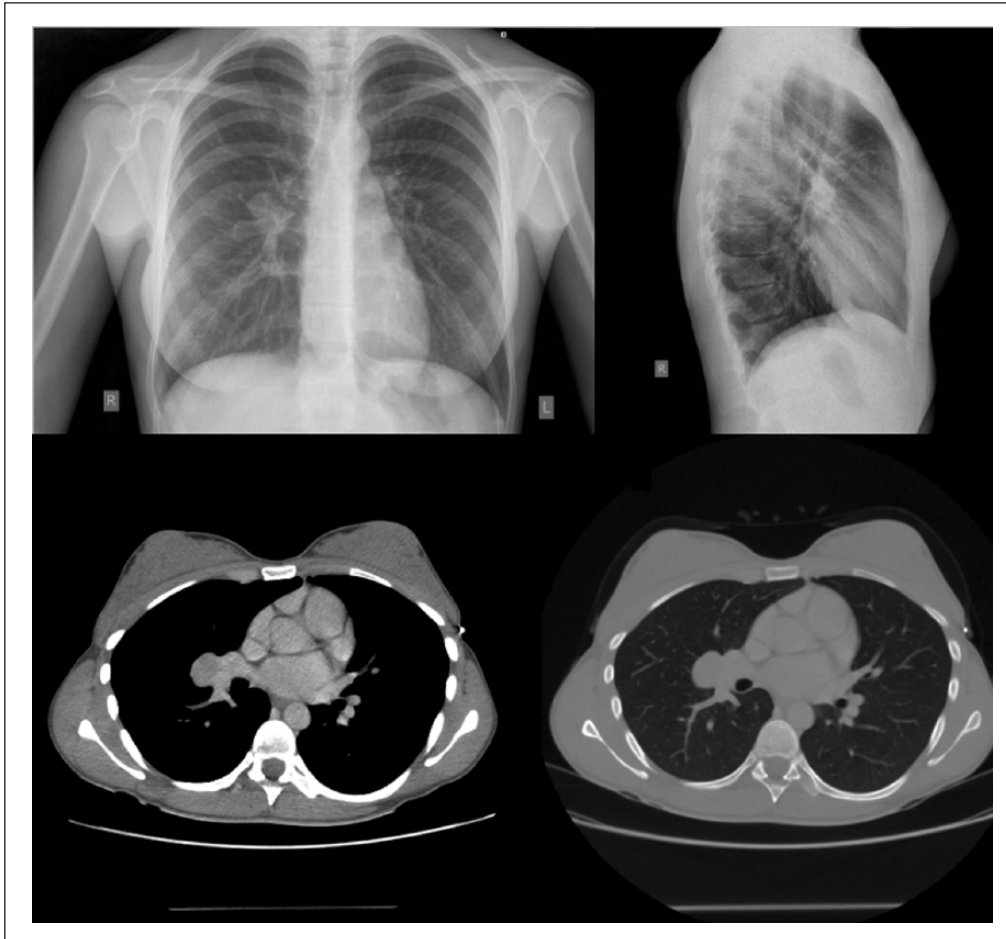
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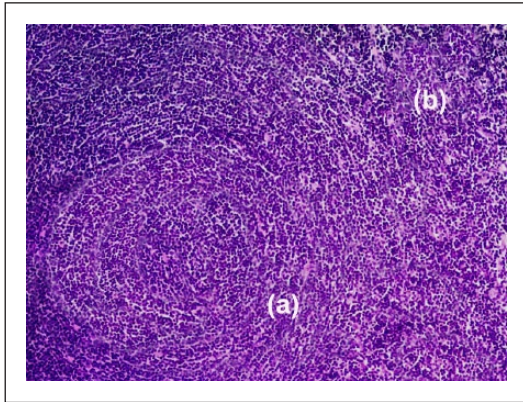
**Figure 1.** Posteroanterior and lateral chest radiographs, demonstrating well-circumscribed right hilar tumour. At chest CT, more detailed view of contrast-enhancing tumour in the right interlobar fissure can be seen.

be used.<sup>8,9</sup> Although a benign disease, recurrences after up to 20 years of follow-up have been described.<sup>10</sup>

### Case report

We report a case of interlobar CD, treated successfully with surgery and adjuvant radiotherapy. A 22-year-old woman, asymptomatic, non-smoker, without history of any chronic illness (including tuberculosis) was admitted to thoracic surgery department due to incidental discovery of right parahilar tumour on prophylactic chest radiograph (Figure 1). No evidence of the lesion was present 3 years earlier. There are no clear indications why chest radiographs were ordered by family physician and occupational specialist at that time. Bronchoscopy did not reveal any endobronchial abnormalities, and the bronchioalveolar lavage cytology was normal. Pulmonary function and routine blood tests were within normal range. Chest CT scan demonstrated well-circumscribed, contrast-enhancing tumour in the right horizontal fissure, tightly adherent to branches of pulmonary artery and upper pulmonary vein (Figure 1). No mediastinal lymphadenopathy, any other intrapulmonary or intraabdominal lesions were detected.

