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Case Report

A rare occurrence of unicentric Castleman disease

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ABSTRACT

Castleman disease (CD) is a rare lymphoproliferative disorder characterized by enlarged lymph nodes involving the neck, mediastinum, and retroperitoneum. Due to its varied presentations, it is often misdiagnosed or underdiagnosed. We present one such case which posed a great diagnostic challenge. An 18-year-old girl presented with fever on and off for 1 year. She was treated for COVID positivity during this time but had persistent evening rise of temperature even after 21 days of COVID negativity. Computed tomography chest done revealed a posterior mediastinal mass on the right side. Biopsy showed features of granulomatous inflammation. She was subsequently started on anti-tuberculous therapy. Repeat imaging showed no significant interval change in the mass and biopsy showed features of cavernous hemangioma. She underwent right posterolateral thoracotomy and excision of posterior mediastinal mass. Histopathology of the mass revealed it to be a hyaline variant of CD. CD is rare in children and the presentation can be very atypical and non-specific. They can be misdiagnosed as various other conditions. In most cases, it is an unanticipated pathological diagnosis.

Keywords: Unicentric Castleman disease, Mediastinal mass, Hyaline vascular variant

INTRODUCTION

A lymphoproliferative disorder, Castleman disease (CD), affects the lymph nodes of either neck, mediastinum, or sometimes the retroperitoneum.[1] Young adults are more commonly affected. Most common site involved is the mediastinum. Clinical presentation in CD can range from being completely asymptomatic to features mimicking autoimmunity. We report a case of an atypical approved presentation of CD which posed a challenge in diagnosis.

CASE REPORT

An 18-year-old girl came with complaints of on and off fever and headache for a duration of 1 year. She was diagnosed with COVID-19 pneumonia 2 days after the onset of fever. However, she continued to have low-grade fever even after testing negative 21 days later. An evening rise of temperature was noted with mild pain in the mid portion of the lower chest. There was no history of cough, difficulty in breathing, abdominal pain, vomiting, night sweats, or significant weight loss. A family member had tuberculosis 10 years ago and was fully treated. General and systemic examinations were unremarkable. With this clinical presentation, the patient was planned for further workup with imaging and blood tests. Blood investigations were unremarkable. High-resolution computed tomography (CT) thorax was done which showed a large lobulated posterior mediastinal soft-tissue lesion measuring 7.3 × 10 ×1.3 cm with central necrotic area abutting the esophagus medially left atrium anteriorly with maintained fat planes. A radiological differential diagnosis of a neurogenic tumor was given. CT-guided biopsy from the mass showed features of chronic granulomatous inflammation. The patient was started on ATT following the biopsy. A follow-up CT chest was done which showed no change in the size of the mass with contrast enhancement raising the possibility of mediastinal paraganglioma. Higher investigation in the form of magnetic resonance imaging thorax was done which showed a large well defined T2 hyperintense lesion in the right paravertebral and prevertebral region [Figure 1]. On contrast administration, the lesion was seen exhibiting progressive centripetal enhancement with central non-enhancing area. Anteriorly, the mass was abutting the pericardium and left atrium. Superiorly, the mass extended up to the arch of aorta, inferiorly till the diaphragm. Medially, the lesion encased one-fourth of circumference of aorta. Arterial feeders were from the descending thoracic aorta [Figure 2]. This raised a possibility of benign vascular neoplasm, mostly a hemangioma. A repeat biopsy of the mass gave a pathological diagnosis of a cavernous hemangioma.

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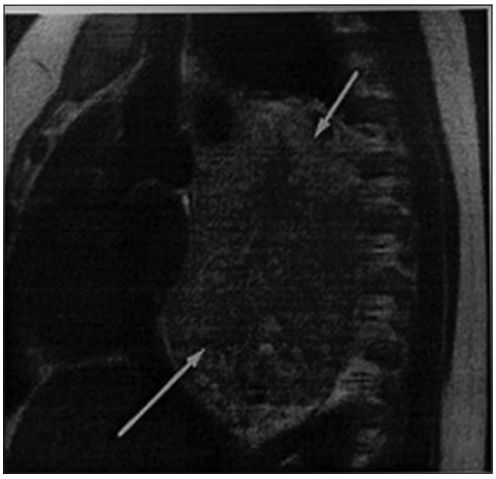

Figure 1: T2 hyperintense mass lesion in the right paravertebral region (indicated by arrow).

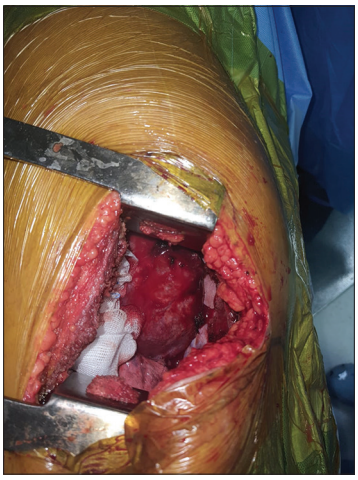

Figure 3: Intraoperative view of the mass.

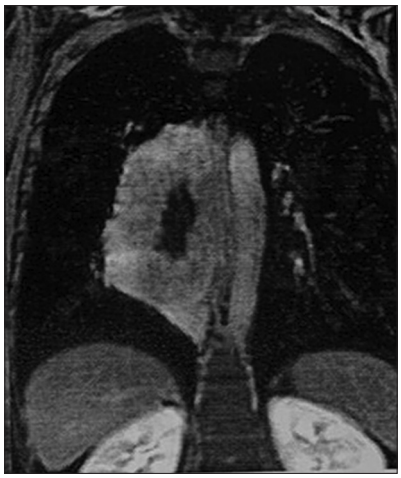

Figure 2: Mass showing a progressive centripetal enhancement with central non-enhancing area.

