Incidentally detected Castleman disease of the thorax and its surgical management

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Abstract

Castleman disease (CD) is a rare lymphoproliferative disorder, the etiology and pathogenesis of which is not clearly established. It presents more commonly as a localised disease or unicentric CD (UCD), and less often as generalised disease or multricentric CD (MCD). The most common site of UCD is in the thorax; however, UCD is rarely included in the differential diagnosis of an intrathoracic mass due to its rarity. The lesion is highly vascular and often has dense adhesions with adjacent organs, making the surgery a difficult task. We report a case of posterior mediastinal mass located in left paraspinal region which was detected incidentally in a 53-year old female and was subsequently resected successfully via left postero-lateral thoracotomy, and was diagnosed post-operatively on histopathological examination (HPE) as CD, hyaline vascular variant (HVV). We review the relevant clinical, pathological and radiological findings of CD, which may act as clinical pointers for establishing a preoperative diagnosis of CD. Suspecting CD preoperatively would guide the surgeon for appropriate surgical planning and may avoid facing such surprise on the operating table.

Title Page

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Abstract

Castleman disease (CD) is a rare lymphoproliferative disorder, the etiology and pathogenesis of which is not clearly established. It presents more commonly as a localised disease or unicentric CD (UCD), and less often as generalised disease or multricentric CD (MCD). The most common site of UCD is in the thorax; however, UCD is rarely included in the differential diagnosis of an intrathoracic mass due to its rarity. The lesion is highly vascular and often has dense adhesions with adjacent organs, making the surgery a difficult task.

We report a case of posterior mediastinal mass located in left paraspinal region which was detected incidentally in a 53-year old female and was subsequently resected successfully via left postero-lateral thoracotomy, and was diagnosed post-operatively on histopathological examination (HPE) as CD, hyaline vascular variant (HVV). We review the relevant clinical, pathological and radiological findings of CD, which may act as clinical pointers for establishing a preoperative diagnosis of CD. Suspecting CD preoperatively would guide the surgeon for appropriate surgical planning and may avoid facing such surprise on the operating table.

Keywords: Castleman disease, lymphoproliferative disorder, posterior mediastinal mass, incidental diagnosis

Text

Introduction

Castleman disease (CD) is a rare lymphoproliferative disorder of uncertain etiology, broadly classified as uncentric CD (UCD) and multicentric CD (MCD). Although thorax is the most common site of UCD, but overall CD a very rare cause of an intrathoracic mass and is usually overlooked as a differential diagnosis of an intrathoracic or mediastinal mass.[1]

Localised Castleman disease of the thorax is known to be quite hypervascular and in close proximity to adjacent organs or great vessels (pulmonary artery or aorta) and often densely adherent to these surrounding structures.[2,3] When overlooked, such tumours may pose surgical challenge (incomplete resection or profuse bleeding) and may lead to adverse outcome. Hence, identifying Castleman disease of thorax preoperatively and careful surgical planning may provide good outcome post-operatively.

We present a case of posterior mediastinal mass detected incidentally in a 53-year old female that was resected successfully via left posterior-lateral thoracotomy with a preliminary possibility of neurogenic tumour/ inflammatory myofibroblastic masss, but was finally diagnosed on histopathology of surgically resected mass as CD, hyaline vascular variant (HVV).

Case Report

Informed written consent was taken from the patient for the publication. A 53-year old female patient presented to us with a provisional diagnosis of hiatus hernia, which was detected incidentally on a CECT chest and whole abdomen while she was being evaluated for fibroid uterus. Outside, CECT chest and abdomen was reported as hiatus hernia with herniation of omental fat through widened hiatus with heterogeneous lobulated lesion within herniated omental fat; in addition there was significant left pleural effusion with passive atelectasis of the left lung. Patient was asymptomatic from hiatus hernia per se. As the diagnosis was in question, we re-evaluated the CT images with in-house radiologist. On re-evaluation, it appeared to be a large well-defined posterior mediastinal mass approximately 8x8 cm in dimension in left hemithorax in left paraspinal region, abutting on the descending thoracic aorta (DTA) with preserved fat planes (?liposarcoma) (Figure 1A, 1B). The mass showed intense enhancement on contrast imaging (Figure 1C). CT- guided biopsy of the mass was reported on histopathology as possibly inflammatory myofibroblastic tumour. No malignant cells were detected on diagnostic pleural aspiration.

After evaluating resectability of the mass, patient was taken up for excision via left posterolateral thoracotomy. Intra-operatively, a large lobulated mass, approximately 10x10x8 cm in dimension, firm in consistency occupying the paraspinal aspect of left hemithorax was detected. It was adherent to DTA with feeder vessels arising from DTA. The mass was densely adherent to the parieties and to left hemidiaphragm. On intraoperative assessment, it appeared to be a malignant tumour. The mass was gently dissected free from DTA after ligating feeder vessels, necessitating application of partial cross clamp for short duration to control bleeders from DTA. Mass was mobilised from left hemidiaphragm and parieties by combination of blunt and sharp dissection. Approximately 700 ml of straw coloured pleural fluid was aspirated. Left lung was completely normal and away from the mass.

