Case Report

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Incidentally Detected Castleman Disease of the Thorax and its Surgical Management: A Case Report

Surgery Section

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ABSTRACT

Castleman Disease (CD) is a rare lymphoproliferative disorder, the aetiology and pathogenesis of which remain unclear. It most commonly presents as a localised form, known as Unicentric CD (UCD), and less frequently as a generalised form, termed Multicentric CD (MCD). The thorax is the most common site of UCD; however, due to its rarity, UCD is seldom included in the differential diagnosis of an intrathoracic mass. Preoperative identification of CD is challenging. On imaging, CD typically appears as a well-defined, homogeneous mass. A characteristic feature of UCD on Contrast-Enhanced CT (CECT) is intense enhancement during the arterial phase, which decreases in the portal venous phase. This intense enhancement is attributed to the hypervascular nature of UCD. Therefore, multiphase CECT may be considered the investigation of choice when UCD is suspected. The lesion is highly vascular and often exhibits dense adhesions with adjacent organs, making surgical resection challenging. Authors report a case of a posterior mediastinal mass located in the left paraspinal region, which was incidentally detected in a 53-year-old female. The mass was successfully resected via a left postero-lateral thoracotomy and was postoperatively diagnosed on Histopathological Examination (HPE) as Castleman Disease, Hyaline Vascular Variant (HVV). The postoperative course was uneventful. Preoperative suspicion of CD would aid surgeons in planning the procedure appropriately and help avoid unexpected findings during surgery.

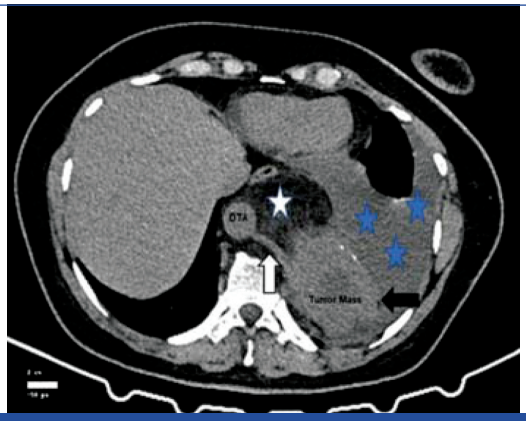
Keywords: Fibroblastic, Liposarcoma, Lymphoproliferative disorders, Thoracotomy, Tumour

CASE REPORT

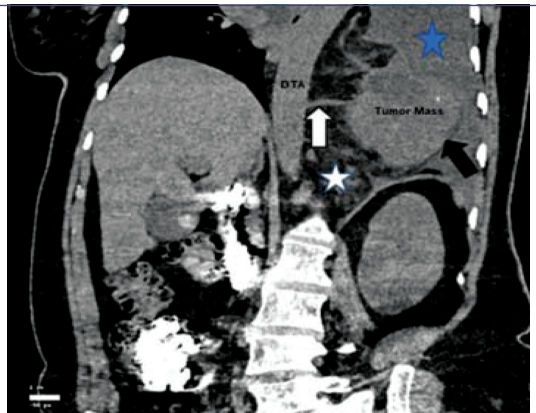
A 53-year-old female patient was asymptomatic and presented with a report of CECT of the chest and whole abdomen, performed at another centre during evaluation for fibroid uterus. The scan was reported as showing a hiatus hernia with herniation of omental fat through a widened hiatus, with a heterogeneous lobulated lesion within the herniated omental fat. In addition, there was significant left pleural effusion with passive atelectasis of the left lung. The patient had no symptoms related to the hiatus hernia.

As the diagnosis was uncertain, re-evaluation of images with our in-house radiologist was done. On review, the findings suggested a large, well-defined posterior mediastinal mass measuring approximately 8×8 cm in the left hemithorax, located in the left paraspinal region and abutting the Descending Thoracic Aorta (DTA), with preserved fat planes (suggestive of liposarcoma) [Table/Fig-1,2]. The mass demonstrated intense contrast enhancement [Table/Fig-3]. A CT-guided biopsy of the mass was reported on histopathology as a possible inflammatory myofibroblastic tumour. No malignant cells were detected on diagnostic pleural aspiration.

