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RESEARCH ARTICLE

CASTELMAN DISEASE REVEALED BY A GIANT ABDOMINAL MASS: A RARE CASE

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Abstract

Introduction:-

Castleman's disease (CD) was first described as hyperplasic mediastinallymph nodes containing interfollicular vascular proliferation in 1956 by Benjamin Castleman etal.[1] CD is a lymphoproliferative disorder and comprises two distinct diseases with differentprognoses, including unicentric and multicentric. Unicentric CD (UCD) is defined as a localizeddisease which involves enlarged lymph node(s) in a single region of the body, whereas multicentricCD (MCD) is a systemic disease with generalized peripheral lymphadenopathy,hepatosplenomegaly, frequent fevers, and night sweats. UCD isa rare disease and its true incidence is not known but it can be seen at any age, especially in youngeradults. The median age at presentation is approximately 35 years.[2] The pathogenesis of UCD isnot well understood, and viral, autoimmune, and neoplastic diseases have all been proposed aspossible etiologies.[3] Unicentric Castlemandisease (UCD) is usually asymptomatic with a 95.3% survival rate. UCD usually affects just one lymph node or a singlelymph node region. While the disease may involve all parts of the body, the mediastinum appears tobe the most common part of involvement. In this study, we present one case of CD causingmechanical intestinal obstruction due to involvement of first ileal loop.

Case Report

A 62-year-old man was admitted to the hospital with abdominal pain, nauseous,fecaloid vomiting, abdominal distention, and absence of gas and stool discharge since 5 days. Hislast bowel movement was 5 days ago, with episodes of chronic constipation. Heshowed no signs of fever or chills. He had no past history of previous hospitalizations. Onphysical examination, he showed signs of diffuse abdominal distention and an abdominal mass was detected. Respiratory soundswere normal. The patient's blood pressure was 130/80 mmHg, heart rate was

76 beats per minute,SpO2 was 96%, and temperature was 37.8 °C.CT scan of the abdomen was done, and showed a mass of ileum measuring 15 cm. The patient was admitted to the block and ileal resection including the mass, was performed.

Gross examination find a hard whitish mass, measuring 15x 12 cm, attached to the ileum.(Fig 1)

Pathological evaluation showed a lymph node with reactive lymphoid hyperplasiacharacterized by regressively transformed germinal centers with increased interfollicularvascularity, consistent with the hyaline vascular variant of

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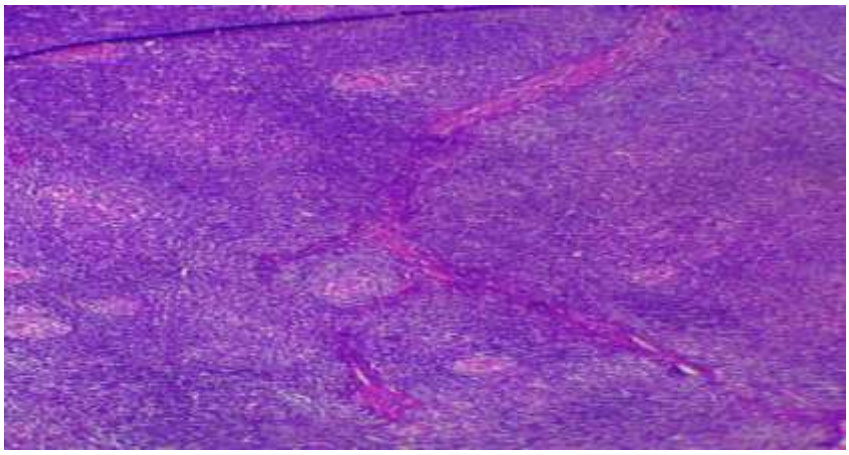
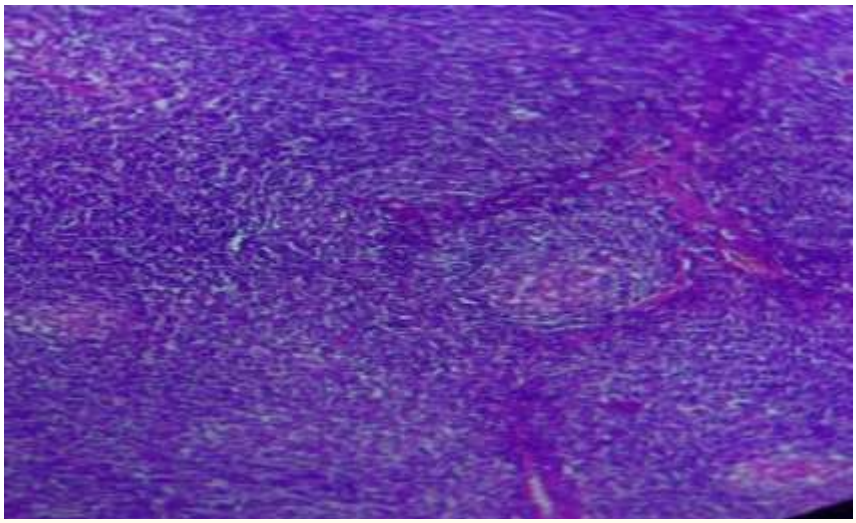
Castleman disease(Fig. 2-3). The postoperative course was uneventful. Onpostoperative day 3, oral food intake was started. The patient was discharged on postoperative day 8 without any problems.

