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Gastrointestinal Imaging *Case Report*

Inflammatory Myofibroblastic Tumor of the Porta Hepatis: A Case Report

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ABSTRACT

A 43-year-old man presented with painless jaundice. Imaging revealed a porta hepatis mass compressing the common bile duct. Endoscopic biopsy was negative for malignancy. Complete surgical resection was performed. Pathological assessment showed IGg4 negative inflammatory myofibroblastic tumor.

Keywords: Inflammatory pseudotumor, Myofibroblastic tumor, Porta hepatis

INTRODUCTION

Inflammatory pseudotumor (IPT) represents a wide spectrum of nonneoplastic and neoplastic entities. These include (a) inflammatory myofibroblastic tumor (IMT), (b) pseudosarcomatous myofibroblastic proliferation of the genitourinary tract, (c) postinfectious/reparative disorders, and (d) IPTs of the lymph node (LN), spleen, and orbit. Although initially described in the lung, extrapulmonary IPTs have been described in many somatic and visceral sites. IMT has emerged as a distinct pathologic entity from the broad category of IPTs.[1,2] Few cases of hepatic IMT have been previously reported, moreover, porta hepatis/bile duct IMT are rarer entity and has been less frequently reported.[3,4]

We present a case of porta hepatis IMT and discuss its CT and MRI imaging features.

CASE REPORT

A 43-year-old man presented to the gastroenterology clinic with 1 month history of progressive yellowish discoloration of his sclera and skin with associated pruritis. His past medical history was unremarkable. On clinical examination, he had no pain or tenderness in the right upper quadrant. Laboratory workup revealed markedly elevated bilirubin 407 umol/L with direct bilirubin of 308 umol/L, he had slightly elevated liver enzymes, as well as alpha-fetoprotein AFP 9.5 IU/ml and carcinoembryonic antigen CEA 3.4 IU/ml. Serum lipase and amylase were within the normal range.

CT examination of the abdomen was performed [Figure 1] and revealed diffuse dilatation of the intra-hepatic biliary system and the common bile duct (CBD), with a well-defined soft-tissue mass compressing the infero-posterior aspect of the CBD. MRI and MRCP were done to further evaluate the mass and the biliary system [Figures 2 and 3].

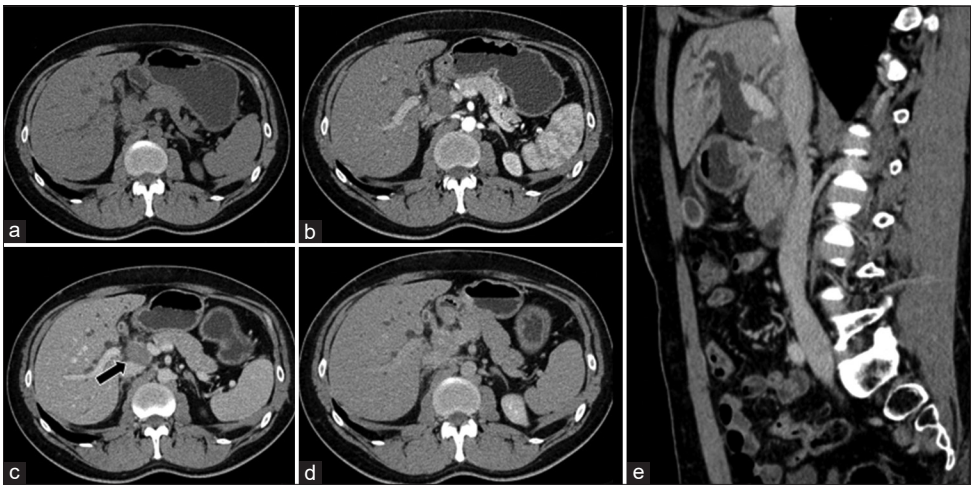

Figure 1: A 43-year-old man presented with porta hepatis inflammatory myofibroblastic tumor who presented with painless jaundice and pruritis. (a) axial pre-contrast CT image, (b) axial post-contrast CT image in arterial phase, (c) axial post-contrast CT image in porto venous phase and (d) axial post-contrast CT image in delayed phase show a heterogeneously enhancing mass at the porta hepatis [black arrow in (c)]. (e) Sagittal reformatted post-contrast CT image shows the mass compressing the common bile duct with upstream biliary dilatation. No vascular invasion was identified. The pancreas and liver were normal.