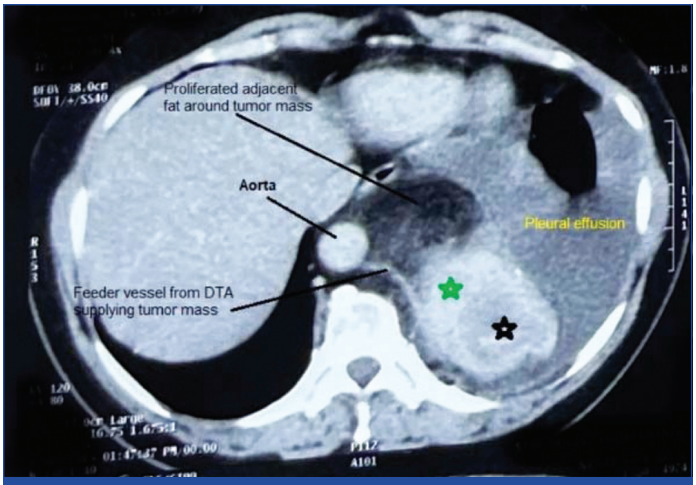
After assessing the resectability of the mass, the patient was taken up for excision via left posterolateral thoracotomy. Intraoperatively, a large lobulated mass measuring approximately 10×10×8 cm, firm in consistency, was identified in the paraspinal aspect of the left hemithorax. It was adherent to the DTA, with feeder vessels arising from it, and densely adherent to the pleura and the left hemidiaphragm. Intraoperatively, the mass appeared malignant. The lesion was carefully dissected free from the DTA after ligation of feeder vessels, which required the temporary application of a partial cross-clamp to control bleeding from the DTA. The mass was then mobilised from the left hemidiaphragm and pleura using a combination of blunt and sharp dissection. Oxygenated regenerated cellulose (Clinicel Knitted, Healthium Medtech, India) was applied to achieve hemostasis after resection. Approximately 700 mL of straw-



[Table/Fig-1]: Non-contrast CT (NCCT) chest (axial view) showing a well-defined hypodense soft-tissue density mass (black arrow) in the posterior mediastinum with eccentric specs of calcification, with proliferation of adjacent fat (white star). Note the presence of a large feeder vessel from DTA (white arrow) and left pleural effusion (blue star).



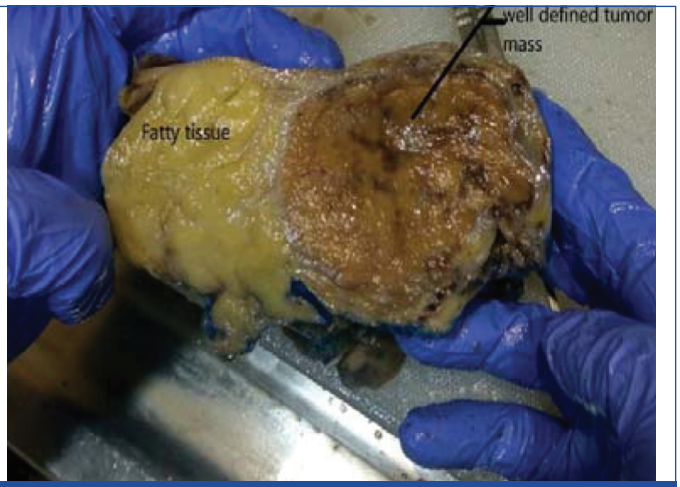
[Table/Fig-2]: NCCT chest (coronal reformatted view) shows a well-defined mass (black arrow) located above left hemidiaphragm with proliferation of adjacent fat (white star). Note the presence of a large feeder vessel from DTA (white arrow) and left pleural effusion (blue star).



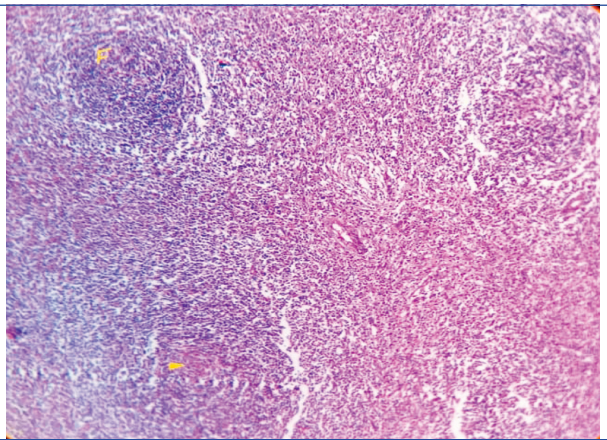
[Table/Fig-3]: CECT chest (axial view) showing avidly enhancing well defined mass (green star) with central non-enhancing areas suggestive of central necrosis (black star). Also note a large feeder vessel from DTA. The CT scan image in arterial phase of contrast enhancement.

coloured pleural fluid was aspirated. The incision was closed using triclosan coated polyglactin 910 Trusynth plus neo suture.