Since malignancy could not be ruled out, the right-sided video-assisted thoracoscopy (VATS) and biopsy of the tumour was planned. VATS was performed using three troacars. Inspection of the right hemithorax revealed relatively well-circumscribed, encapsulated solitary mass in horizontal fissure tightly adherent to adjacent interlobar vasculature. Tumour showed hypervascular features, and despite careful dissection, noticeable diffuse bleeding occurred. Complete macroscopic excision of the tumour was considered to be possible, but too risky thoracoscopically. Conversion to conventional axillary thoracotomy was made and tumour excised. Total blood loss was approximately 300 mL. The postoperative period was uneventful and the patient was discharged home on the fifth postoperative day. A pathological examination revealed a  $2.5 \times 2.5 \times 2$  cm<sup>3</sup> tumour with bright-rose colouration, morphologically consistent with hyaline vascular type of CD without any evidence of malignant transformation (Figure 2). Although the tumour was macroscopically removed, we were doubtful about the radicality of the operation. Margins of apparently tumour-free tissue around the lesion was hard to assess microscopically due to avoidance of extended resection; thus, there was a risk of potential R1 – resection. Because of the tight adherence to



**Figure 2.** Histopathological features of CD – hyaline vascular type lymphoid follicle (H&E, 100×) which contains (a) onion-skinned mantle zone and (b) interfollicular lymphohistiocytic cells.

pulmonary vessels, as well as to lung parenchyma and possible invasive nature of the tumour, the pulmonary oncology council suggested adjuvant radiation therapy. The patient received three-dimensional conformal radiotherapy to the site of operation in once daily 2 Gy fractions to cumulative radiation dose of 44 Gy. During 6-year follow-up period, the patient is free from recurrence. No complication from surgery or adjuvant radiotherapy has occurred.

## Discussion

CD can occur in many different lymph node stations within the chest or in other regions of the body.<sup>2,4</sup> Although mediastinal lymph nodes are most commonly affected, we observed CD in interlobar fissure, which is a very rare location – only single cases have been reported in the literature.<sup>4,6</sup> This location makes the preoperative diagnostics rather difficult as it is not easily accessible for either endobronchial ultrasound or transthoracic fine needle biopsy. Therefore, VATS biopsy or excision of the tumour is often needed to rule out malignancy and to establish the diagnosis.

In majority of cases, surgical excision of the tumour is considered to be the best option for the treatment of localized CD. Thoroscopic resection of unicentric CD has been successfully performed alternatively to conventional open thoracotomy;<sup>6</sup> however, in several cases, conversion to open approach had to be performed.<sup>4,11</sup> Main reasons for conversion are inability to perform complete resection (tumour invasion to adjacent structures) or intraoperative bleeding due to hypervascularity of the tumour.<sup>4,6,11</sup> Therefore, it is probably advisable to start the operation thoroscopically, but without hesitation to convert it into open approach in case of dense adherence of the tumour or risk of serious bleeding. According to previous series of CD, in majority of operations, remarkable bleeding had occurred with mean blood loss value of 620 mL.<sup>6</sup> Intraparenchymal or interlobar location of the CD makes the excision even more demanding. For

example, lobectomy has been performed due to location of the tumour inside the lobe parenchyma or pneumonectomy due to profuse tumour bleeding and extension of the tumour to the adjacent lobes. Moreover, in several reports, dense adhesions<sup>4</sup> and even invasion into adjacent anatomical structures, including myocardium, pericardium, right atrium, coronary arteries, great vessels, peripheral pulmonary parenchyma, vertebral body, rib and chest wall,<sup>6,12,13</sup> necessitating more extensive resections have been described.

In most unicentric CD cases, radical resections can be performed since hyaline vascular type of tumour is generally encapsulated and well-circumscribed. Incomplete resection margins may, however, result in a slow enlargement of the remaining tissue and the development of recurrence,<sup>3,4,10,13</sup> indicating the need for postoperative additional therapy. Adjuvant chemotherapy has not been used in the treatment of unicentric CD. Previous experience with adjuvant radiotherapy is rather limited, but in situations where completeness of the tumour removal is questionable, it is reasonable to assume that adjuvant radiotherapy decreases the risk of tumour recurrence. In fact, published case reports have confirmed excellent local control rates of unicentric CD with adjuvant radiotherapy (27–50 Gy) in thoracic,<sup>8,14</sup> as well as in other regions of the human body including brain.<sup>15</sup> Similarly, in our case of interlobar CD, adjuvant radiotherapy (44 Gy) was used due to the avoidance of the resection of lung parenchyma, tight tumour adherence to pulmonary vessels and possible invasive nature of the disease. After 6 years of follow-up, our patient is still free from local recurrence and do not experience any treatment-related side effects.

## Conclusion

Combining surgery and postoperative radiotherapy in the management of unicentric CD allowed us to avoid resection of lung parenchyma and resulted in well-tolerated curative outcome.

## Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

## Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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## Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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