CASE REPORT

Castleman's disease presenting with mechanical intestinal obstruction: A rare case

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ABSTRACT

Castleman's disease (CD) is a lymphoproliferative disorder and the occurrence of CD in the small bowel is rare. In this study, we present one case of CD causing mechanical intestinal obstruction due to involvement of terminal ileum. A 50-year-old man was admitted to the hospital with signs and symptoms of mechanical intestinal obstruction without history previous surgery. After examination and obtaining abdominal computed tomography, diagnosis of mechanical intestinal obstruction was reached and emergency surgery was performed with a median incision. On abdominal exploration a tumor like mass that also held distal small intestine mesentery, and ileocecal valve causing complet intestinal obstruction was observed. Ileocecal resection and ileocolonic anastomosis were performed. CD is a rare entity and should be kept in mind during the differential diagnosis of mechanical intestinal obstruction provided that wall thickening in terminal ileum mimicking mass, and accompanying enlargement mesenteric lymph nodes observed during preoperative investigations or intraoperative exploration.

Keywords: Castleman's disease; lymphoproliferative disorder; mechanical intestinal obstruction; mesenteric lymph nodes; terminal ileum.

INTRODUCTION

Castleman's disease (CD) was first described as hyperplastic mediastinal lymph nodes containing interfollicular vascular proliferation in 1956 by Benjamin Castleman et al.[1] CD is a lymphoproliferative disorder and comprises two distinct diseases with different prognoses, including unicentric and multicentric. Unicentric CD (UCD) is defined as a localized disease which involves enlarged lymph node(s) in a single region of the body, whereas multicentric CD (MCD) is a systemic disease with generalized peripheral lymphadenopathy, hepatosplenomegaly, frequent fevers, and night sweats. CD may also present with a number of malignancies, including Kaposi sarcoma, Hodgkin lymphoma and non-Hodgkin lymphoma. UCD is a rare disease and its true incidence is not known but it can be seen at any age, especially in younger adults. The median age at presentation is approximately 35 years.[2] The pathogenesis of UCD is not well understood, and viral, autoimmune, and neoplastic diseases have all been proposed as possible etiologies.[3] Although some familial cases of UCD have been reported, genomic sequencing has not been performed to identify inherited mutations.[4,5] Patients with UCD are usually asymptomatic and come to clinical attention when an enlarged lymph node is noted on physical examination or imaging studies. UCD usually affects just one lymph node or a single lymph node region. While the disease may involve all parts of the body, the mediastinum appears to be the most common part of involvement. In this study, we present one case of CD causing mechanical intestinal obstruction due to involvement of terminal ileum.

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CASE REPORT

A 50-year-old man was admitted to the hospital with abdominal pain, nauseous, fecaloid vomiting, abdominal distention, and absent of gas and stool discharge since three days. His last bowel movement was three days ago and he reported no flatulance for three days, as well. He showed no signs of fever or chills. He had no a past history of previous hospitalizations. On physical examination, he showed signs of abdominal distension and bowel sounds were hypoactive. Tenderness was observed in all quadrant and there was rebound and guarding. Respiratory sounds were normal. The patient's blood pressure was 140/90 mmHg, heart rate was 90 beats per minute, SpO_2 was 92%, and temperature was 37.6 °C. Rutin blood tests and abdominal computed tomography (CT) were obtained. Results of blood tests revealed total protein 7.3 g/dL, albumin 4.5 g/dL, AST 10 IU/L, ALT 9 IU/L, glucose 165 mg/dL, CRP 7.6 mg/dL, WBC 16.570 /µL, neutrophils 72%, HGB 13.74%, HCT 44.17%, PLT 328.000 /µL, INR 1.43, total bilirubin 0.7 mg/dL, BUN 21 mg/dL, creatinin 1.65 mg/dL, Na 141 mEq/L, K 4.3 mEq/L. Abdominal CT images and findings were shown in Figure 1.

