CASE REPORT

EMBRYONAL RHABDOMYOSARCOMA OF BILIARY TRACT MASQUERADING AS HYDATID CYST/WORM INFESTATION

Muhammad Fasih Mansuri, Marya Hameed, Kanwal Laique, Ashraf Amir Ali, Syed Shariq Ullah

Department of Radiology, National Institute of Child Health (NICH), Karachi, Pakistan.

PJR January - March 2022; 32(1): 55-58

ABSTRACT

Rhabdomyosarcoma of childhood is a rare malignant soft tissue tumor of mesenchymal origin with an incidence of 4.5 cases per 1 million children. Obstructive jaundice is the most common presentation dueto this neoplastic biliary obstruction. Among it s histological subtypes embryonal subtype remains the most common with an incidence of around 54%. It commonly occurs in genitourinary tract and head and neck region with a rare occurrence in retroperitoneum and biliary tract. A 5-year-old child presented to us with obstructive jaundice and itching. On imaging, the peri-ampullary mass was misdiagnosed as biliary ascariasis/hydatid cyst; however, operative excision and histopathological correlation of the mass steered us towards the diagnosis of embryonal rhabdomyosarcoma. The patient was referred to the pediatric oncology department for chemotherapy and further follow-up. Despite being a rare entity, embryonal rhabdomyosarcoma of the biliary tract must be suspected in children presenting with obstructive jaundice and should get evaluation through multiple diagnostic tools in order to get accurate diagnosis and avoid a possible misdiagnosis.

Keywords: Embryonal rhabdomyosarcoma, biliary tract, hydatid cyst, obstructive jaundice, soft tissue tumor.

Introduction

Childhood rhabdomyosarcoma is a rare malignant soft tissue tumor of mesenchymal origin with only 50 cases reported in the literature.[1] Obstructive jaundice is the most common presentation due to this neoplastic biliary obstruction. It commonly occurs in genitourinary tract and head and neck locations with rare occurrence in retroperitoneum and biliary tract.[2] Among it s histological subtypes embryonal subtype remains the most common with an incidence of around 54%.[3]

We were presented with a case of embryonal rhabdomyosarcoma of biliary tract which was misdiagnosed initially as hydatid cyst/worm infestation, however; further work-up lead us towards the actual diagnosis of embryonal rhabdomyosarcoma. Our case report aims at highlighting the importance of multiple diagnostic tools in evaluating this condition which presents with a presenting complain that mimics with other hepatobiliary conditions in order to avoid a possible misdiagnosis.

Case Report

A 5-year-old male child presented to us in outpatient department with a complain of jaundice and itching for 3 months and 1 week respectively. On examination, the patient was very irritable with continuous itching and scratch marks on whole body. The liver was palpable 3cm below the right costal margin along with no evidence of ascites and splenomegaly.

Correspondence : Dr. Muhammad Fasih Mansuri
Department of Radiology,
National Institute of Child Health (NICH),
Karachi, Pakistan.
Email: fasih_mansuri@live.com

Submitted 20 December 2021, Accepted 29 January 2022

The liver function test showed elevation of alkaline phosphatase, Gamma GT, total and direct bilirubin levels. Viral markers including Anti HCV and HBsAG were negative. Ultrasound of the whole abdomen revealed dilated intrahepatic biliary duct giving the picture of obstructive jaundice. A solid mass with small cystic component is identified on epigastric region near porta hepatis measuring 4.4x3.8cm showing colour flow at periphery giving the picture of mass in healing of pancreas/duodenal mass. Further imaging through CT scan suggested moderate intra-hepatic duct dilatation with a possibility of worm infestation in extra-hepatic biliary system. ERCP showed external compression at ampulla causing bulky ampulla with ulceration at verge. A plan of surgery was made for the removal of mass with subsequent biopsy to rule out malignancy. The biopsy report of the peri-ampullary mass turned out to be positive for WT1 and Ki-67 with malignant spindle cells confirming the diagnosis of embryonal rhabdomyosarcoma. The patient was referred to pediatric oncology department for chemotherapy and further follow-up.

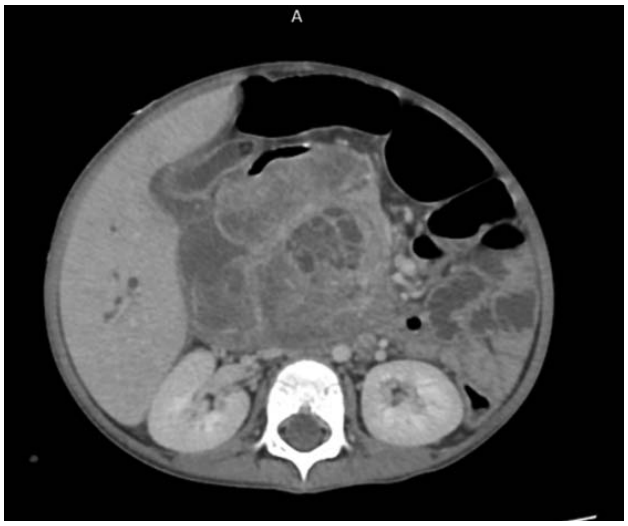

Figure 1: Showing heterogeneously enhancing mass with non-delineation of normal common hepatic and common bile duct.