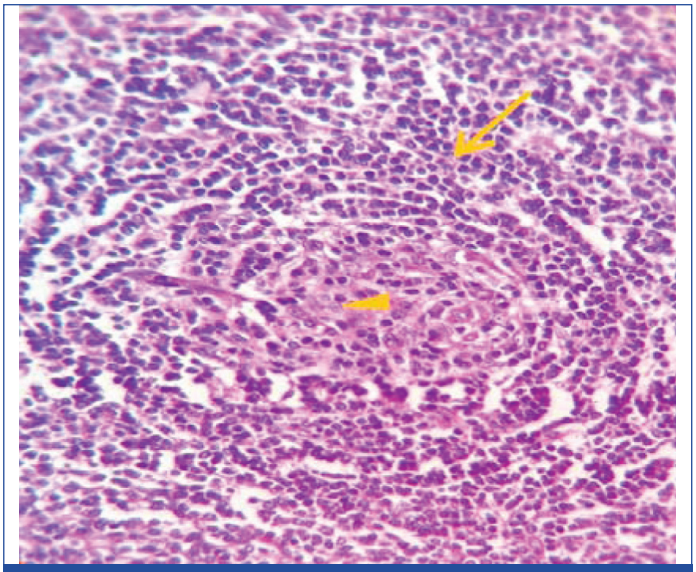
The postoperative course was uneventful, and the patient was discharged in stable condition on the 5th postoperative day (POD). Gross examination of the resected specimen revealed a well-defined tumour surrounded by fatty tissue on one end [Table/Fig-4]. Histopathological Examination (HPE) of the excised mass showed the characteristic "onion-skin" pattern of lymphoid follicles, with concentric layers of lymphocytes surrounding an atrophic germinal centre, consistent with Castleman Disease, Hyaline Vascular Variant (HVV) [Table/Fig-5,6].



[Table/Fig-4]: Cut-section of the operative specimen shows a well-defined tumour surrounded by fatty tissue on another end.



[Table/Fig-5]: Photomicrograph shows lymphoid follicles (F) with atrophic germinal centres and central hyalinisation (yellow arrowhead). Interfollicular stroma shows vascular and fibroblastic proliferation along with numerous lymphocytes and plasma cells (H&E, 20x).



[Table/Fig-6]: Photomicrograph shows lymphoid follicle having concentric layering of lymphocytes in an onion-skin pattern (yellow arrow) encircling an atrophic germinal centre (yellow arrowhead). This so-called targetoid onion-skin appearance and is highly suggestive of CD (H&E, 100x).

DISCUSSION

Castleman Disease (CD) is a rare, non malignant lymphoproliferative disorder of uncertain aetiology, first described by Benjamin Castleman in 1954 [1]. It is also referred to as giant lymph node (LN) hyperplasia or angiofollicular hyperplasia. Although CD can present at any age, its peak incidence occurs in the 3rd and 4th decades of life [2]. Clinically, CD can be classified (based on location) into two types: Unicentric CD (UCD, localised disease) and Multicentric CD (MCD, diffuse or systemic disease). Histopathologically, CD is divided into two variants: Hyaline Vascular Variant (HVV) and Plasma Cell Variant (PCV). UCD is more common (68–98%) than MCD. Furthermore, the majority of UCD cases are of the HVV type, whereas most MCD cases are PCV [2,3].

UCD is usually asymptomatic and detected incidentally, whereas MCD often presents with constitutional symptoms such as fever, night sweats, and weight loss. Occasionally, UCD may cause symptoms due to mass effect on adjacent structures, such as cough, dysphagia, dyspnoea, or pain [4]. Although CD can occur at any site where LN tissue is present, the thorax is the most common site (70%). Other sites include the abdomen and pelvis (10-15%) and the neck (10-15%). Within the thorax, the mediastinum is the most frequent location, although UCD may also arise in the lung hilum, pleura, or chest wall [5].