Because of the severe tenderness, signs of peritoneal irritation, and CT findings compatible with mechanical intestinal obstruction, emergency surgery was performed the same day with a median incision. On abdominal exploration, a tumor-like mass that also held distal small intestine mesentery, and ileocecal valve causing complet intestinal obstruction was observed. Ileocecal resection and ileocolonic anastomosis was performed. Pathological findings revealed Castleman hyaline vascular type reactive lymphoid hyperplasia with ulcerated active chronic inflammation (Fig. 2).

The postoperative course was uneventful. On postoperative day 3, oral food intake was started. The patient was discharged on postoperative day 7 without any problems.

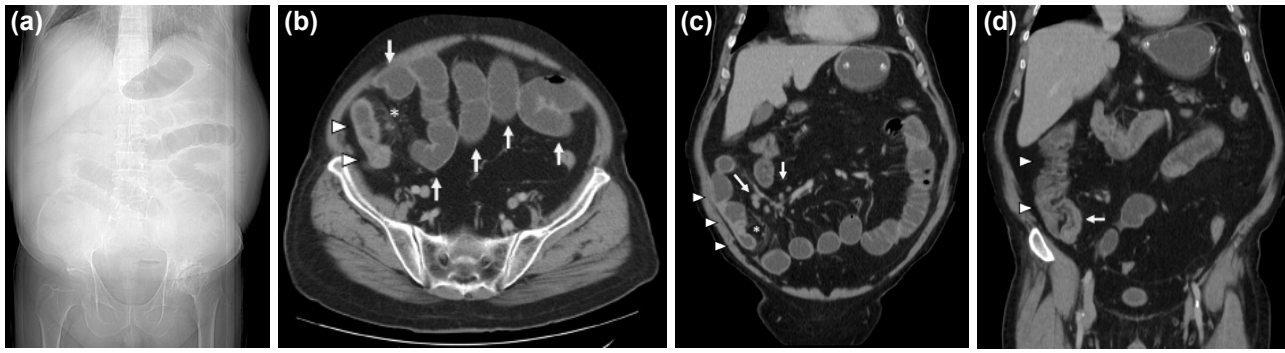

Figure 1. Abdomen CT scout image of patient in supine position **(a)** demonstrates distended stomach and small intestine segments with air, and diameter of the small intestine segments increased as well, compatible with intestinal obstruction. Axial contrast enhanced abdominal CT **(b)** demonstrates dilated small intestine segments (arrows), confirms scout image findings. Diffuse bowel wall thickening and contrast enhancement in the thickened bowel wall (arrowheads) with increased density adjacent mesentery (*) are seen in right lower quadrant, just before the terminal ileum (not shown in image). In coronal reformatted image **(c)**, bowel wall thickening, long segmental involvement (arrowheads), and enlarged lymph nodes (arrows) in adjacent mesentery with increased density (*) are seen at right lower quadrant. In addition, involvement of terminal ileum (arrow) and lack of cecal and ascending colon involvement (arrowheads) are also seen in this coronal reformatted image **(d)**.