Postoperative period was uneventful and patient was discharged in stable condition on 5th post operative day (POD). The cut section of the operative specimen showed well-defined tumour surrounded by fatty tissue on other end (Figure 2). On HPE of the excised mass, it showed characteristic onion-skin pattern of lymphoid follicles (concentric layer of lymphocytes surrounding an atrophic germinal centre) and was diagnosed as CD, hyaline vascular variant (Figure 3A, 3B).

Discussion

Castleman Disease (CD) is a rare non malignant lymphoproliferative disorder of uncertain etiology, first reported by Benjamin Franklin in 1954.[1] It is also known as giant lymph node (LN) hyperplasia or angiofollicular hyperplasia. Although it can present at any age, its peak incidence is in 3rd and 4th decade.[4]

It can be classified clinically (on the basis of location) into two types: Unicentric CD (localised disease) and multricentric CD (diffuse or systemic disease). Histopathologically, CD is of two types: hyaline vascular variant (HVV) and plasma cell variant (PCV). UCD is more common (68-98%) than MCD. Further, majority of UCD are HVV, whereas majority of MCD are PCV.[4,5]

UCD is usually an asymptomatic disease detected incidentally whereas MCD often presents with constitutional symptoms (fever, night sweats, weight loss etc). However, occasionally UCD may present with features of mass effect on adjacent on adjacent organs such as cough, dysphagia, dyspnoea or pain.[6]

Although CD can arise at any site where lymph node tissue is present, but its most common site is in the thorax (70%). Other sites are abdomen and pelvis (10-15%), neck (10-15%). In the thorax, most common site of CD is the mediastinum. Other intrathoracic sites of UCD are the hilum of lung, pleura, and chest wall.[7]

Identifying Castleman disease preoperatively is difficult. In the present case, a well-defined posterior mediastinal mass was incidentally detected on CECT chest in an asymptomatic 53-year old female which on preoperative biopsy on histopathology was considered as an inflammatory myofibroblastic tumour. Intraoperatively, the appearance was like that of a malignant tumour. Preoperative contrast imaging (CT/ MRI) may help to differentiate CD from other more common posterior mediastinal masses (neurogenic tumours e.g. schwannoma, neurofibroma, ganglioneuroma). On imaging, CD presents as a well-defined, homogeneous mass; however, the characteristic feature of UCD on CECT is *the intense enhancement during arterial phase* which decreases during portal venous phase. This intense enhancement is due to hypervascular nature of localised CD (UCD). Hence, *multiphase* CECT may be performed as the investigation of choice if UCD is suspected.[8] Further CT may reveal relation of mass to adjacent organs and major vessels of thorax, helping in surgical planning.

Identifying or suspecting UCD preoperatively may be useful for surgical planning. Localised/Unicentric CD (UCD) is amenable to complete surgical resection which is curative. However, UCD is known to be hypervascular and have dense adhesions with adjacent organs or major vessels[2] which may pose a surgical challenge. In such cases, either preoperative embolisation may be done (to reduce vascularity) [9] or mass may be excised on cardiopulmonary bypass (CPB) if dense adhesions with major vessels is present.[10] Video-assisted thoracoscopy (VATS) may be associated with less postoperative pain and less hospital stay; however, due to dense adhesions and vascular nature of CD, VATS does not appear to be a lucrative option in CD.[10]

Prognosis of UCD is good as it behaves as a benign disease and recurrence is rare; hence complete surgical resection is curative. On the other hand, MCD can present as an aggressive lymphoproliferative disease and debulking surgery and/or immunochemotherapy are treatment options in such cases.[7]

In conclusion, UCD is a rare cause of intrathoracic mass, the etiology and pathogenesis of which is unclear. It mimics both benign as well as malignant lesions in the neck, thorax and abdomen. A high index of clinical suspicion followed by focused evaluation with multiphase CECT chest can help to establish its diagnosis preoperatively. Resection of UCD may be difficult owing to hypervascularity and dense adhesions. Hence, preoperative presumptive diagnosis of CD may help in proper preoperative optimisation (embolisation), deciding surgical approach (open thoracotomy vs VATS vs need for CPB).

Author contributions

Dr. Sudhansoo Khanna - Data analysis, Drafting article.

Dr. Rana Sandip Singh - Critical revision of article.

Dr. Priyanka Goyal- Data Collection

Dr. Harsh Mohan – Critical revision of article.

Dr. Poonam Bhaker - Data collection.

Dr. Shaleen Rana - Data collection.

Dr. Sunil Sanga – Critical Revision of the article.

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Figure legends

Figure 1A: Non contrast CT (NCCT) chest (axial view) showing a well-defined hypodense soft tissue density mass (black arrows) 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 and left pleural effusion (blue star).

Figure 1B: NCCT chest (coronal reformatted view) shows a well defined, mass located above left hemidiaphragm. Rest of the findings are same.

Figure 1C : 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.

Figure 2 : Cut section of the operative specimen shows well-defined tumour surrounded by fatty tissue on other end.

Figure 3A : Photomicrograph shows lymphoid follicles (F) with atrophic germinal centres and central hyalinisation (yellow arrow head). Interfollicular stroma shows vascular and fibroblastic proliferation along with numerous lymphocytes and plasma cells.

Figure 3B : Photomicrograph (Inset image) shows lymphoid follicle having concentric layering of lymphocytes in an onion-skin pattern (yellow arrow) encircling an atrophic germinal centre (yellow arrow head). This so called targetoid onion-skin appearance and is highly suggestive of CD.