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Inflammatory Myofibroblastic Tumor of the Distal Bile Duct Mimicking Cholangiocarcinoma: A Case Report

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Abstract **Case Report**

Inflammatory myofibroblastic tumor has been described in various locations. However, its appearance in the periampullary region is uncommon and has rarely been reported in the literature. We report a case of a 37-year-old Moroccan male with painless jaundice and pruritus. Endoscopic retrograde cholangiopancreatography and magnetic resonance cholangiopancreatography showed a 2-cm-long stricture in the distal bile duct that suggests a cholangiocarcinoma. There was no involvement of adjacent structures at computed tomography of abdomen. The patient underwent a pancreaticoduodenectomy. The tumor histology showed regular spindle cells arrayed in fascicles, admixed with lymphocytes, plasma cells and eosinophils. Immunohistochemical analysis showed positive staining for smooth muscle actin, H-caldesmon. In contrast, the tumor cells did not express cytokeratin, PS100, CD68, CD23, CD117 and ALK. Based on these data, the diagnosis of inflammatory myofibroblastic tumor of distal bile duct was made. We report this new case in the aim of shedding more light on this entity and the challenges in making the diagnosis in such location.

Keywords: Inflammatory myofibroblastic tumor, distal bile duct, cholangiocarcinoma.

INTRODUCTION

Inflammatory myofibroblastic tumors (IMT) of the biliary tract are extremely rare and heterogeneous by etiology and clinical presentation. They might cause biliary obstruction and mimic cholangiocarcinoma and their final diagnosis is usually achieved only after surgical excision. Its localization in the periampullary region is exceedingly rare with few cases reported in the literature [1]. We report this new case in the aim of shedding more light on this entity and the challenges in making the diagnosis as clinical presentation and radiological features are similar to cholangiocarcinoma.

Patient Information

We describe a 37 year old Moroccan man admitted to our department in 2014 with painless jaundice and pruritus. His medical history was unremarkable.

CLINICAL FINDINGS

The physical examination doesn't reveal any abnormalities except for his icterus. Our patient denied any weight loss or other constitutional symptoms and he did not take any conventional or alternative medications. His liver, spleen and superficial lymph nodes were not enlarged and there was no sign of any palpable mass.

Diagnostic Assessment

Serum tumor markers CEA and CA-19.9 were normal. The liver function tests were markedly raised. A trans- abdominal ultrasonography revealed dilatation of the intrahepatic and extrahepatic bile ducts with no visible obstacle. Endoscopic retrograde cholangiopancreatography was performed. It showed a 2-cm-long stricture in the distal bile duct that suggests a cholangiocarcinoma with dilatation of the intrahepatic and extrahepatic bile duct. Magnetic resonance cholangiopancreatography confirmed the presence of an irregular stricture in distal bile duct that was suggestive of cholangiocarcinoma. There was no evidence of metastatic disease at computed tomography of abdomen.

Therapeutic Intervention

The patient was considered for surgical resection of a suspected cholangiocarcinoma of distal bile duct. He underwent a pancreaticoduodenectomy. At gross examination, the pancreas doesn't show any

macroscopic abnormality. The distal bile duct measured was completely filled by a yellowish-white and firmed tumor measuring 2 x 1,7 cm. (figure 1). Histologically, the tumor was made by a proliferation of regular spindle cells arrayed in fascicles, admixed with lymphocytes, plasma cells and eosinophils (Figures 2). Immunohistochemical analysis showed positive staining for smooth muscle actin (Figure 3), and H-caldesmon (Figure 4). In contrast, the tumor cells did not express CK (Figure 5), ALK1, CD23, CD68, PS100 and CD117. Based on these data, the diagnosis of inflammatory myofibroblastic tumor of distal bile duct was retained. There is no inflammatory or tumoral lesion in pancreatic parenchyma. The surgical margins were free of tumor.

