Indian Journal of Thoracic and Cardiovascular Surgery (November–December 2021) 37(6):691–693
https://doi.org/10.1007/s12055-021-01179-0

CASE REPORT

Mediastinal Castleman disease with supraclavicular extension—a diagnostic dilemma and surgical challenge

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Received: 28 January 2021 / Revised: 9 March 2021 / Accepted: 11 March 2021 / Published online: 26 May 2021

Abstract

Castleman's disease, also known as angiofollicular lymph node hyperplasia, is a rare lymphoproliferative disorder, with unicentric or multicentric variety. Herein, we present an unusual case of unicentric Castleman disease in a 19-year-old boy presenting as a left posterior mediastinal mass with supraclavicular extension, a rare form of presentation, with very few cases reported in the past. It can be misdiagnosed as other malignant pathology and mismanaged as in our case; hence, a high index of suspicion is necessary.

Keywords Castleman's disease · Unicentric · Mediastinal · Supraclavicular extension · Hyaline vascular

Introduction

Castleman's disease (CD) is an uncommon disorder characterized by a benign proliferation of the lymphoid tissue that may be localized or unicentric (UCD), or disseminated or multicentric (MCD). Histologically, CD can be classified as hyaline-vascular type, plasma cell type, or a mixed type [1]. Patients with localized hyaline-vascular type are usually asymptomatic and are diagnosed during routine imaging studies. The definitive diagnosis is based on postoperative pathological findings. The aim here was to describe a case of unicentric Castleman's disease, within the mediastinum and extending into the neck, its diagnostic tools, and the perioperative management.

Case report

A 19-year-old boy presented with prolonged hoarseness of voice and recurrent oral ulcers. On evaluation in his hometown, contrast-enhanced computed tomography (CECT) of the chest (Fig. 1) revealed a left hemithorax mass, which was interpreted as adenocarcinoma on biopsy. He received 3 cycles of gemcitabine and carboplatin. On persistence of symptoms, he came for a second opinion to the medical oncologist in our centre. On evaluation, significant findings were presence of clubbing, reduced breath sound intensity in the left supraclavicular area, with no palpable lymph nodes in the neck, bilateral axillae, or groin; serum tumour markers—alpha fetoprotein (AFP), beta human chorionic gonadotrophin (HCG), lactate dehydrogenase (LDH)—were normal. A fluorodeoxyglucose (FDG)-positron emission tomography (PET) (Fig. 1) revealed a mildly FDG-avid heterogeneously enhancing 9.2 × 9.0 × 10.0 cm mass arising from the left posterosuperior mediastinum, extending into the left supraclavicular region and no significant mediastinal lymph nodes. Given the doubtful primary diagnosis, an image-guided core biopsy performed at our centre revealed benign lymph node tissue, with no evidence of malignancy or granulomas. Immunohistochemistry was positive for CD20 (follicles), CD3 (interfollicular zone), and terminal deoxynucleotidyl transferase (TdT); negative for Pan CK. Videolaryngoscopy was normal but for mild bowing of the left vocal cord. Following workup for surgery, we resected the mediastinal mass via a left sternothoracotomy incision extending onto the neck along the sternocleidomastoid. It was 12 × 10 × 10 cm, well-circumscribed mass in the left posterosuperior mediastinum extending into the neck, in close proximity to the brachial plexus roots and left subclavian vessels (Fig. 2). Postoperatively, chylothorax was suspected on postoperative day 4 due to the nature and progressively increasing quantity of chest tube drainage, which was confirmed with elevated triglycerides in pleural

