Hilar Inflammatory Pseudotumour with Hepatic Artery Atheroma- mimicker of Klatskin Tumour

Archana Rastogi · Chhagan Bihari · Nalini Gupta · Pranjal Deka ·
Arvind Kumar · Sanjay Singh Negi · Ankur Arora

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Abstract Inflammatory pseudotumour of hilar biliary structures is an extremely rare benign lesion that can mimic hilar cholangiocarcinoma. Clinical presentation and imaging findings often pose diagnostic difficulties. Main histopathological findings are the presence of myofibroblastic spindle cells, plasma cells, macrophages, and lymphocytes without cellular atypia or atypical mitotic figures. We describe a case of 62 year old male who presented with surgical obstructive jaundice. Imaging revealed a mass lesion involving the biliary confluence with upstream dilatation of biliary tree. Diagnosis of hilar cholangiocarcinoma with type III hilar block was made. Intraoperatively hilar mass lesion was found which was encasing right hepatic artery with no evidence of metastasis. The patient underwent Right hepatectomy with caudate lobectomy with complete common bile duct (CBD) excision with Roux en Y hepaticojejunostomy. Unexpectedly histopathological examination showed no evidence of malignancy and revealed hilar inflammatory pseudotumour with hepatic artery atherosclerosis. Preoperative imaging, operative management, pathologic diagnosis and literature review are being presented in view of rarity of the case.

Keywords Hilar · Inflammatory pseudotumour · Cholangiocarcinoma · Artery · Atheroma

A. Rastogi (✉) · C. Bihari · N. Gupta
Department of Pathology, Institute of Liver and Biliary Sciences,
D-1 Vasant Kunj, New Delhi 110070, India
e-mail: drarchanarastogi@gmail.com

P. Deka · A. Kumar · S. S. Negi
Department of Hepatopancreatobiliary Surgery, Institute of Liver
and Biliary Sciences, D-1, Vasant Kunj, New Delhi 110070, India

A. Arora
Department of Radiology, Institute of Liver and Biliary Sciences,
D-1, Vasant Kunj, New Delhi 110070, India

Introduction

Inflammatory pseudotumor (IPT), also known as inflammatory myofibroblastic tumors or plasma cells granuloma, is benign, rare and poorly understood entity and is an extremely rare cause of a hilar mass lesion.

Resection and in selected patients, transplantation offers the best chance of cure in hilar cholangiocarcinoma, also known as Altemeier-Klatskin tumour. The clinical presentation and radiological appearance of hilar IPT are similar to those of a Klatskin tumour; hence preoperative diagnosis of the tumor is difficult and often depends on pathological examination after operation.[1].

Biliary inflammatory pseudotumour (IPT) located at porta hepatis are typically diagnosed after aggressive surgical intervention and usually present with jaundice, rarely with pain, fever and anemia. We report a case of IPT located at the porta hepatis, mimicking Klatskin tumour and associated with peculiar findings of coexistent hepatic artery atherosclerosis. The Surgical result and post-operative follow-up was good.

Case Report

A 62 year old gentleman presented with surgical obstructive jaundice. He was apparently well 20 days back when he noticed yellowish discoloration of eyes. It was gradually progressive in nature and was associated with pruritus, clay colored stools and high colored urine. There was no history of pain, fever, vomiting, upper or lower gastrointestinal bleed or altered bowel habits. No history of prodromal symptoms. There was no history of loss of weight or similar episodes in the past. He was not a known case of diabetes, hypertension, COPD asthma, tuberculosis and there was no history of previous surgery. No history of similar illness in the family. He was evaluated in a private hospital with USG abdomen and

Fig. 1 (A) Coronal (3D) MRCP image shows a hilar block (white arrow) with separation of the right and left hepatic ducts and resultant bilobar biliary dilatation. (B) Axial contrast enhanced CT image reveals a soft-tissue mass at the hepatic hilum (black arrow)

referred to our institute for further management. He was moderately built and nourished, conscious and cooperative. Icterus was present. There was no pallor, cyanosis, clubbing or edema. There was no supraclavicular lymphadenopathy. Vitals were normal. Abnormal laboratory investigations included raised total bilirubin (6.6 mg/dl), raised alkaline phosphatase (343 IU/L) and raised alanine transaminase levels (284 IU/L). Hepatitis serology for HbsAg, anti-HCV, HIV1 & II were negative.

On evaluation with CECT & MRCP he was found to have mass lesion involving the biliary confluence with upstream dilatation of biliary tree. (Fig. 1A and B) Provisional diagnosis of hilar cholangiocarcinoma with type III hilar block was made. In view of possible requirement of major liver resection percutaneous transhepatic biliary drainage was done. The patient was taken up for surgery after cardiopulmonary evaluation and clearance. Intraoperatively mass lesion was found which was encasing right hepatic artery with no evidence of

metastasis. Intraoperative proximal and distal bile duct resection margins were sent for histopathological frozen section examination and both were reported as negative for dysplasia and malignancy. In view of type III hilar block and right hepatic artery encasement the patient underwent Right hepatectomy with caudate lobectomy with complete CBD excision with Roux en Y hepaticojejunostomy. The patient tolerated the surgery well.

