Biliary Rhabdomyosarcoma in a 5-year Child - A Rare Case Report

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Abstract:
Jaundice in children is not an uncommon disease. However, paediatric jaundice is more often due to hepatic disease than biliary obstruction. The possible differential considerations for obstructive jaundice in children include choledocholithiasis, choledochal cysts and rare neoplasms. The most common soft tissue sarcoma in paediatric patients is rhabdomyosarcoma, typically involving the head and neck, genitourinary system and extremities but rarely occurs in biliary tree. We present a case of a 5-year-old boy with abrupt onset obstructive jaundice sent to our department. Initially USG and later MRCP was done and was diagnosed as rhabdomyosarcoma of hepatic duct confluence. Post-operative immunohistochemistry proved the case as a embryonal rhabdomyosarcoma (Botryoides sarcoma). This case illustrates the importance of considering malignant etiologies in cases of obstructive jaundice, particularly in toddlers.

Key words: *Biliary obstruction, choledochal cyst, obstructive jaundice, pediatric, rhabdomyosarcoma, hepatic duct confluence, MRCP.*

Introduction:

Obstructive jaundice in paediatric patients beyond the neonatal period can be caused by choledocholiths, choledochal cysts, strictures in the setting of chronic cholangitis and rarely neoplasms such as biliary rhabdomyosarcoma. Rhabdomyosarcoma, a malignancy of skeletal muscle, is the most common soft tissue tumor in the pediatric population, accounting for 1% of malignancies among children ages 0–14 years.[1] It is rarely found in the biliary tract, where it typically presents with signs and symptoms of obstructive jaundice. The low incidence of biliary rhabdomyosarcoma makes diagnosis challenging and imaging evaluation particularly important. We present a case of a child with embryonal rhabdomyosarcoma involving the hepatic duct confluence.

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Case History:

A 5-year-old boy presented to us with a history of jaundice, right upper abdominal pain and distension. He was constipated and his stools recently became pale. On general examination the boy was icteric, ill looking, temperature- 99p F, Pulse – 100/min, respiratory rate – 24/min. Local abdominal examination revealed mild tenderness over right upper quadrant. Blood picture revealed a total bilirubin level of 9.1 mg/dl and mostly conjugated; transaminases level mildly elevated; alkaline phosphatase and GGT markedly elevated; lipase was normal and white blood cell count was mildly elevated. In our radiology department, USG of abdomen was performed at first which revealed; one irregular inhomogeneous mass in the porta hepatis region measuring about 38 x 30 mm causing upstream biliary obstruction and non-visualization of common bile duct. Mild intra-lesional vascularity was evident on color Doppler. Subsequently MRCP was done and it showed – A mixed signal intensity mass at peri-portal region involving and compressing hepatic duct confluence resulting upstream biliary dilation. It was reported as a case of biliary rhabdomyosarcoma with biliary obstruction.

The patient was advised for surgical resection & he underwent laparotomy. Surgical excision of the mass was done & sent for histopathology. Histopathological report revealed embryonal rhabdomyosarcoma (Botryoides sarcoma). Immunohistochemistry was also done where it was also diagnosed as embryonal rhabdomyosarcoma (Botryoides sarcoma). The patient was discharged with a plan for neoadjuvant chemotherapy.