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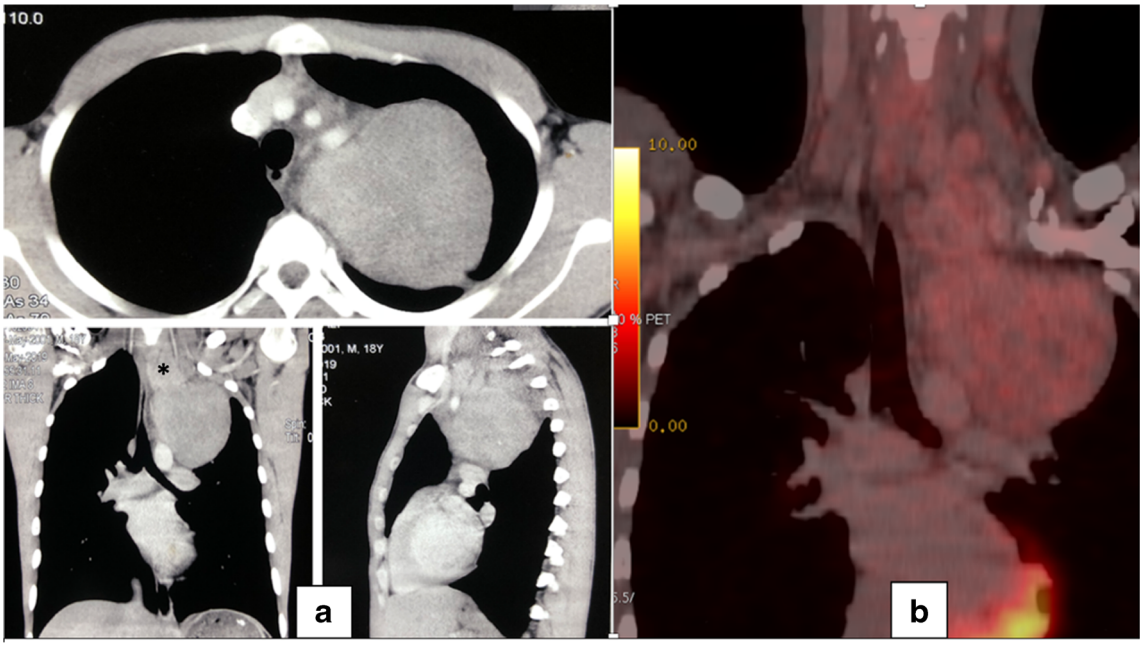

Fig. 1 **a** CECT chest depicting the mass in the left posterosuperior mediastinum with extension into neck ('asterix' seen in the coronal section in bottom left corner); **b** PET-CT revealed a heterogenously mild FDG uptake within the mass

fluid (> 800 mg/dl). He underwent uniportal video-assisted thoracoscopic surgery thoracic duct ligation, following which the drainage gradually subsided, and he was discharged on post-operative day 11. Histopathological examination showed preserved lymph node architecture with a capsule, widely spaced follicular germinal centres, and interfollicular vascular proliferation with perivascular hyalinization (Fig. 3). The features strongly favoured Castleman's disease of hyaline vascular type. The patient has been doing well with no recurrence at 1-year follow-up.

Discussion

Castleman's disease was first reported by Dr. Benjamin Castleman in 1956 as lymph node masses with capillary proliferation and follicular hyperplasia [2]. Based on the location, it has two clinical varieties: unicentric and multicentric Castleman's disease (UCD and MCD respectively). Histologically, it is classified into hyaline vascular, plasma cell, or mixed type [3]. Diagnosis is based on histopathology, as imaging features show considerable overlap, thus posing diagnostic difficulties as seen in our case. It can affect individuals from adolescence to the seventh decade of life. The hyaline vascular type is usually unicentric and asymptomatic, but occasionally may present with mass effect similar to our case. The multicentric variant is usually associated with plasma cell histomorphology and involves extra-thoracic sites, and affected individuals tend to have a more complicated course [4]. Most patients of unicentric variety are usually asymptomatic, and a mass is an incidental finding. Occasionally, they may present with chest heaviness, breathlessness, cough, and dysphagia. About 70% of UCD cases are seen in the thorax, 10–15% in the neck, and 10–15% in the abdomen, retroperitoneum, and pelvis [5]. Although occurrence of UCD in the mediastinum or neck is common, such lesions extending across both the compartments have been scarcely reported in the literature [6]. In our patient, it was seen as a posterior mediastinal mass extending into the neck. It may be misdiagnosed as neurogenic tumour due to its location [7]. The aetiology and pathophysiology have not been clearly elucidated. Surgery is usually curative, with favourable outcomes seen even in partially resected masses which remain asymptomatic for many years. Systemic steroids can provide symptomatic relief but do not predictably reduce tumour size. Radiation therapy shows response in few cases but makes subsequent surgical intervention more difficult [8].

