

# Castleman's disease: A rare indication for endovascular therapy for hemoptysis

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## Abstract

Castleman's disease (CD) is a rare lympho-proliferative disorder due to faulty immune regulation resulting in proliferation of lymphatic tissue. The vascular supply to these lesions have been reported to arise from the bronchial, internal mammary and the intercostal arteries. We report a case of hemoptysis secondary to intrathoracic CD with vascular supply arising from the left inferior phrenic artery which was successfully embolised with polyvinyl alcohol (PVA) particles.

**Key words:** Atypical radiological features; Castleman's disease; hemoptysis; inferior phrenic artery

## Introduction

Castleman's disease (CD) is a rare lymphoproliferative disorder caused by faulty immune regulation resulting in excessive B-lymphocyte and plasma-cell proliferation in lymphatic tissue.<sup>[1]</sup> Marked interfollicular vascular proliferation leads to mass formation, which can encase bronchi and sometimes cause mural erosion leading to life-threatening hemoptysis. Vascular supply to these lesions have been reported to arise from the bronchial, internal mammary, and intercostal arteries.<sup>[2]</sup> We report a case of hilar intrathoracic CD with atypical radiological features where the left inferior phrenic artery and bronchial circulation provided codominant blood supply.

## Case Report

A 24-year-old female with a 5-month history of wheeze and nonproductive cough underwent chest X-ray, which showed a well-defined mass in the left hilar region [Figure 1A-D]. A contrast-enhanced computed tomography (CT) scan of

the chest revealed a heterogeneously enhancing 7 × 5 cm mass containing flecks of calcification at the left hilum. 18 fluorodeoxyglucose positron emission tomography (18-FDG PET) scan demonstrated an inhomogeneous hypermetabolic lesion with no evidence of metastatic uptake. A CT-guided percutaneous core biopsy revealed chronic inflammatory tissue with no evidence of malignancy. No definitive histological diagnosis was possible. Due to worsening cough and wheeze, the patient underwent a thoracotomy and surgical resection. During the operation there was massive blood loss of 2000 ml. Histopathology confirmed the mass to have marked lymphoid infiltrate with irregular B cell follicles and hyaline vascular-like change consistent with CD.

Two months after the thoracotomy, the patient presented to the emergency department with a 1-day history of massive hemoptysis (>200 ml). A CT pulmonary angiogram for clinically suspected pulmonary embolism showed no pulmonary embolism, however, multiplanar

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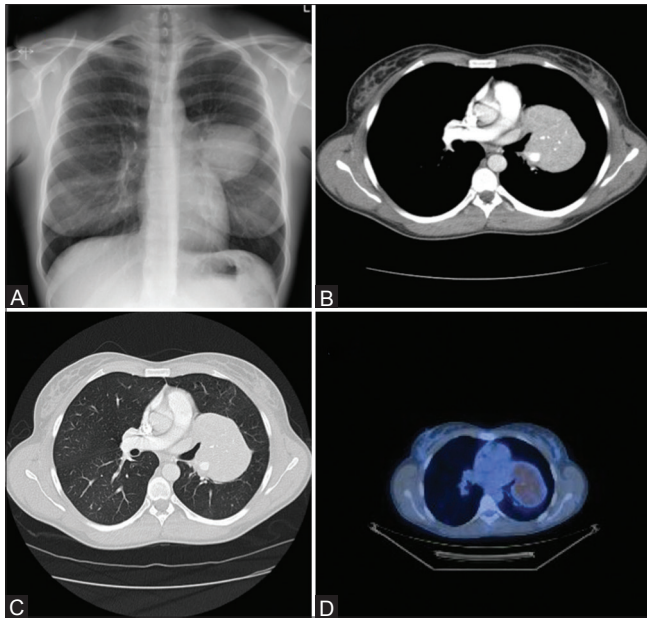
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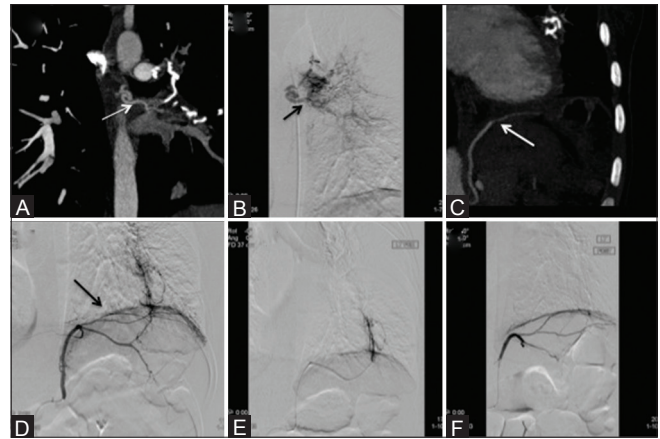