Preoperative identification of CD is challenging. CD typically presents as a solitary, well-circumscribed, enhancing mass. Feeding vessels arising from major arteries are characteristic, and adjacent fat proliferation may be seen. The differential diagnosis includes:

a) Lymphoma – usually associated with systemic symptoms (fever, weight loss, night sweats), often lacks intense vascularity, commonly presents with multiple lymphadenopathies, and typically does not show feeding vessels from the aorta.

b) Thymoma/Thymic carcinoma – usually arises in the anterior mediastinum, may contain calcifications or cystic changes, and is less likely to demonstrate prominent feeding vessels or intense enhancement on CT.

c) Paraganglioma – appears as a highly vascular, intensely enhancing lesion with possible feeding vessels on CT. It may mimic CD radiologically but is more likely to secrete catecholamines, detectable clinically or biochemically.

On imaging, certain features help differentiate CD from other mediastinal masses: A well-defined, homogeneous mass; proliferation of adjacent fat; absence of necrosis or calcification; and

no invasion of surrounding structures (unlike aggressive tumours). The hallmark feature of UCD on CECT is intense enhancement in the arterial phase with decreased enhancement in the portal venous phase. This finding reflects the hypervascular nature of UCD. Thus, multiphase CECT may be considered the investigation of choice when UCD is suspected [6].

Recognising or suspecting UCD preoperatively is valuable for surgical planning. Localised UCD is amenable to complete surgical resection, which is usually curative. However, UCD is highly vascular and often demonstrates dense adhesions with adjacent organs or major vessels, making surgical resection challenging. In such cases, preoperative embolisation may be considered to reduce vascularity [7], or resection may be performed under Cardiopulmonary Bypass (CPB) if there are dense adhesions to major vessels [8]. Although Video-Assisted Thoracoscopic Surgery (VATS) is associated with less postoperative pain and shorter hospital stay, its role in CD appears limited due to the dense adhesions and vascularity of the disease [8].

Recently, most benign mediastinal masses have been managed with VATS. Compared with ultrasound-guided endoscopic fine-needle aspiration and percutaneous transthoracic puncture biopsy—which are less invasive and easier to perform but carry the risk of damage to critical structures such as large blood vessels or airways when sampling mediastinal tumours—VATS provides adequate exposure of the thoracic cavity, allowing for either biopsy or complete excision.

For mediastinal and posterior mediastinal masses of unknown origin, particularly in patients with compressive symptoms involving adjacent structures such as dysphagia, VATS can be used as an exploratory approach. However, it should be promptly converted to thoracotomy if thoracoscopic dissection proves difficult [9]. VATS is generally not recommended for resection of localised mediastinal CD when there are dense adhesions between the tumour and surrounding vital structures. The tight adhesions and hypervascularity of localised mediastinal CD can lead to profuse intraoperative bleeding [8,9]. Iyoda A et al., reported conversion to open thoracotomy during the treatment of a posterior mediastinal CD due to excessive bleeding [10].

The prognosis of UCD is favourable, as it behaves like a benign disease, and recurrence is rare; thus, complete surgical resection is considered curative. By contrast, MCD often presents as an aggressive lymphoproliferative disorder, for which debulking surgery and/or immunochemotherapy may be required [5].

CONCLUSION(S)

In conclusion, UCD is a rare cause of intrathoracic mass, with unclear aetiology and pathogenesis. Collaboration with radiology and pathology teams is essential for accurate diagnosis and effective management. UCD can mimic both benign and malignant lesions of the neck, thorax, and abdomen. A high index of clinical suspicion, followed by focused evaluation with multiphase CECT of the chest, can aid in establishing a preoperative diagnosis. Surgical resection of UCD may be challenging due to its hypervascularity and dense adhesions. Therefore, a presumptive preoperative diagnosis of CD may facilitate appropriate preoperative optimisation (e.g., embolisation) and inform the choice of surgical approach (open thoracotomy vs. VATS vs. the need for cardiopulmonary bypass).

Statements and Declarations

Preprint: This article was previously posted to the Research Square preprint server on February 15, 2022 [11].

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