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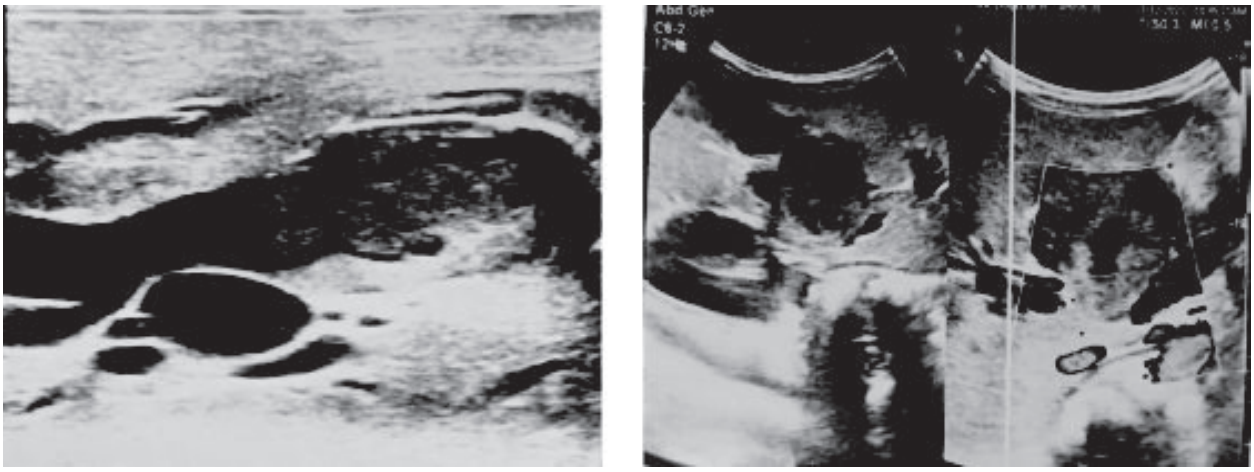

Fig.-1: *USG through upper abdomen showing a solid mass with internal necrosis in hepatic duct confluence. Biliary radicles are markedly dilated.*

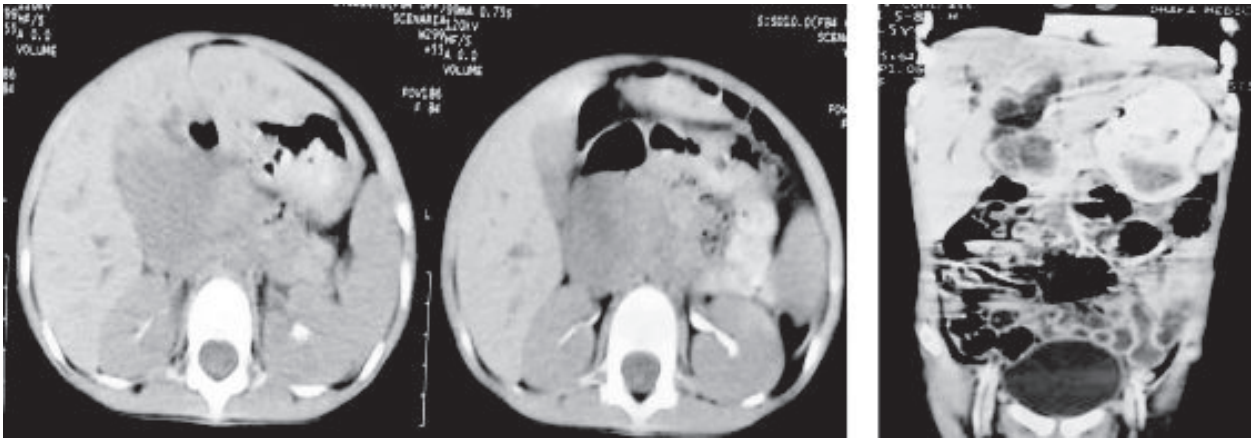

Fig 2: *Post contrast axial & coronal images of CT scan showing a large mass in hepatic duct confluence.*