**Figure 1 (A-D):** Chest X-ray of a 24-year-old female (A) showing well-defined left hilar mass. Contrast-enhanced CT scan (B and C) shows heterogeneously enhancing mass with coarse calcifications within this hilar mass. 18-FDG PET/CT scan (D) shows hypermetabolic uptake within the lesion

reformats showed low attenuation soft tissue in the surgical resection bed at the left hilum receiving blood supply through hypertrophied left bronchial and left inferior phrenic arteries [Figure 2A-F]. Flexible bronchoscopy showed fresh blood in the left lower lobe bronchus. The patient had an embolization through a right common femoral artery approach and the common (conjoined) origin of the right broncho-superior intercostal trunk and left bronchial arteries were catheterized with a 5 Fr (Cordis, USA) left Amplatz coronary I catheter (Cordis, USA). The sharply angulated left bronchial artery could not be superselectively catheterized with a range of microcatheters. The main trunk was embolized with 300–500 micron polyvinyl alcohol (PVA) particles (Cook, USA). The medial division of the inferior phrenic artery was seen to supply the mass via its pericardial branch. This was super-selectively catheterized with the same 5 Fr catheter and a Renegade microcatheter (Boston Scientific, USA) and embolized with the same PVA particles. No spinal, esophageal, or coronary supply was demonstrated from the bronchial and left inferior phrenic arteries. There were no procedure-related complications and the patient had an uneventful recovery with no further hemoptysis during the 3 years follow-up period.

## Discussion

CD may be localized or multicentric and primarily involves the mediastinum, neck, and mesentery.<sup>[1]</sup> There are three known histological variants namely (a) hyaline-vascular, (b) plasma-cell, and (c) mixed variant. CD can present



**Figure 2 (A-F):** Contrast-enhanced CT with MIP images (A and C) showing blood supply to the lesion with bronchial artery and left inferior phrenic artery (arrows). Catheter angiogram demonstrates bronchial artery (arrow in B) and left inferior phrenic artery (arrow in D) and subsequent embolization with PVA (E and F)

as a mass in the mediastinum, neck, or mesentery. Classic features on a CT scan is a solitary, intensely, and homogeneously enhancing mass with no local invasion. Calcification is present in only 5–10% of the cases.<sup>[2]</sup> Our patient, however, had atypical findings of inhomogeneous enhancement and calcifications.

The standard treatment of the localized form is surgical resection, however, where there is encasement or invasion of the adjacent structures, preoperative embolization has been advocated.<sup>[3,4]</sup> Recently, embolization alone without surgical resection has been shown to terminate hemoptysis and alleviate shortness of breath due to airway compression in patients with unresectable disease.<sup>[5]</sup>

In our patient, the diagnosis of CD was only made after thoracotomy and resection. Preoperative embolization, which may have reduced the large volume intraoperative blood loss, was not considered. It was only when the patient presented with hemoptysis after surgical resection that the vascular supply was identified and an emergency referral to interventional radiology was made.

Previous reports have identified bronchial, internal mammary, and intercostal arteries as feeding vessels to CD.<sup>[2]</sup> Inferior phrenic supply to CD has not been reported before to the best of our knowledge. When basal lung segments are suspected as the source of hemoptysis, inferior phrenic supply should always be considered. Inferior phrenic supply to hilar masses is much rarer, but when it occurs, the supply is usually via the pericardial branch of its medial division. The left inferior phrenic medial branch may supply the esophagus, and it is important to exclude any esophageal supply before particulate embolization. It commonly communicates with the pericardiophrenic branch of the internal mammary artery.

Careful review of multiplanar reformats of the CT scan aided procedure planning and the consent process by identifying the hypertrophied arterial feeders.

## Conclusion

CD should be considered in the differential diagnosis for all benign intrathoracic masses. Where a mass shows avid enhancement, whether homogeneous or heterogeneous, a CT angiogram, should be considered to identify hypertrophied feeding arteries. Preoperative embolization can reduce intraoperative blood loss in appropriately selected patients.

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## Conflicts of interest

There are no conflicts of interest.

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