Macroscopically, cut surface of the resected specimen showed a greyish-white firm tumour like lesion measuring 25x25 mm and involving confluence of hepatic ducts, common hepatic duct, cystic duct, neck of gall bladder and part of common bile duct. (Fig. 2) Histologically, the lesion showed hyalinized fibrocollagenous tissue, plump fibroblasts, histiocytes, lymphocytes, myofibroblasts and sheets of plasma cells, involving all the layers of affected biliary system. (Fig. 3A and B) Hyperplastic nerve bundles, focal vasculitis and perivascular collagenization was also seen. Hepatic artery

Fig. 2 Gross specimen showing a greyish-white firm tumour like lesion involving confluence of hepatic ducts, common hepatic duct, cystic duct, neck of gall bladder and part of common bile duct

Fig. 3 (A) Histopathology of hilar mass composed of dense hyalinized fibrocollagenous tissue with large number of plasma cells (HE, $\times 200$) (B) High power view shows sheets of plasma cells with plump fibroblasts, myofibroblasts and histiocytes in fibrocollagenous tissue (HE, $\times 400$)

showed presence of atherosclerotic plaque and recanalization. (Fig. 4) Hilar lymph nodes showed reactive lymphoid hyperplasia. Sections from the liver showed early bilirubinostasis without significant inflammation or fibrosis. Gallbladder showed chronic cholecystitis. No evidence of malignancy, atypical mitosis or necrosis was seen in the multiple sections obtained after extensive dissection of the entire resected biliary system. Immunohistochemical analysis showed SMA, Vimentin and CD34 immunostaining in plump myofibroblasts. The inflammatory plasma cells did not reveal monoclonality on kappa and Lambda immunostaining. Very few plasma cells revealed IgG4 positive staining. Histopathological diagnosis of Hilar Inflammatory Pseudotumour with Hepatic Artery Atherosclerosis was rendered.

His condition at discharge was good with stable vitals, was tolerating normal diet well, afebrile, ambulatory and normal LFT and is doing well on follow-up of 6 months.

Fig. 4 Hepatic artery shows partial luminal obliteration by atherosclerotic plaque (Masson Trichrome, $\times 100$); composed of sheets of foam cells, cholesterol clefts with histiocytic giant cell reaction and fibroblasts (Inset HE, $\times 400$)

Discussion

Inflammatory pseudotumor (IPT) is a term used to describe a benign and rare process most commonly involving the lung and orbit but found in nearly every site in the body.[2].

IPT was first recognised in 1939 in lungs, hepatic IPT was first described by Pack & baker in 1953.[3] Many etiologies including infection & immunological/allergic reactions are suggested, however exact etiology unclear.

IPT was so named by Umiker et al.[4] in 1954, because of its propensity to clinically and radiologically mimic a malignant process.

Hilar cholangiocarcinoma, also known as Altemeier-Klatskin tumour occurring within 2 cm of the hilar confluence, accounts for about 50–70 % of all cholangiocarcinomas. Resection and in selected patients, transplantation offers the best chance of cure in Klatskin tumours.[1] Klatskin tumours are difficult to differentiate from a number of benign pathologies and some malignant lesions that mimic the clinical presentation and the radiological appearances, adding to the complexity of management in patients suspected to have cholangiocarcinoma.[5].

Inflammatory pseudotumor of the biliary tract is a benign disease, and is extremely rare. Diagnosis often depends on pathological examination after operation. Hepatic IPT

Table 1 Cases of IPT involving Hilar biliary confluence

Author	Year	Journal
Deng FT et al.	2010	Hepatobiliary Pancreat Dis Int
Nonomura A et al.	1997	Pathology- Research & Practice
Gohy S et al.	2007	Hepatogastroenterology

accounts for 0.4 % of all focal liver lesions treated by hepatectomy [6] and there are very few reports of IPT involving hilar biliary confluence.[7–9] (Table 1).

The clinical manifestations and imaging appearance of the tumor are similar to those of a neoplasm, but tumor differs from the malignant one in benign biological behavior and the property of spontaneous regression.[10,11] Patients with IPT of the hepatic hilar biliary.

Duct present with fever, jaundice, and manifestations indistinguishable from those of cholangiocarcinoma.[8,9].

At histologic analysis, IPT is characterized predominantly by an inflammatory infiltrate consisting of lymphocytes, plasma cells, and histiocytes admixed with a variable proportion of fibroblasts and myofibroblasts.[5].

One peculiar findings in our case was presence of coexistent hepatic artery atherosclerosis. There is a single study by Saiura et al., in which benign left duct stricture in a 79 year old male was associated with atherosclerosis of hepatic artery.[12].

It not always possible, to established diagnosis of hilar cholangiocarcinoma before embarking on a radical resection. Negative report on intraoperative frozen section examination of resection margins does not necessarily mean the absence of malignancy. Few similar previously published reports of Klatskin mimicking IPT have highlighted the pre- and intraoperative difficulty in the diagnosis of this benign entity.[7–9].

Conclusion

Hilar IPT although rare, constitutes a diagnostic and therapeutic challenge as clinical manifestations and imaging appearance of IPT are similar to those of Klatskin tumour and diagnosis often on pathological examination after operation. IPT should be kept in mind as a differential diagnosis of Klatskin mimicking lesions.

Conflict of interest None.

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