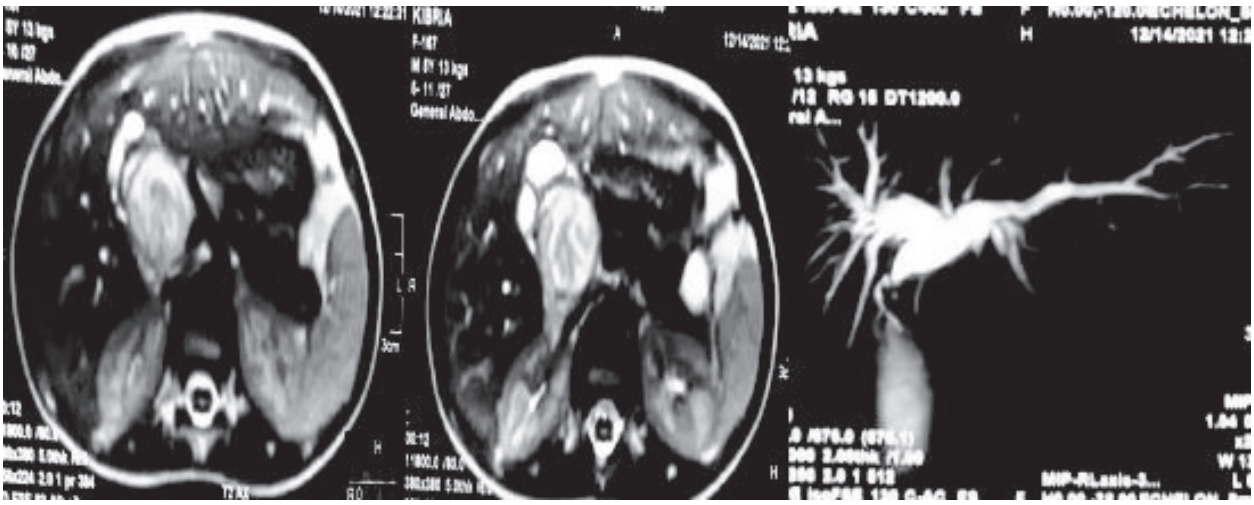

Fig 3: *T2WI axial MRI scan & MIP image of MRCP showing an irregular oval shaped mass at the confluence of hepatic ducts with intrahepatic biliary dilatation.*

Discussion:

Biliary obstruction in paediatric population is an uncommon disease. The differentials for obstructive jaundice in children includes choledolcolithiasis, choledochal cysts, strictures and rarely neoplasms. Imaging plays an important role in defining the etiology with ultrasound typically the first imaging modality.

Rhabdomyosarcoma is the most common soft tissue tumor in the paediatric population but sarcoma of the biliary tract is a rare cause of obstructive jaundice. Of all cases of rhabdomyosarcoma in the Intergroup Rhabdomyosarcoma Studies I-IV between 1972 and 1998, only 0.5% of cases (25 of 4291) involved the intrahepatic or extrahepatic biliary tree.[2,3]

Biliary rhabdomyosarcoma typically presents with obstructive jaundice caused by the tumor, which can be accompanied by abdominal distention and possibly hepatomegaly. Elevated liver enzymes and conjugated bilirubin are frequently seen. Infrequent features include abdominal pain, nausea, vomiting and fever.

Pathological subtypes of rhabdomyosarcoma include alveolar pleomorphic, and embryonal, which is further subdivided into Botryoid and spindle-cell variants. Embryonal type is three times more common than alveolar, and the Botryoid subtype has the best prognosis.[3,4] The Intergroup Rhabdomyosarcoma Study Group (IRSG) has developed a modified tumor-node-metastasis staging system that separates patients into low, medium and high-risk groups which are used to guide therapy.[4] The system considers site of tumor as well as regional nodal and distant metastases. In the absence of distance metastases, as seen in our case, biliary tract rhabdomyosarcoma is considered stage 1, placing our patient in the IRSG low risk group.

Ultrasound, typically the initial imaging study in children with obstructive jaundice, shows dilation of the intra- and extrahepatic bile ducts with an intra-ductal mass. On MRI, these tumors follow the signal of skeletal muscle on T1, are hyperintense on T2 and enhance heterogeneously. Large lesions may demonstrate hemorrhage and necrosis.[5] The imaging appearance of a biliary tree rhabdomyosarcoma with central tumor necrosis can mimic the appearance a choledochal cyst as was seen with this patient. Indeed, previous case reports have remarked that biliary rhabdomyosarcoma is readily misdiagnosed as a choledochal cyst on imaging.[6,7]

The surgical management of biliary rhabdomyosarcoma has historically been determined by tumor location and extent, ranging from aggressive resection necessitating choledocho-jejunostomy or pancreatico-duodenectomy to surgery for staging purposes only via exploratory laparotomy or laparoscopy. Presently, resection of residual tumor following neoadjuvant chemotherapy is advocated and has been associated with good outcomes.[8]

Conclusion:

Diagnostic imaging plays a critical role in the diagnosis and management of this malignancy. Rhabdomyosarcoma should be included in the differential diagnosis for children with obstructive jaundice.

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Conflicts of interest: There are no conflicts of interest.

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