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Extrahepatic bile duct fibroma in an infant: a case report

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Abstract

Background Extrahepatic bile duct tumor in neonates and infants is extremely rare. Fibroma in neonates and infants has been reported in different body parts but not in the bile duct. We report a 2-month-old girl with common bile duct (CBD) fibroma.

Case presentation A 2-month-old girl presented with jaundice and pale stool for 15 days. Her perinatal period was uneventful. On examination, we found the girl deeply icteric and having hepatomegaly but no palpable lump. Laboratory investigations revealed direct hyperbilirubinemia with elevated liver enzymes. Ultrasonography revealed hepatomegaly with focal dilatation of the extrahepatic bile duct (2.1 × 0.5 cm). Our preliminary diagnosis was a type I choledochal cyst. Considering the clinical scenario, we planned for exploration without further imaging. On exploration, we found the liver blackish and a hard mass involving the common CBD and cystic duct. The gall bladder was filled with clear mucus. After excising the mass, the gall bladder, and CBD, we reconstructed the anatomy with Roux-N-Y hepaticojejunostomy. The postoperative period was uneventful except for minor bile leakage during the first four postoperative days, which stopped spontaneously. Histopathology revealed spindle-shaped fibrous tissue admixed with collagenous tissue, compatible with a fibroma. A liver biopsy showed the features of cirrhosis. However, liver function tests were normal 2 weeks after surgery. The baby was thriving at 6 months of follow-up, hepatomegaly was resolved, and there was no clinical or radiological sign of recurrence.

Conclusions CBD fibroma is a rare cause of obstructive jaundice in neonates and infants. The earliest possible excision of the tumor and bile drainage restoration are necessary to halt the progression of liver damage.

Keywords Bile duct tumor, Bile duct fibroma, Obstructive jaundice in infants, Roux-N-Y hepaticojejunostomy

Background

Bile duct tumor in children is rare. The clinical presentation mimics other common pathologies of the bile duct in children. Therefore, it results in difficulty in diagnosis and surgical decision-making. There are reports of malignant bile duct tumors in children, mostly in teenagers [1]. Only two extrahepatic bile duct tumor cases have been reported in neonates and infants [2, 3]. A few cases have been reported in older children [2–4]. We report a case of bile duct fibroma in an infant.

Case presentation

A 2-month-old girl presented with jaundice and pale stool for 15 days. Her perinatal period was uneventful. On examination, we found the girl deeply icteric and having hepatomegaly but no palpable lump. Laboratory investigations revealed direct hyperbilirubinemia with elevated liver enzymes (total bilirubin 7.05 mg/dl, direct bilirubin 5.0 mg/dl, indirect bilirubin 2.05 mg/dl, SGPT 200U/L, alkaline phosphatase 558 U/L, gamma-glutamyl transferase (GGT) 273 U/L). Ultrasonography revealed

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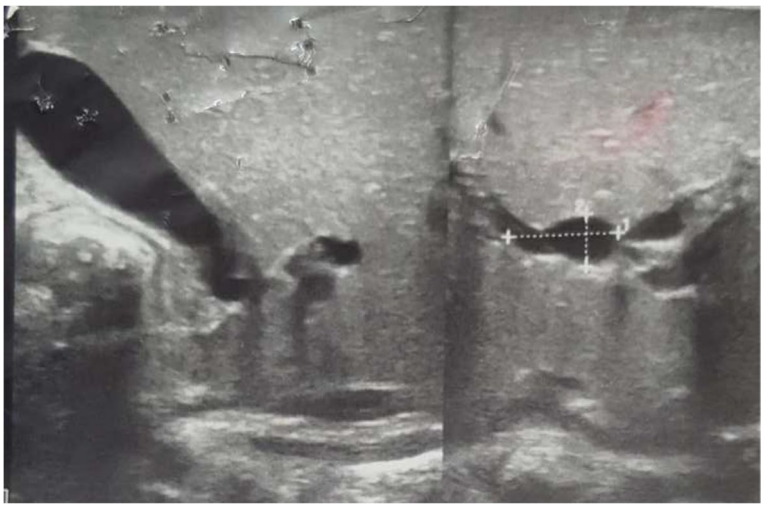

Fig. 1 Ultrasonogram of the hepatobiliary system. It shows the distended gall bladder and cystic dilation of the bile duct