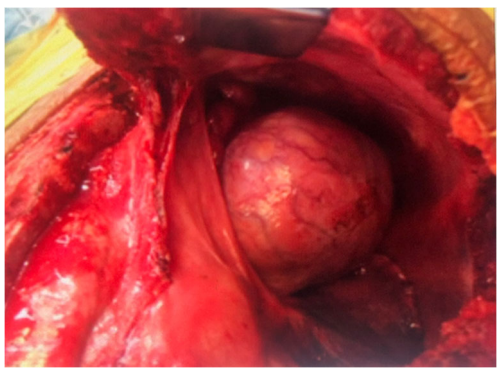

Fig. 2 Mass in the left posterosuperior mediastinum extending into the neck

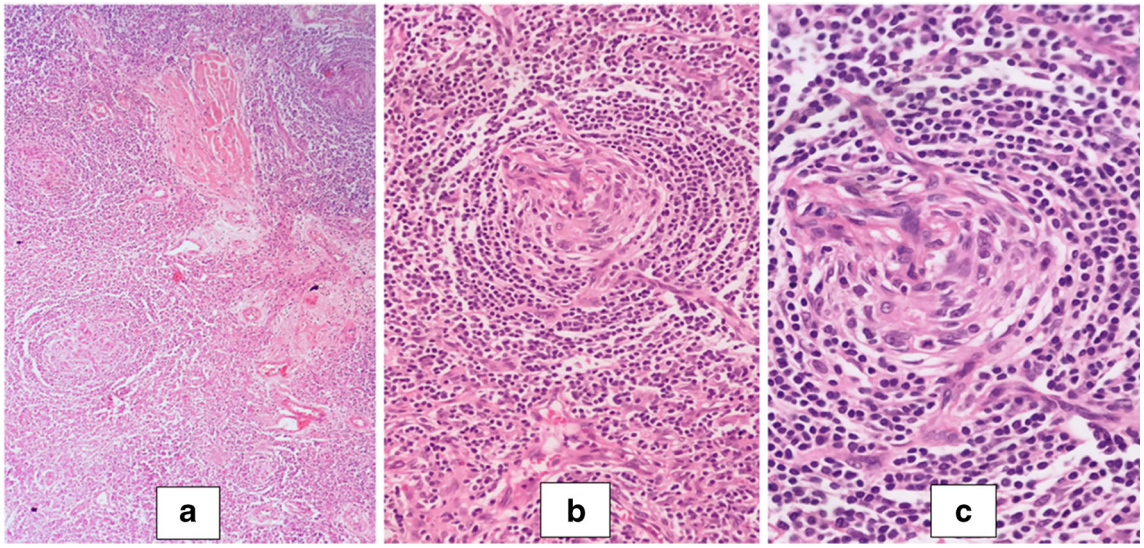

Fig. 3 Histopathological examination showed (**a**) preserved lymph node architecture with a capsule (**b**), widely spaced follicular germinal centres (**c**), and interfollicular vascular proliferation with perivascular hyalinization

Conclusion

Castleman disease is a relatively rare entity. The clinical and radiological features, such as those seen in our case, where the mass was crossing two different anatomical compartments can add to diagnostic challenge. Surgery is the mainstay of therapy and diagnosis is confirmed only with typical histopathological features; a high index of suspicion is mandatory. Chylothorax and neurovascular injuries are potential postoperative complications while dealing with such large masses. Overall prognosis is good, particularly in the unicentric variety of disease.

Author contribution All the authors have provided their intellectual contribution to the case report. The final draft was read and approved by each of them.

Funding None.

Declarations

Ethics approval The patient's personal information was anonymized and procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (Helsinki Declaration 1975, revised in 2000 and 2008). Accordingly, the Narayana health Academics Ethics Committee (NHAEC) approved this case report for publication prior to submission (letter no. NHH/AEC-CL-2020-597 dated 14/10/2020).

Consent for publication Informed patient consent for publication of this case report has been obtained.

Conflict of interest The authors declare no competing interests.

Meeting presentation Presented as Case Report in Indian Society of Thoracic Surgery Annual Meet January 2020.

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