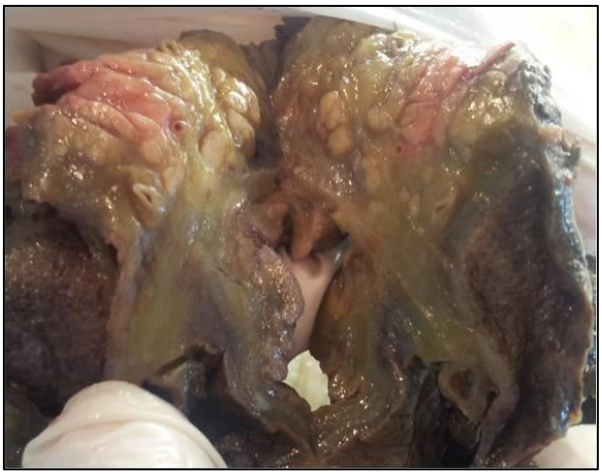

Figure 1: Macroscopic finding: The distal bile duct is completely filled by a yellowish-white and firmed tumor

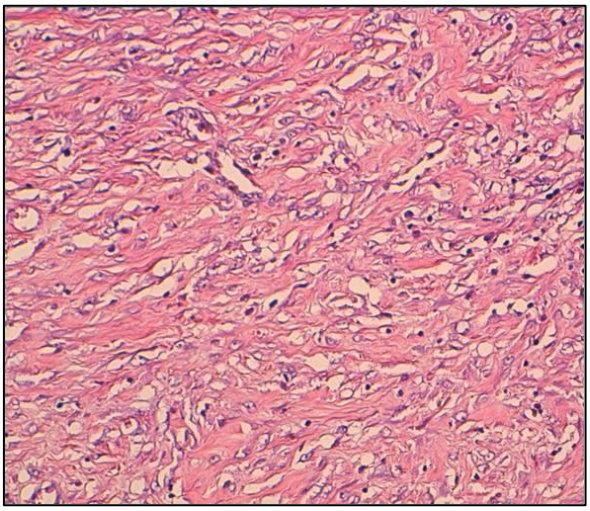

Figure 2: Regular spindle cells arrayed in fascicles, admixed with lymphocytes, plasma cells and eosinophils (hematoxylin-eosin × 100)

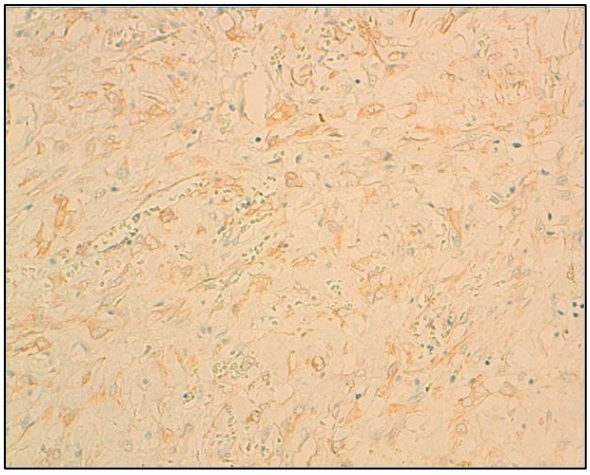

Figure 3: Immunohistochemical analysis showed that Spindle cells express smooth muscle actin

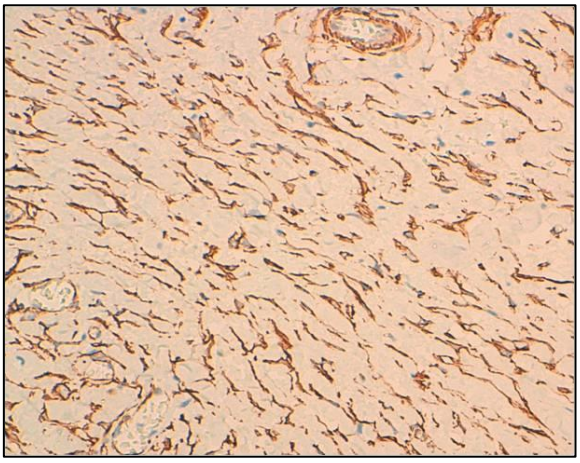

Figure 4: Immunohistochemical analysis showed that Spindle cells express H-caldesmone