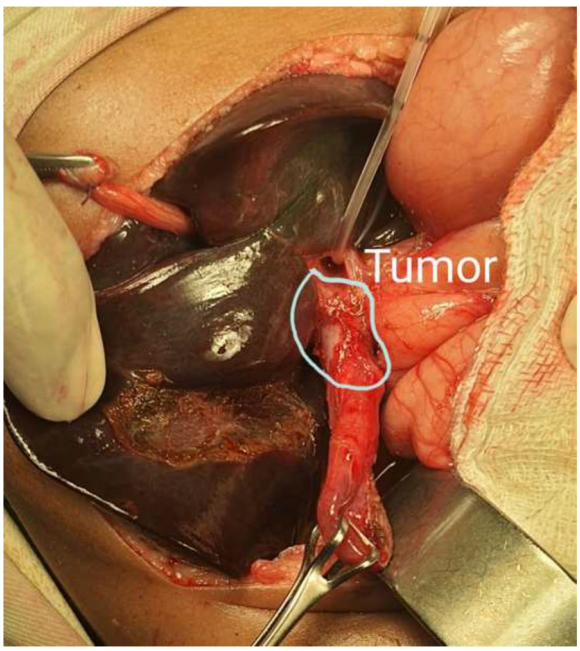

Fig. 2 Per-operative picture of the tumor. The liver is blackish. The tumor involved the common bile duct and the cystic duct

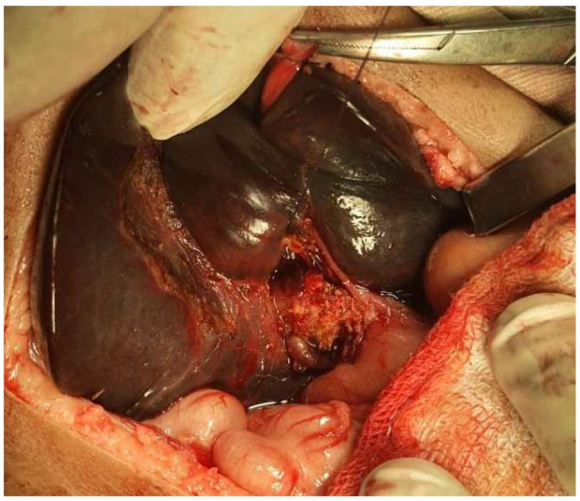

Fig. 3 Per-operative picture after tumor resection. The hepatic duct is mildly dilated

hepatomegaly with focal dilatation of the extrahepatic bile duct measuring 2.1×0.5 cm (Fig. 1), distended gall bladder, and mildly dilated intra-hepatic biliary channels. The pancreatic duct was not dilated. Our preliminary diagnosis was a type I choledochal cyst, and differentially, we thought about the cystic variety of biliary atresia. Considering the clinical scenario, we planned for exploration without further imaging. On exploration, we found the liver blackish and a hard mass involving the common CBD and cystic duct (Fig. 2). The gallbladder was distended and filled with clear mucus. Proximal to the mass, the common hepatic duct was mildly dilated (Fig. 3). After excising the mass, the gallbladder, and CBD, the residual common hepatic duct drained bile. We reconstructed the anatomy with Roux-N-Y hepaticojejunostomy. A drain tube was kept in the sub-hepatic space. The breastfeeding was resumed gradually from the second postoperative day. The postoperative period was uneventful except for minor bile leakage in the drain during the first four postoperative days. The postoperative stool color was yellow. The drain collection stopped spontaneously after 4 days. Histopathology revealed spindle-shaped fibrous tissue mixed with collagenous tissue, compatible with a fibroma (Fig. 4). A liver biopsy

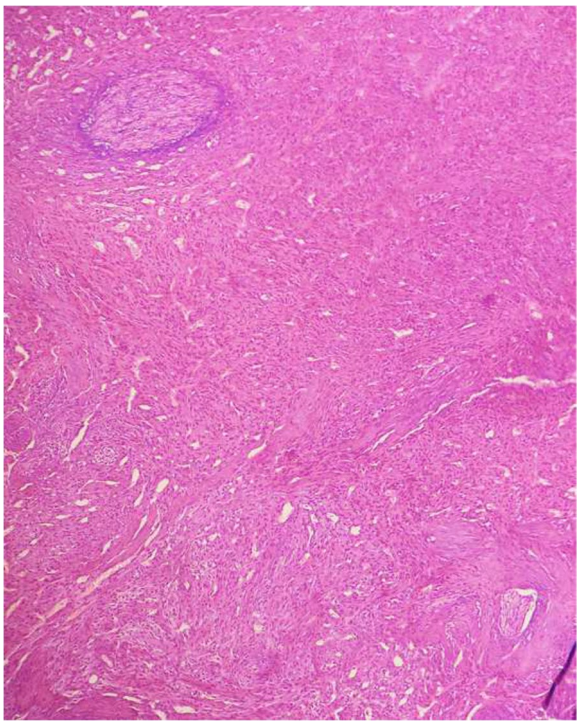

Fig. 4 Histopathology of the tumor. H&E stain, low-power image reveal uniform, bland spindle cells arranged in short intersecting fascicles and storiform pattern admixed with collagen

showed the features of cirrhosis. However, liver function tests were normal 2 weeks after surgery. The baby was thriving at 6 months of follow-up, hepatomegaly was resolved, and there was no clinical or radiological sign of recurrence.

