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Radiology

Imaging of Botryoid Rhabdomyosarcoma of the Biliary Tract: Case Report

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Abstract **Case Report**

Botryoid Rhabdomyosarcoma of biliary tree is a disease of young children who may arise in any part of the biliary tree or in a choledochal cyst. It is a rare tumor; however, it is the most common cause of obstructive jaundice due to neoplastic biliary obstruction in children, following nonneoplastic conditions such as choledochal cyst, congenital anomalies of the biliary tree, biliary atresia, and gallstones. The management and prognosis of this tumor are constantly changing as new imaging techniques and chemotherapy as initial treatment become available. We will report highlights certain imaging features that may help in early diagnosis of Botryoid Rhabdomyosarcoma and differentiation from other mimicking conditions.

Keywords: Biliary choledochal cyst, obstructive jaundice, pediatric, rhabdomyosarcoma.

INTRODUCTION

Rhabdomyosarcoma (RMS) accounts for 10 to 15% of solid tumors in children under ten years of age. It primarily affects the urogenital tract and soft tissues of the head and neck region. RMS of the biliary tract is rare and has a poor prognosis as it represents 1% of all rhabdomyosarcomas in children and 0.04% of pediatric cancers [1–3].

However, it is the most common cause of malignant jaundice in children [4]. It is often of botryoid type and localized in the main bile duct or intrahepatic ducts.

In order to diagnose a biliary tract RMS, a set of clinical, biological and radiological arguments is necessary. Thus, we will focus here on the radiological aspects of biliary RMS through this case report, before discussing it in the light of the literature.

CASE REPORT

A 4-year-old boy with non-existing medical history presented with a cholestasis syndrome consisting of mucocutaneous icterus, pruritus, and discolored stools that had been evolving for 1 month. The initial clinical examination revealed a fever of 39°C and hepatomegaly.

On the biological level, a cholestasis syndrome was found with elevated bilirubinemia with a predominance of conjugated bilirubin and elevated GGT as well as a slight hepatic cytolysis.

Pancreatic enzymes were measured and returned normal.

An abdominal ultrasound was performed and showed:

- A moderate dilatation of the intrahepatic bile ducts.
- A segmental cystic dilatation of the main bile duct measuring 24 mm in diameter, within which was a tissue-like formation, vascularized on Doppler mode, measuring 17x14 mm.