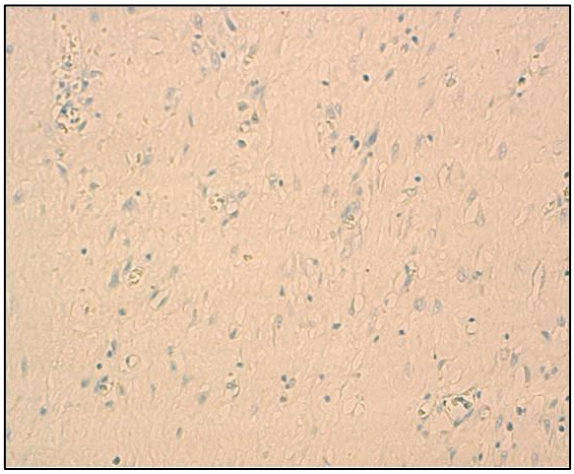

Figure 5: Immunohistochemical analysis showed negative staining for cytokeratin

Follow-up and Outcomes

At 8 years from his surgery, the patient is doing well and there are no radiological findings of recurrent disease.

DISCUSSION

IMT (synonym plasma cell granuloma, inflammatory myofibroblastic tumour) are extremely rare benign lesions of unknown etiology. They have been described in many localization including lymph nodes, spleen, brain, spinal cord, larynx, thyroid gland, breast, pancreas, gastrointestinal tract and bladder, but most frequently it occurs in the lungs. IMTs of the biliary tree are less frequent and, there is very few cases that have been reported (table 1) [2]. Among these cases, only two cases have been described in the distal bile duct [3, 4].

Table 1: Characteristics of patients with inflammatory pseudotumours of the biliary system

Authors	Age at presentation and symptoms	Location	Therapy	Follow-up
Haith *et al.,*	6-year-old, boy, vomiting, diarrhea, fever and obstructive jaundice	Distal common bile duct	Surgical resection	5 months, no recurrence
Stamatakis *et al.,*	13-year-old girl, abdominal pain and obstructive jaundice	Proximal common bile duct, cystic duct, common hepatic duct	Surgical resection	21 months, no recurrence
Ikeda *et al.,*	43-year-old man, fever, weight loss and obstructive jaundice	Intrahepatic ducts, common hepatic duct, common bile duct	Surgical resection	7 months, no recurrence
Pokorny *et al.,*	43-year-old man, nausea, epigastric pain and obstructive jaundice	Right intrahepatic duct, anterior right hepatic lobe, porta hepatis, proximal common bile duct	Surgical resection	5 year, no recurrence
Ozeki *et al.,*	73-year-old man, obstructive jaundice	Bile duct bifurcation Surgical resection with an extended right hepatic and caudate	lobe resection	Not applicable
Fukushima *et al.,*	58-year-old woman, obstructive jaundice	Mid and distal common bile duct	Surgical resection with duodenopancreatectomy	Not applicable
Walsh *et al.,*	50-year-old man, obstructive jaundice	Proximal common bile duct	Surgical resection	19 years, recurrent disease submandibular gland, pancreatic tail, kidne
Walsh *et al.,*	69-year-old woman, obstructive jaundice	Bile duct bifurcation	Chemotherapy followed by surgical resection	21 years, recurrent disease left intrahepatic bile duct. Treated with resection and multiple endoscopic stents
Saint-paul *et al.,*	53-year-old man, jaundice and weight loss	Intrahepatic bile duct	Surgical resection: partial hepatectomy	Not applicable
Sobesky *et al.,*	51-year-old woman	Distal common bile duct	Surgical resection: duodenopancreatectomy	No recurrence
Ashcroft *et al.,*	50-year-old woman, epigastric pain, obstructive jaundice and significant weight loss	Common hepatic duct	Stent placement followed by surgical resection	8 years, no recurrence
Ashcroft *et al.,*	61-year-old man, right upper quadrant abdominal pain and obstructive jaundice	Common hepatic duct and cystic duct	Stent placement followed by surgical resection	6.5 years, no recurrent disease
Abu-wassel *et al.,*	55-year-old man, obstructive jaundice	Common bile duct, common hepatic duct	Stent placement, right portal vein embolisation, right extended hepatic and extrahepatic bile duct resection	16 months, no recurrent disease
Current case	37 year old man, painless jaundice	Distal common bile duct	Surgical resection: duodenopancreatectomy	8 years , no recurrent disease