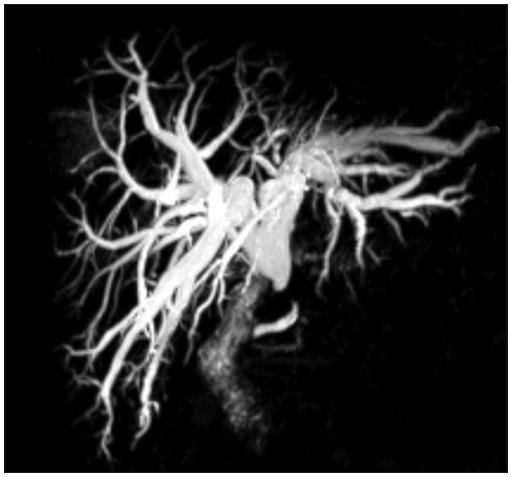

Figure 2: A 43-year-old man presented with porta hepatis inflammatory myofibroblastic tumor who presented with painless jaundice and pruritis. MRCP image shows diffuse dilatation of the intrahepatic biliary radicles and proximal common bile duct (CBD) with narrowing noted at the middle third of the CBD.

ERCP and biopsy from the mass were performed, with placement of CBD stent [Figure 4]. The histopathological result revealed no malignant cells or lymphoid tissue.

After stent placement, the bilirubin level decreased (total 163 umol/L, direct 136 umol/L), however, remained above the normal limits.

The case was discussed in the tumor board, the main radiological differential diagnosis was extra-hepatic cholangiocarcinoma and porta hepatis LN enlargement either primarily or metastatic, however, negative biopsy results, regular borders of the mass, the absence of infiltrative features or other primary tumors or LNs involvement was against these diagnoses. The third differential of IPT was suggested. Initial decision of short-term follow-up was taken. Two months later, CT examination was repeated (not shown) and revealed no significant changes. After multidisciplinary rediscussion, as well as discussion with the patient, the decision was to perform open surgical biopsy and removal instead of repeating the endoscopic biopsy.

At surgery, there was soft-tissue mass at the inferior posterior aspect of the middle/distal CBD, frozen sections were taken from the mass and confirmed its benign fibrotic nature, the mass was infiltrating the CBD, so excision of the mass with preserving the CBD was not possible. The mass was dissected and excised together with the distal CBD, choledochojejunostomy was performed as well as cholecystectomy. The final pathological results revealed bland spindle cell proliferation with marked lymphoplasmacytic infiltrate as well as eosinophils and rare lymphoid aggregations. The spindle cells were diffusely positive for Vimentin and negative for ALK and beta-catenin. Immune staining for IgG4 was negative with the final diagnosis of IMT. The margins of the CBD were invaded by the mass as well. The gall bladder was normal.

DISCUSSION

IPTs are characterized by an inflammatory infiltrate consisting of lymphocytes, plasma cells, and histiocytes mixed with a variable proportion of fibroblasts and myofibroblasts. IPTs have also been described as plasma cell granulomas, histiocytomas, fibroxanthomas, and inflammatory fibrosarcomas. IMT has previously been

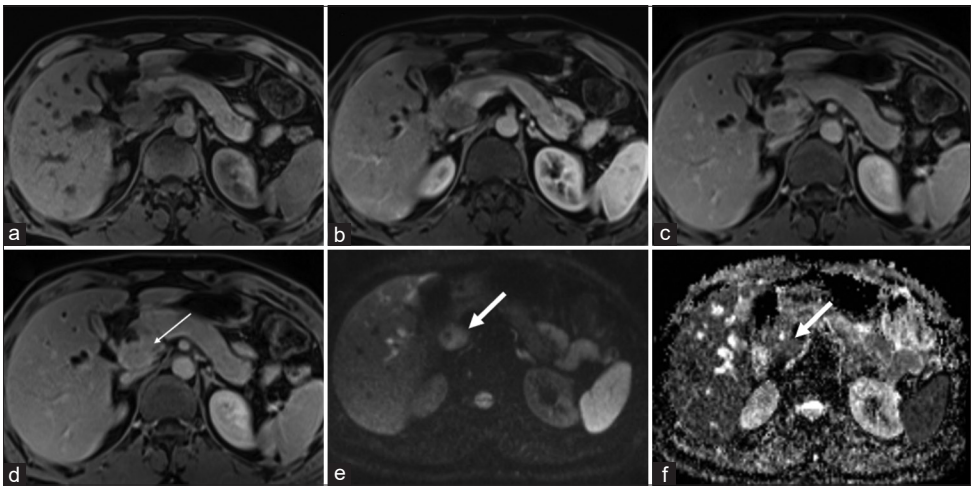

Figure 3: A 43-year-old man presented with porta hepatis inflammatory myofibroblastic tumor who presented with painless jaundice and pruritis. (a) axial pre contrast MR image, (b) axial post-contrast MR image in the arterial phase, (c) axial post-contrast MR image in porto venous phase, and (d) axial post-contrast MR image in delayed phase show a heterogeneously enhancing mass at the porta hepatis. The mass is of intermediate T1 signal intensity with no fat component. The mass shows mild arterial enhancement with progressive enhancement throughout the delayed phases [thin white arrow]. (e) axial DWI image and (f) corresponding axial ADC image show mild diffusion restriction [bold arrow]. No vascular invasion was identified.