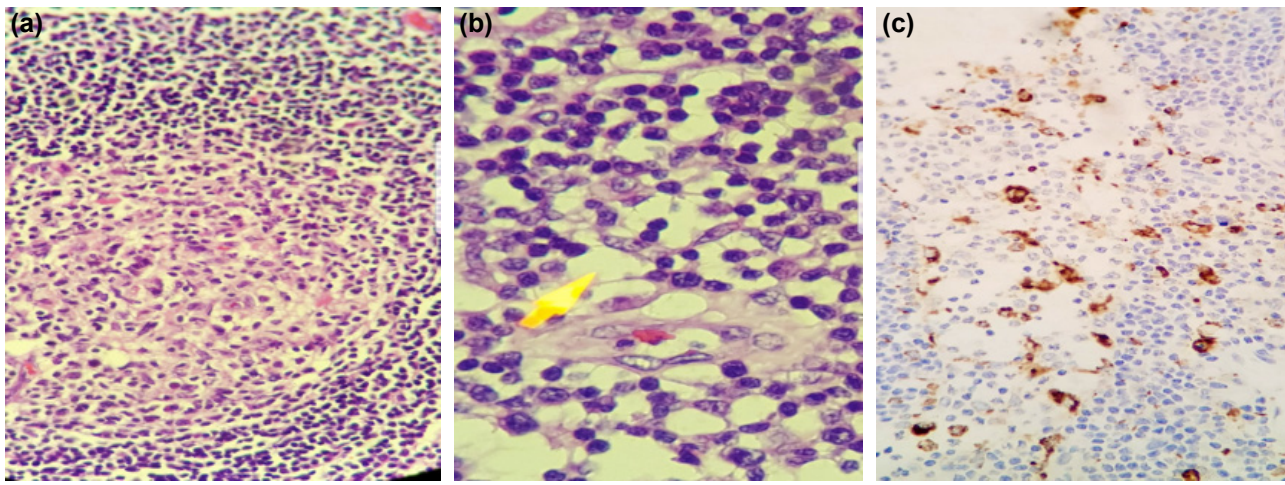

Figure 2. Castleman disease of hyalin vascular type. Germinal centers are rather prominent and they Show well-developed changes **(a)**. Scattered reticular/dendritic cells (arrow) some show dysplastic changes. Hematoxylin-Eosin stain. 100× **(b)**. CD68 positive plasmacytoid dendritic cells in the interfollicular stroma. CD68 (514H12) Leica BOND-III mouse anti-human monoklonal antibody **(c)**.

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.[10] Our patient had localized CD, and presented with clinical signs and symptoms of mechanical intestinal obstruction. After surgical treatment, the patient was followed clinically and did not have a problem.

Complete resection of the involved node or organ is curative and the gold standard approach for the treatment of UCD.[11] Talat 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.[12] 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.[13] 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 differential diagnosis of mechanical intestinal obstruction provided that wall thickening in terminal ileum mimicking mass, and accompanying enlargement mesenteric lymph nodes observed during preoperative investigations or intraoperative exploration. In cases of UCD, complete surgical resection should be performed.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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OLGU SUNUMU - *ÖZET*

Mekanik bağırsak tıkanmasıyla başvuran Castleman hastalığı: Nadir bir olgu

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Castleman hastalığı, lenfoproliferatif bir hastalıktır ve ince bağırsakta görülmesi nadirdir. Bu çalışmada, terminal ileum tutulumuna bağlı mekanik bağırsak tıkanıklığına neden olan bir Castleman hastalığı olgusu sunuldu. Daha önce geçirilmiş karın cerrahisi öyküsü olmayan 50 yaşında erkek hasta, mekanik bağırsak tıkanıklığı bulguları ve semptomları ile hastaneye başvurdu. Muayene ve abdominal bilgisayarlı tomografi ile mekanik intestinal obstrüksiyon tanısı konuldu ve median insizyon ile acil cerrahi girişim yapıldı. Abdominal eksplorasyonda, distal ince bağırsak mezenterini ve ileoçekal valvi tutan tümör benzeri kitlesel lezyonun intestinal obstrüksiyona neden olduğu görüldü. İleoçekal rezeksiyon ve ileokolik anastomoz yapıldı. Castleman hastalığı nadir görülen bir durumdur. Görüntülemede ya da ameliyat sırasında büyümüş lenf bezlerine eşlik eden distal ileumda kitle benzeri görünüm tespit edilmesi durumunda mekanik bağırsak tıkanıklığının ayırıcı tanısında akılda bulundurulmalıdır.

Anahtar sözcükler: Castleman hastalığı; lenfoproliferatif bozukluk; mekanik intestinal obstrüksiyon; mezenterik lenf nodları; terminal ileum.

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