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Inflammatory Myofibroblastic Tumor (IMT) of Biliary Ducts: A Pediatric Case

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Abstract

The Inflammatory Myofibroblastic tumor (IMT) are a rare group of solid lesions of mesenchymal origin with intermediate biological potential; in children, they predominate in the first decade of life. They present as single lesions in 60% of cases, and the abdomen is the second most frequent site of presentation. The etiology of IMT is unclear, and the diagnostic is not easy. The treatment of choice is complete surgical resection, which constitutes the only curative treatment, liver transplant has been described when the IMT cannot be resected from this organ. The history of the patient that we are going to talk about is like what is described in the literature, with gastrointestinal involvement; and the extension of the lesion limiting the possibility of surgical resection. Our patient had a mass involving the convergence of the hepatic ducts, was diagnosed with IMT; without possibility of surgical resection, making the patient a candidate for liver transplant. Liver transplant was performed successfully, and the patient has recovered.

Keywords: Myofibroblastic tumor; Liver tumor; Pseudotumor

Introduction

Inflammatory myofibroblastic tumor is a lesional pattern of inflammatory pseudotumour. It is characterized by a mix of inflammatory cells, e.g. plasma cells, lymphocytes and eosinophils etc. The liver is the most commonly involved organ among the intra-abdominal organs. It is diagnosed based on their appearance under the microscope. The treatment of choice is complete surgical resection. This case report present about a 10-year-old patient with two month medical history of jaundice.

Case Report

A 10-year-old patient presented with a two-month history of jaundice, choluria, acholia, and intermittent right upper quadrant abdominal pain, without fever [1]. Ultrasound of the liver and bile ducts and abdominal computed tomography were performed, showing dilation of the intrahepatic biliary tract, predominantly involving its central portion. Abdominal nuclear magnetic resonance imaging (MRI) showed amputation of the hepatic ducts prior to the confluence of the common hepatic duct. Blood tests showed direct hyperbilirubinemia, increased gamma glutamyl transferase and alkaline phosphatase, negative infectious disease profile, and negative tumor marker results [2,3].

Cholangiopancreatography showed a 21 mm mass involving the convergence of the hepatic ducts, with dilation of the intrahepatic biliary tract, findings were compatible with cholangiocarcinoma *versus* rhabdomyosarcoma. The patient was referred for a pediatric oncology consult, considering the potential diagnosis of biliary rhabdomyosarcoma.

Cholecystectomy and endoscopic retrograde cholangiopancreatography were performed to bypass the bile ducts, but stenting was not possible, an external biliary bypass and tricot biopsy were performed, resulting in decreased jaundice and reduced bilirubin level.

Preliminary biopsy report revealed a myofibroblastic proliferation compatible with a benign tumor. The possibility of surgical resection was limited, with a high risk of insufficient remnant liver tissue. The patient was thus a candidate for liver transplant.

Liver transplant was performed from a cadaveric donor. Intraoperatively, a nodule was noted on the gastric greater curvature, which was resected. Both lesions were sent for histological analysis. The biliary lesion was described as inflammatory myofibroblastic tumor associated with IgG4 disease of the biliary confluence, with obstruction of the right and left hepatic ducts, and secondary sclerosing cholangitis. The immunophenotype was negative for ALK. The gastric lesion was described as a calcifying fibrous tumor of the stomach, with slight lymphoplasmacytic inflammatory infiltrate. Six months after transplant the patient was asymptomatic, with immunosuppressant treatment and decreasing gamma glutamyl transferase levels.

Discussion

Myofibroblastic tumor was first described by Brumm in 1939, in a case of lung tumor, and the first description of myofibroblastic tumor of the liver was published by Pack and Beacker [1]. IMT are a group of solid lesions of mesenchymal origin with intermediate biological potential [2]. They present at any age, predominantly in the third decade of life; in children, they predominate in the first decade of life [1]. There is a male predominance, with a 3.5: 1 ratio [1].

IMT occur from the brain to the bladder. The abdomen is the second most frequent site of presentation [3]. Extrapulmonary forms have been reported to be associated with Wilms tumor, chronic sclerosing cholangitis, Hodgkin's lymphoma, acute myelomonocytic leukemia, and adrenocortical tumors [3]. They present as single lesions in 60% of cases [1]. They tend to be locally invasive, with a high probability of recurrence and a low risk of distant metastases [4].

The etiology of IMT is unclear, but the following have been proposed: genetic factors, trauma, surgery, use of steroids, autoimmune reactions, radiation therapy, viral and bacterial infections [1-6]. Fifty percent of cases demonstrate ALK gene rearrangement, with this feature being more frequent in women and children. ALK rearrangement is associated with a better prognosis due to its less aggressive behavior [6,7].

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Tumor regression and control have been achieved in neoplasms showing ALK translocations with the use of ALK-directed therapy [6]. Infiltrating IgG4 positive plasma cells can be seen. It has been proposed that this IgG4 related tumor, not associated with autoimmune pancreatitis, is part of the spectrum of IgG4 related diseases, and should be considered in the differential diagnosis of hepatic tumors in children, and, therefore, the study of these tumors should be widened to include their association with serum immunoglobulins [8-10].

Diagnostic tools include ultrasound and computed tomography, which can show a dilated intrahepatic biliary tract and liver masses. Cholangiopancreatography is not always specific. Biopsy is the gold standard for diagnosis [1].

The treatment of choice is complete surgical resection, which constitutes the only curative treatment. In cases of incomplete resection, combined treatment with chemotherapy, steroidal and non-steroidal anti-inflammatory drugs, and biological therapy (anti-TNF), has been reported [1-8]. Liver transplant has been described when the IMT cannot be resected from this organ. The use of ALK inhibitors in selected inflammatory myofibroblastic tumors is under investigation [6].

Calcifying gastric tumors are infrequent, benign, mesenchymal lesions, usually asymptomatic, and are commonly diagnosed as incidental findings during endoscopic and radiologic procedures [11,12].

Retrospective studies suggest that the incidence of gastric submucosal lesions is 0.36% [12,13]. Their etiology is unknown. Some authors suggest that calcifying fibrous tumors represent a sclerosing phase of inflammatory myofibroblastic tumor. However, the characteristic expression of ALK1 associated with myofibroblastic tumors is not seen in calcifying tumors [14]. Our patient's IMT is similar to what is described in the literature in terms of age at presentation and systemic clinical signs, with gastrointestinal involvement. Immunophenotypically, it was positive for IgG4 and negative for ALK. ALK negativity gives this tumor a worse prognosis due to its increased aggressiveness, as demonstrated in this case, with localized extension of the lesion preventing the possibility of surgical resection.

Conclusion

The presence of a calcifying fibrous gastric tumor with similar immunohistochemical characteristics to the biliary tumor, and which seems to correspond to the involution of an inflammatory myofibroblastic tumor of the stomach, probably prior to the biliary tumor, was incidentally documented.

This report describes the case of a patient with IMT of the biliary ducts treated with liver transplant. We believe that our report makes a significant contribution to the literature because this case demonstrates the diagnostic difficulty of this rare neoplasm and suggests the need to consider this diagnosis in the pediatric population. This case also demonstrates the potential for locally aggressive behavior.

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