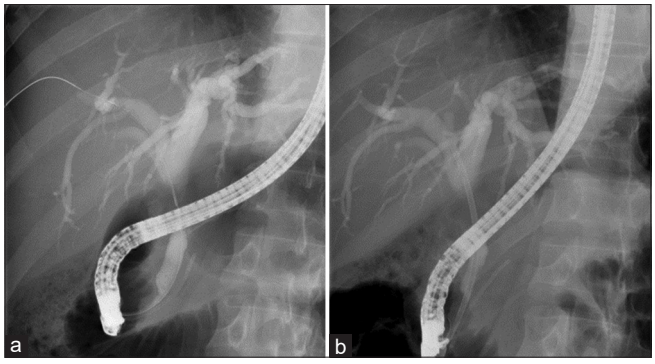

Figure 4: A 43-year-old man presented with porta hepatis inflammatory myofibroblastic tumor who presented with painless jaundice and pruritis. (a) ERCP image after cannulation of the common bile duct (CBD) shows stricture at the middle third of the CBD with subsequent proximal CBD and intrahepatic biliary radicles diffuse. (b) ERCP image after stent placement.

included in IPT and both terms were used interchangeably in the literature. However, recent histologic descriptions have separated IMTs from the general group of IPTs with their own biology and behavior. IMTs have a tendency for local recurrence and low risk of metastasis, thus they are currently classified as tumors of intermediate biological potential by the World Health Organization.[1]

IMTs are characterized histologically by a spindle cell proliferation in a myxoid to collagenous stroma with a prominent plasma cell and lymphocyte infiltration.[1,3]

IMTs have been attributed to different etiologies including trauma, infection, inflammatory, and postoperative conditions. In our case, the patient did not have any of these predisposing conditions. Hepatic IPT was first reported by Pack and Baker.[5] Intrahepatic IPTs has been frequently reported, the largest case series was reported by Yang *et al.*[4] Hepatic hilum or bile duct IMTs have been rarely reported. To the best of our knowledge, 24 cases have been reported in the literature.[3,6,7]

The clinical presentation of IMTs is non-specific and depends on the site of involvement. Obstructive jaundice has been the most common clinical finding followed by nausea, vomiting, abdominal pain, and fever. The frequent presentation with jaundice explains the usual overlap in diagnosis between IMTs and cholangiocarcinoma.[3]

The imaging appearance of IMTs is variable. In their analysis of 114 cases of hepatic IMTs, Yang *et al.*[4] reported that the most common CT findings are well-defined hypodense masses with heterogeneous arterial enhancement. Similar to our case, most of the masses were T1 hypointense and T2 isointense or hypointense. Variable post-contrast enhancement patterns were reported and the most common was delayed enhancement due to the desmoplastic reaction. Kang *et al.*[8] investigated gadoxetic acid-enhanced MRI and FDG-PET-CT in 18 intrahepatic IMTs, in their study, all cases showed diffusion restriction and hyperaccumulation of FDG, none of the cases showed intralesional fat. They classified the enhancement pattern into five categories, the most common pattern was a well-defined target-like hypervascular mass with peripheral hypointensity rim on arterial and hepatobiliary phases, central necrosis was common in these hypervascular masses. The other less common patterns included hypovascular mass, heterogeneous enhancing mass,

sclerosing mass, and non-target hyperenhancing mass. The differential diagnosis of hilar IMTs is cholangiocarcinoma. Due to the non-specific imaging features, intra-hepatic IMTs should be considered in the differential of hepatic focal lesions such as hepatocellular adenoma and carcinoma, especially in young patients with normal tumor markers. In rare cases, hepatic IMTs can mimic liver abscess if there is central necrosis.[1,9] Surgical resection is the treatment of choice for IMTs, depending on the site of involvement and respectability of the tumor. Resection usually requires reconstruction due to the frequent involvement of the biliary tree and hepatic vessels.[6] In our patient, the infiltration of the CBD by the tumor required resection with choledochojejunostomy. Vascular invasion has been frequently reported in hilar IMTs and was treated by liver transplant or pancreaticodudenectomy. Berumen *et al*. reported combined liver transplant and pancreaticodudenectomy with vascular reconstruction due to hepatic artery involvement.[1] Anti-inflammatory drugs and chemotherapy have been used in previous studies with variable reported success rates. Although there is no consensus regarding follow-up after treatment, some individual cases reported aggressive nature of the disease with recurrence after treatment.[1,3,7,9]

CONCLUSION

IMTs are rare, benign tumors that can affect multiple organs. Bile duct IMTs were rarely reported. Imaging diagnosis of biliary IMTs is challenging due to overlapping features with neoplastic conditions, especially cholangiocarcinoma. Surgical resection is the curative treatment of choice.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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