

Castleman disease presenting as obstructive jaundice

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ABSTRACT

A 48-year-old Saudi male was admitted with features of obstructive jaundice. Endoscopic retrograde cholangiopancreatography showed stricture in distal common bile duct (CBD). Computed tomography scan revealed lymphadenopathy along CBD and in porta hepatis. Cholangiocarcinoma, lymphoma or metastatic deposits were suspected but no definite preoperative diagnosis could be established. Laparotomy disclosed lymph node enlargement in porta hepatis and along the CBD and lesser curvature of the stomach. Triple bypass procedure was performed to relieve the obstruction. Pathological examination of the lymph nodes showed Castleman disease of hyaline vascular type.

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Castleman disease (CD), first described in 1954, is a rare lymphoproliferative disorder of unknown etiology.¹ Most cases of CD involve mediastinum but may occur anywhere where lymph node tissue is found.² We report a case of Castleman disease presenting as obstructive jaundice and distal common bile duct (CBD) stricture with multiple locoregional lymph nodes enlargement. To the best of our knowledge, only one case of CD causing obstructive jaundice has been previously reported in the English literature.³ This case is reported to highlight this rare clinical presentation of CD and to review the literature for the current clinicopathological spectrum and management options of this uncommon clinical entity. Castleman disease should be included in the list of long differential diagnoses of obstructive jaundice due to regional lymphadenopathy.

Case Report. A 48-year-old Saudi male was admitted through the emergency room to the surgical unit of Riyadh Medical Complex, Riyadh,

Kingdom of Saudi Arabia with mild to moderate continuous pain in the right upper abdominal quadrant for one month. The severity of pain had increased over the last 2 days and he had developed fever in the last 24 hours. He had some degree of mental retardation since his childhood. Positive findings on physical examination were fever (38.3°C), jaundice and right upper abdominal quadrant tenderness. Laboratory data revealed hemoglobin of 10.3 g/dl, leucocytosis (13.8 x 10⁹/L) and biochemical features of obstructive jaundice (total bilirubin 23 mmol/L, direct bilirubin 20 mmol/L, alkaline phosphatase 354 U/L). Other liver enzymes and amylase were within normal range. Treatment of cholangitis was instituted and the patient was further investigated. Ultrasonographic examination showed gallstones, CBD dilatation (11 mm) and a soft tissue growth around the distal end of CBD. Endoscopic retrograde cholangiopancreatography (ERCP) confirmed stricture of distal CBD with proximal dilatation and dilatation of the pancreatic duct. Papilla of Vater

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was seen bulging with normal looking overlying mucosa. Cholangiocarcinoma was suspected but report of brush cytology did not support it. Computed tomography scan demonstrated enlarged lymph nodes in porta hepatis and along the CBD suggesting lymphoma or metastatic deposits. At laparotomy, lymph nodes enlargement was noted in the porta hepatis, alongside CBD and the lesser curvature of the stomach. Common bile duct was dilated (12 mm). Gallbladder was thick-walled, shrunken and contained multiple stones. Frozen section of pericholedochal lymph node was reported as "reactive changes" only. Triple bypass, comprising Roux-en-Y choledochojejunostomy, retrocolic gastro jejunostomy and enteroenterostomy, was performed after cholecystectomy and multiple biopsies of involved lymph nodes along CBD, porta hepatis and gastric lesser curvature were obtained. The lymph nodes were firm and the cut surface was homogeneously gray. Paraffin histological examination demonstrated follicular hyperplasia with increased amount of hyaline material within lymphoid follicles and the interfollicular tissue (Figure 1). The findings were consistent with hyaline vascular type of angiofollicular lymph node hyperplasia or Castleman disease. Postoperative course of the patient was unremarkable. His biochemical parameters returned to normal. The patient is symptom free after a follow up of 9 months.

Discussion. Castleman disease is a benign disease of unknown etiology. It is characterized by proliferation of mature lymphocytes and plasma cells. Castleman and Towne¹ first reported hyperplasia of mediastinal lymph nodes and described these as resembling those of thymoma.⁴ Although exact pathogenesis of CD is unknown; chronic low-grade inflammation, hamartomatous origin, an immunodeficiency state and autoimmunity have all been implicated as possible etiologies.⁵ The most common location of CD is mediastinum. However, extrathoracic sites have been reported in the neck, axilla, shoulder, pelvis, pancreas, retroperitoneum and mesentery.^{3,5} Castleman disease localized to lymph nodes in porta hepatis and along the CBD resulting in obstructive jaundice is exceedingly rare.³ There are 2 histological types of CD: the hyaline vascular type (90%) and plasma cell type (10%). The hyaline vascular type is characterized by small hyaline follicles and extensive capillary proliferation, whereas, plasma cell type has sheets of proliferating plasma cells without vascularization or hyalinization.⁶ Clinically, hyaline vascular type is usually asymptomatic, whereas, patients with plasma cell type are likely to have symptoms of

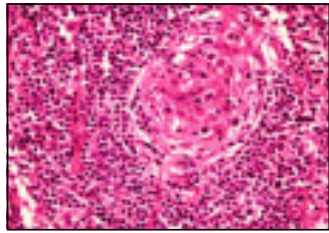


Figure 1 - Photomicrograph showing flattened cells making concentric ring pattern, hyalinized germinal center and capillary proliferation in hyaline vascular type of Castleman disease.

anemia, fever, sweating and fatigue.⁷ Other presentations may be related to the site of origin, like by virtue of its location, CD in the present report presented with features of obstructive jaundice. Preoperative diagnosis of CD is usually difficult to establish. Cytology and core histological biopsy may misdiagnose it as lymphoma.^{5,8} The lesion usually appears as an enhancing mass on CT and should be considered in the differential diagnosis of such radiological lesions.⁹ Castleman disease is commonly a well localized disease process.⁹ The solitary form usually does not progress and surgical resection is curative. Various treatment options for rare multicentric form includes surgery, chemotherapy, radiotherapy and steroids with variable results.¹⁰

Castleman disease is an uncommon disease of obscure etiology. Castleman disease of the lymph nodes around the biliary tree is extremely rare and may present as obstructive jaundice. The disease should be considered in cases of obstructive jaundice due to extrinsic compression by multiple local and regional lymph nodes enlargement of uncertain origin.

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