Figure 1:- Gross image of the mass.





Figures 2- 3:- Pathology of the mass showing small follicles with a partially lymphocyte-depleted germinal center with hyaline deposits. Capillaries and small vessels are present in the germinal center within the interfollicular space, showing the unique lollipop configuration.

Discussion:-

CD, which was reported by Castleman in 1954, is a highly heterogeneous clinicopathological entity belonging to the family of lymphoproliferative disorders.[6] Although the etiology and pathophysiology of CD's remain unknown, potential contributors to disease development are chronic inflammation, immune deficiencies, and some autoimmune diseases. Additionally, Epstein-Barr virus, Toxoplasma, and Mycobacterium tuberculosis are among the infectious agents that are responsible for disease. Clinical and laboratory abnormalities noted in the presence of CD are associated with inflammatory mediators, particularly with interleukin-6.[7,8] CD is clinically classified into two types: unicentric and multicentric. UCD (90%) is the most commonly seen type. There are three subtype of UCD: The hyaline vascular histopathologic subtype, the plasma cell histopathologic subtype and the mixed histopathologic subtype. Multicentric type is associated with a poor prognosis. A patient diagnosed with multicentric CD should be considered as systemic, and combination treatment, should be initiated as soon as possible.[9] Patients with UCD are reported to be commonly asymptomatic and come to clinical attention when an enlarged lymph node is noted on physical examination or cause a problem. In the present study, the problem was mechanical intestinal obstruction due to enlargement of mesenteric lymph nodes and involvement of terminal ileal wall.[2] Although the radiological appearance of CD is nonspecific, UCD may also present on imaging modalities. The most common radiologic presentation is mediastinal or hilar mass on computed tomography. Our patient had localized CD, and presented with clinical signs and symptoms of mechanical intestinal obstruction. Completeresection of the involved node or organ is curative and the gold standard approach for the treatmentof UCDTalat et al. in their systematic review, investigated 404 published cases of CD, and concluded that surgery is the gold standard for treatment of UCD. A systematic review of the role of surgical resection in localized CD showed that surgical resection was the most effective treatment for localized CD. However, aggressive surgical treatment of CD is not recommended as this may increase the rates of morbidity and mortality among CD patients. CD is a benign disease, which may also be treated by chemoradiotherapy as an alternative to surgery or after surgery.

Particularly, in the presence of unresectable unicentric CD, neoadjuvant rituximab and neoadjuvant radiotherapy can allow resection to be performed with a lower rate of morbidity given that these treatments may result in tumor shrinkage and reduced vascularity. Total resection provided cure in our case presented here. While mesenteric involvement in our case was unicentric, histopathological investigations indicated hyaline-vascular type CD. CD is difficult to diagnose preoperatively. The disease often presents with a solitary mass. Enlarged solitary lymph node showing homogeneous intense enhancement upon administration of contrast agent in computerized tomography should remind the diagnosis of the CD. The disease is frequently confused with malignancy as unicentric CD does not have specific radiological findings and appears as a solitary mass on radiological images.

Conclusion:-

CD should be kept in mind during the differentialdiagnosis of mechanical intestinal obstruction provided that wall thickening in terminal ileummimicking mass, and accompanying enlargement mesenteric lymph nodes observed duringpreoperative investigations or intraoperative exploration. In cases of UCD, complete surgicalresection should be performed.

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