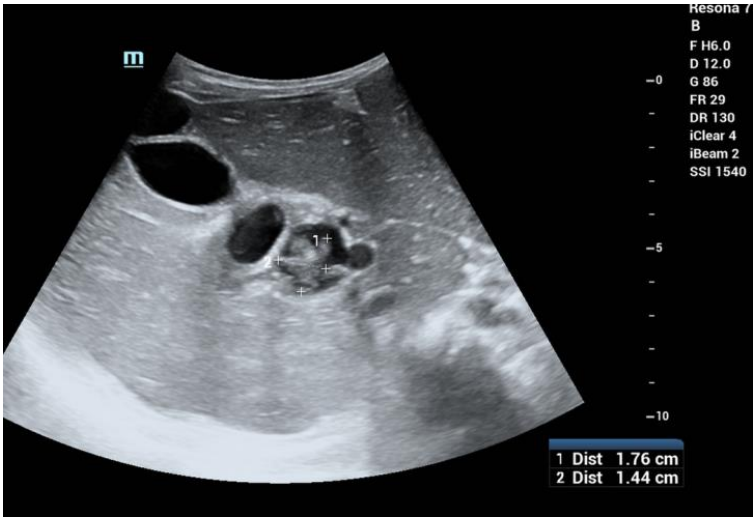

Figure 1: Cystic dilatation with tissue-like formation

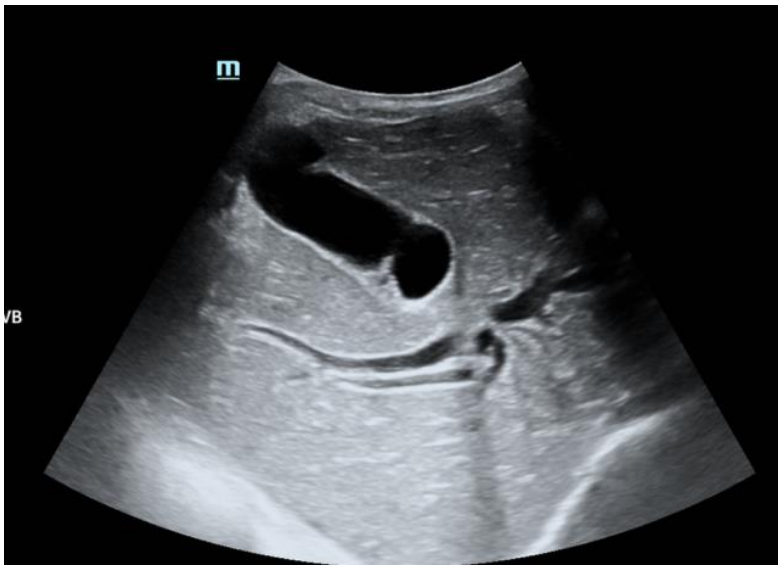

Figure 2: Dilated bile ducts

An MRI of the biliary tract was conducted and showed:

- Dilatation of the intrahepatic bile and main bile ducts, with the presence of a hypointense T1 formation, slightly enhanced after Gadolinium injection.

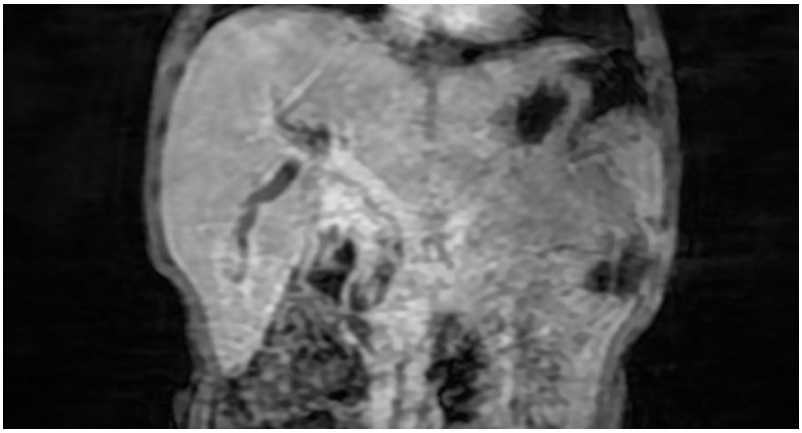

Figure 3

DISCUSSION

Biliary tract rhabdomyosarcoma is a rare disease often seen in young infants aged between 1 and 9 years old (median age being 3) with a slight male predominance.

The diagnosis should be made as soon as possible to increase chances of survival, as the survival prognosis is very poor. It is often revealed by a jaundice with an obstructive cholestasis syndrome. The biology may show and increased level of conjugated bilirubin, and slightly elevated hepatic enzymes. Imaging plays a decisive role in the rapidity of the diagnosis, but can also determine the extent of the tumor, so that an early and appropriated treatment can be started to maximize survival.

In most cases, the abdominal ultrasound is the first radiological examination to be conducted, and it should be performed by an experienced operator.

It typically shows a cystic image within the bile ducts which is seat of a solid tissue like material, vascularized on Doppler mode, as in the present case, which strongly suggested the possibility of a biliary tract tumor. Literature describes this network/lace-like appearance as characteristic of a biliary tract RMS. It can evaluate the extension to portal vein and other structures in the case of voluminous tumors and thus indicating or not a possibility of surgical resection.

The ultrasound being a relatively easy, non-invasive and available exam, it can be performed repeatedly, thus making it the first choice examination for follow-up evaluation under and after treatment to assess the decrease or increase in tumor size for patients under chemotherapy. There is no consensus for a sonographic surveillance rhythm.

CONCLUSION

Rhabdomyosarcoma (RMS) is the most common tumor of biliary tree in childhood. Biliary tree is an uncommon site for RMS to occur, accounting for about 1% of all pediatric RMS. The key to diagnosis is to distinguish solid component of the tumor from organized sludge seen in choledochal cyst. Imaging plays a vital role for preoperative staging as well.

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