The etiology of IMTs remains unclear. Some authors believe that auto immunogenic processes represent the underlying etiology of IMT. Moreover, many recent studies, suggest that it might be a true neoplasm due to the presence, at the myofibroblastic component, of a fusion gene involving the ALK gene, a

tyrosine kinase oncogen located on chromosome 2p23, initially found to be arranged in anaplasic large cell lymphomas. This fusion leads to constitutive over-expression of the ALK, causing cell proliferation. These changes are most frequent in abdominal and pulmonary IMTs in the first decade of life and are associated with a higher frequency of recurrence [1, 2]. In our case the tumor doesn't show reactivity with ALK1.

The clinical manifestations and imaging appearance of IMT of biliary tree are similar to those of a cholangiocarcinoma, but it differs in benign biological behavior and the property of spontaneous regression. The most common clinical finding is obstructive jaundice. Other symptoms may include abdominal pain, nausea, pruritus vomiting and fever [5-8].

Radiological aspects are variable and nonspecific. At magnetic resonance images of hepatobiliary, it's difficult to make a differentiation between IMT and cholangiocarcinoma or lymphoma because there are considerable similarities [9].

Macroscopically, the tumor is firm, well-defined, white, grey or brownish-yellow, and non-encapsulate. Histologically, IMT includes three histological subtypes: one is a richly vascularized and myxoïde resembling fasciitis or granulation tissue. Another is a more compact fascicular spindle cell proliferation with variable collagenized regions and lymphoid nodules, resembling fibromatosis and finally a very sclero-hyaline, slightly cellular pattern, looking more like a desmoid tumor. Immunohistochemistry showed reactivity for vimentin and smooth muscle actin. Immunohistochemical positivity for ALK is detectable in just over half of the cases with cytoplasmic staining, more rarely at the nuclear membrane. The main pathological differential diagnoses includes other spindle cells tumor like leiomyosarcoma, fibrosarcoma, Malignant peripheral nerve sheath tumours, sarcomatoid carcinoma, stromal tumor and follicular dendritic cells tumor [2, 10].

In all previously reported IMT cases, surgical resection was the treatment of choice [4, 10]. This can be attributed to the radiological findings that are suspicious for cholangiocarcinoma in addition to inconclusive cytology and in some cases even on histological inspection. The extent of the resection depends on the intra-operative findings. The indicator of benign disease is the negative margin shown by fast frozen section analysis. Thus some authors recommend intra-operative examination of fast frozen section during IMT surgery [2].

The reported long-term outcome for these patients when managed by surgical excision and biliary-enteric anastomosis appears to be excellent [1, 5]. Only one report has raised the possibility of local or distant recurrence with long-term follow-up [11].

CONCLUSION

The diagnosis of biliary tree IMTs is challenging as both its clinical presentation and radiological features are similar to biliary tract malignant neoplasms. The final diagnosis of the tumor relies mainly on the pathological examination of the surgical specimen. Surgical resection represents the treatment of choice, ensuring a satisfactory prognosis.

Patient Perspective

The patient is satisfied with the treatment he received.

Informed Consent

The patient has given informed and written consent for publication.

COMPETING INTERESTS

The authors declare that they have no competing interests.

Author's Contributions

MS and AB retrieved clinical information, performed the literature review, wrote and approved the final manuscript.

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