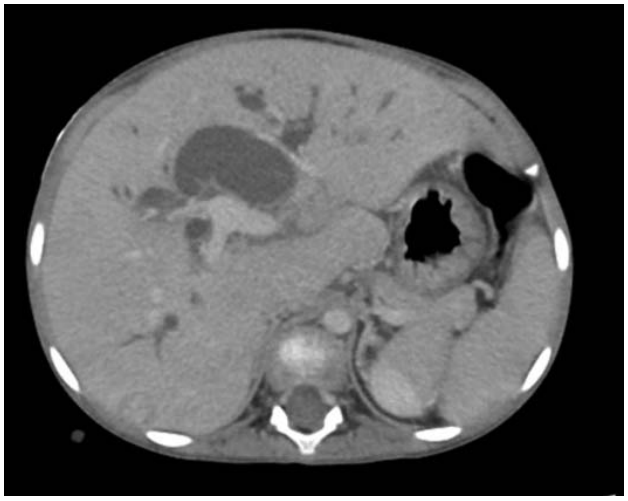

Figure 2: Showing cystic dilatation of common hepatic duct and intrahepatic biliary radicals

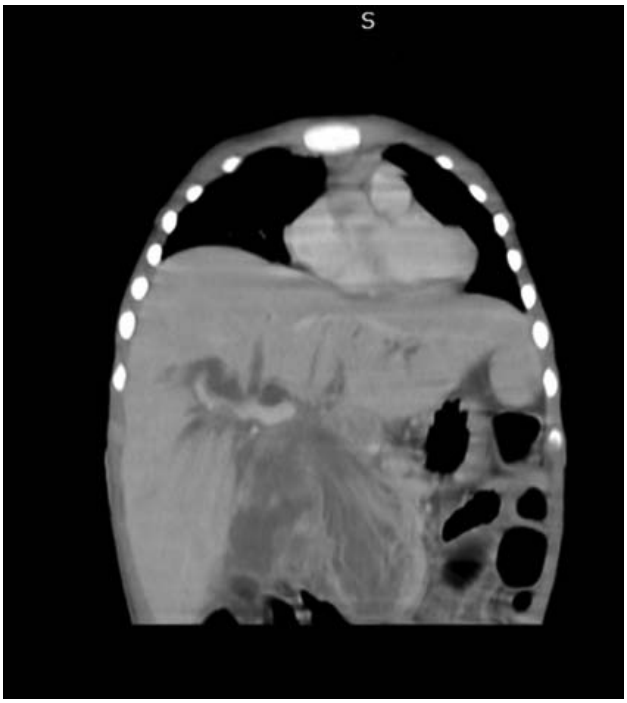

Figure 3: Showing infiltration of biliary rhabdomyosarcoma into adjacent structures.

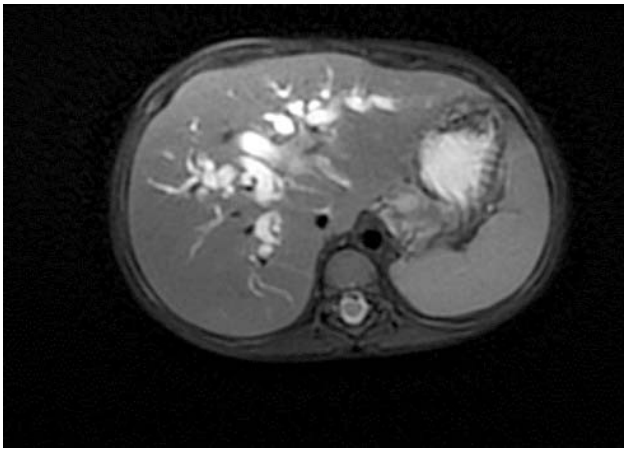

Figure 4: MRCP axial section reveals intrahepatic biliary dilatation secondary to mass effect.