Discussion

Tumor or tumor-like lesions are uncommon in children, especially young infants. Therefore, diagnosis could be difficult based on clinical presentation. Because of tumor position, clinical features mimic other common conditions with obstructive jaundice and cholangitis, e.g., choledochal cyst, and biliary ascariasis [2–6]. The preoperative diagnosis in our patient was a choledochal cyst.

Sonography detected the proximal bile duct dilatation but could not detect the mass in our patient. Similar sonographic findings have been reported in children with extrahepatic bile duct tumors; even a CT scan has missed the mass in a 10-year-old boy with bile duct tumor [2, 7]. This might be due to the unfamiliarity of these masses to the radiologist, which makes the preoperative diagnosis more confusing. However, a preoperative radiological delineation of the extrahepatic bile duct should be attempted for proper surgical planning. We perform an MRCP in most children when a USG indicates the presence of a choledochal cyst. However, in this particular patient, we decided to skip this investigation due to several reasons. Firstly, our hospital lacks the facilities to conduct an MRCP; patients must bear the cost of undergoing the procedure at private facilities. Additionally, the 2-month-old girl would require anesthesia outside our hospital, which could pose a risk. Thirdly, the investigation cost, which was higher than 150 USD, was not affordable for the parents.

Though rare, cholangiocarcinoma has been reported in older children. This malignant tumor has a poor prognosis in children [1]. Therefore, preoperative diagnosis, at least per-operative confirmation of the histopathological type is of utmost importance. Per-operative frozen section biopsy of the mass helps in decision-making for pre-operatively unidentified tumors [2, 7]. At our center, facilities for a frozen section biopsy were not available. Fortunately, we could excise the tumor completely, which later turned out to be a benign fibroma. We have re-established the bile flow with a Roux-N-Y hepatico-jejunostomy. A malignant mass would have required a more extensive resection and reconstruction.

Chung et al. reported 61 cases of infantile fibromatosis; 16 were multicentric. Most cases were noted at birth or shortly after birth. Head and neck was the most common site, followed by trunk and extremities. Only four children had visceral involvement, and three died within 48 h because of extensive multi-organ fibromatosis. The alive baby had fibromatosis in the colon. None of them had bile duct involvement. Three of the solitary lesions recurred [8]. Our patient had no evidence of multi-organ involvement.

A few cases of bile duct fibroma have been reported in adults. Fibromas of various sizes and shapes led to obstructive jaundice in those patients. This presentation is similar to our patient's. The morphology of bile duct fibromas was similar to fibromas elsewhere [9].

We are following the patient at 3-month intervals with liver function tests and sonography to detect recurrence early, if any. There is no sign of recurrence at 6 months. We advised the parents to do a half-yearly follow-up. One case has been reported to recur 1 year after bile duct teratoma excision at neonatal age [3]. Recurrence of childhood fibroma has been reported at 15 years of age [8].

The gross appearance of the liver and its histopathology showed early cirrhotic change. This might be due to cholestasis caused by the tumor. Therefore, early resection of the tumor and restoration of bile flow is recommended to avoid the progression of liver damage.

Conclusion

Bile duct fibroma in young infants is extremely rare, but the presentation mimics the common pathologies of the bile duct. So, it should be considered a differential diagnosis in infants presenting with jaundice and pale stools. Per-operative frozen section biopsy can differentiate it from malignant conditions and help in operative decision-making. Early resection and restoration of bile flow are empirical to prevent liver damage.

Abbreviations

CBD Common bile duct
CT Computed tomography
H&E Hematoxylin and eosin

Authors' contributions

Conceptualization: Dr. Md. Samiul Hasan and Dr. Ashrarur Rahman. Data collection/surgery: Dr. Md. Samiul Hasan, Dr. SM Nazmul Islam, and Dr. Refoyez Mahmud. Manuscript preparation/editing: Dr. Md. Samiul Hasan, Dr. Umama Huq, and Dr. Hasiba Mahshed Khan. Histopathological evaluation and review: Dr. Mashud Parvez. Manuscript review and critical appraisal: Dr. Mashud Parvez and Dr. Ashrarur Rahman.

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Availability of data and materials

All relevant information and documents have been provided in the manuscript. For further queries, the corresponding author will be available for explanation.

Declarations

Ethics approval and consent to participate

Ethical approval has been obtained from Bangladesh Shishu Hospital and the Institute's ethical review board (ERB).

Consent for publication

Informed written consent has been taken from the patient's parents.

Competing interests

The authors declare that they have no competing interests.

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