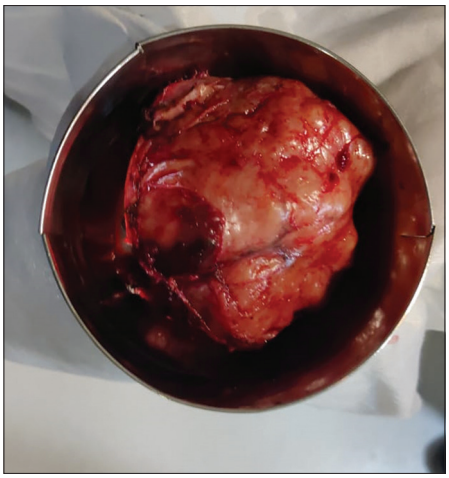

Figure 4: Excised mass.

With the initial biopsy showing features of chronic granulomatous inflammation, the patient was started on anti- tuberculous therapy. However, there was no improvement clinically and radiologically. Later, with the pathological diagnosis of cavernous hemangioma, she was started on propranolol therapy. Medical treatment showed no improvement and with persisting symptoms, the patient underwent right posterolateral thoracotomy with excision of the mediastinal mass. Intraoperative findings showed a soft to firm mass of size 12 × 12.5 cm in the right posterior mediastinum adherent to the lower lobe of the right lung, descending aorta, inferior vena cava, diaphragm, and esophagus and was excised in toto [Figures 3 and 4]. Intraoperative blood loss was managed with transfusions. Postoperatively, the patient was managed in the intensive care unit with inotropic supports and gradually weaned off. Intercostal drains were removed on post-operative day 4. The patient was discharged on post-operative day 7 in a hemodynamically stable condition. The histopathology of the excised mass showed angiofollicular lymphoid hyperplasia with extensive hyalinization, sclerosis, and multifocal granulomas suggestive of hyaline vascular (HV) variant of CD and unicentric type.

DISCUSSION

CD includes a heterogeneous group of disorders involving lymph nodes. Histopathologically, it is classified into unicentric CD (UCD) and multi-centric CD (MCD) types. UCD affects a single enlarged lymph node or multiple nodes of a single nodal group. It was first described by Castleman and Towne in a patient with a mediastinal mass in 1954 and thereafter by Castleman. in a series in 1956.[2] MCD affects multiple nodal stations along with systemic involvement. Symptoms such as fever, weight loss, and fatigue occur due to interleukin-6 and other cytokines.[2]

Castleman who originally reported the disease found that the disease was not gender specific. However, recent studies have shown that there is a slight female preponderance. Incidence is estimated to be around 16 in 1,000,000. UCD exhibits as asymptomatic lymphadenopathy. If symptoms do occur, they are often due to compression of vital structures due to the enlarged lymph nodal mass. In a series by Talat *et al*., the most common sites of involvement were the mediastinum (29%), neck (23%), abdomen (21%), and retroperitoneum (17%). UCD can also occur in the axilla and inguinal regions as well as orbits, nasopharynx, and small bowel.[3]

Unicentric type of CD when asymptomatic in most instances is almost always underdiagnosed or misdiagnosed. CD of the thorax can occur in the mediastinum or in other areas in the thorax such as chest wall, pleura, and lung. Symptoms such as chest pain, breathlessness, and cough may occur due to compression from the mass. Challenges to complete resection of the mass include adherence to blood vessels and bronchi.[4]

The pathogenesis of UCD is not clear. Recent evidence suggests that it may be caused by a clonal expansion of lymph node stromal cells and must be distinguished from other lymphoproliferative disorders common in children. These disorders include infectious and autoimmune disorders such as infectious mononucleosis and autoimmune lymphoproliferative syndrome and others such as Kawasaki disease and Rosie-Dorfman disease.[5] The vague symptomatic profile poses a great challenge in the diagnosis and can lead to various investigations and biopsies which still could be inconclusive.

Histopathological variants of CD are (1) HV variant; (2) plasma cell variant (PCV); (3) mixed variant; and (4) plasmablastic variant.[6] HIV and HHV-8 are commonly associated with MCD CD. The plasmablasts harboring HHV-8 in histopathologic evaluation predict the severity and outcome of the disease. MCD, PCV, and HHV-8 positivity indicate poor prognosis.[7]

There is no recognized treatment guideline for CD. However, surgery and complete resection remain the mainstay of treatment of UCD.[5] Complete surgical resection is generally considered curative in unicentric disease. MCD disease, however, may require chemotherapy and radiotherapy in addition to surgery as part of the treatment. Incomplete resection in UCD might require additional chemoradiotherapy. Baek *et al*. reported a case of an adolescent with relapsed unicentric-PCV CD after incomplete resection who was eventually cured with complete surgical resection.[7]

It is essential that the diagnosis of CD be made based on clinical presentation and radiological investigation before surgery. The complete removal of the lesion with appropriate chemotherapy provides excellent prognosis in children with CD.[5]

CONCLUSION

We report a case of mediastinal UCD with HV variant in an adolescent girl who presented with on and off fever and non-specific chest pain. All attempts of definitive pre-operative diagnosis failed in our case. Complete surgical resection was successful in our patient in providing a cure. CD should always be kept as a differential diagnosis in mind when evaluating a mass with non-specific symptoms.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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