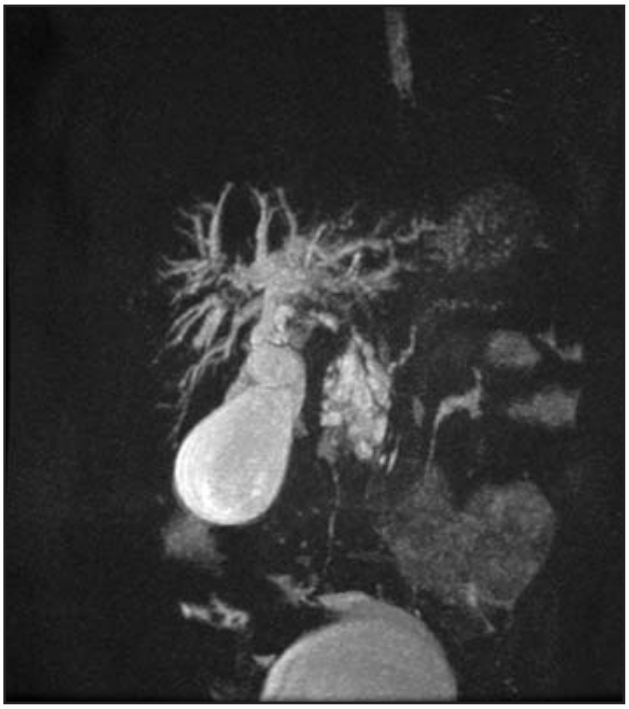

Figure 5: MIP image shows distended gall bladder and multicystic dilatation of common bile duct.

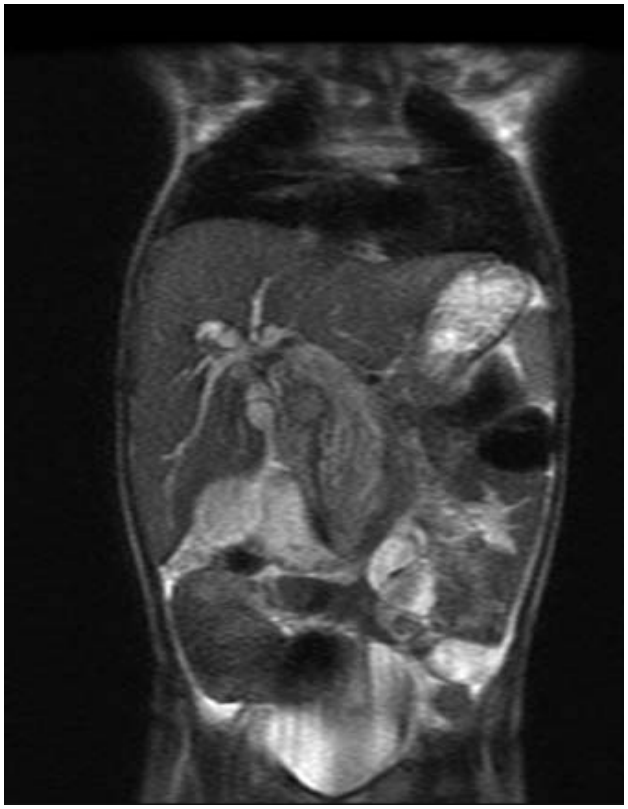

Figure 6: MRCP coronal T2 weighted image showing large heterogeneously enhancing mass responsible for voluminous common bile duct dilatation.

Discussion

Childhood rhabdomyosarcoma is a rare malignant soft tissue tumor of mesenchymal origin with obstructive jaundice is the most common presentation due to this neoplastic biliary obstruction. It commonly occurs in genitourinary tract and head and neck locations with rare occurrence in retroperitoneum and biliary tract.[1,2] Among it s histological subtypes embryonal subtype remains the most common with an incidence of around 54%.[3] It is slightly predominant in male population with an average age of occurrence of 3 years.[4] Apart from obstructive jaundice the other notable clinical symptoms include abdominal pain, fever acholic stools and decreased appetite.

The diagnosis of rhabdomyosarcoma is usually confirmed through surgery and biopsy of the mass. Thus, before histological examination, and on the basis of laboratory criteria and radiological examination rhabdomyosarcoma is often misdiagnosed as worm infestation as well as hydatid and Choledochal cysts. Nevertheless, imaging remains an integral part of the diagnostic work-up as it reveals the presence of a mass and ductal system dilatation.[5]

We presented a case of a 5-year-old boy who has symptoms of obstructive jaundice and itching which directed us towards imaging studies. On subsequent imaging studies and ERCP our diagnosis went in a direction of a hydatid cyst/worm infestation. Eventually, the surgery and histopathological examination of the mass steered us towards embryonal rhabdomyosarcoma.

The treatment of rhabdomyosarcoma remains multimodal. A multidrug chemotherapy and radiotherapy with IRS protocols along with second look surgery for residual or recurrent disease results in a long-term survival.[6]

Conclusion

Despite being a rare entity, embryonal rhabdomyosarcoma of the biliary tract must be suspected in children presenting with obstructive jaundice and should get evaluation through multiple diagnostic

tools in order to get accurate diagnosis and avoid a possible misdiagnosis, which along with multimodal treatment will eventually result in a long-term survival of the patient.

Conflict of Interest: None

References

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