Unusual mesenchymal hamartoma with uncommon biliary anatomy: Case report and literature review

Mehdi Tahiri*, Noura Alhassan, Rita Chaouni, Alana Beres, Damian Maxwell, Pramod Puligandla

Montreal Children’s Hospital, Division of Pediatric Surgery, McGill University, Montreal, Quebec, Canada

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ABSTRACT

We report a case of a mesenchymal hamartoma of the liver in a twenty-month-old girl. The patient initially consulted her pediatrician, and was then referred to the emergency room for evaluation of a palpable upper midline abdominal mass. The initial referral questioned splenomegaly. Radiologic workup consisted of an abdominal ultrasound, followed by a magnetic resonance imaging (MRI) which showed a left hepatic lobe mass measuring 10 × 7 × 12 cm. The patient eventually underwent a left hepatectomy for excision of the mass. Postoperative course was complicated by a biliary leak secondary to an anatomic variant. The goal of this case report is to review the main features of liver hamartomas, and to discuss the possible anatomical variants of the biliary system that can be encountered in children.

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Primary tumors of the liver are uncommon in children. The most common presentation of pediatric liver neoplasm is an asymptomatic abdominal mass. Within these tumors about one third are benign and two third are malignant. Mesenchymal hamartomas represent one third of the benign neoplasm in the children [1].

1. Case report

We present the case of a 20-month-old girl previously healthy, originally from Mexico who presented to the emergency department of the Montreal Children’s Hospital. She was referred by her pediatrician for evaluation of the presence of a palpable upper midline mass on physical examination. She had poor oral intake and was not gaining weight. In the emergency department, an abdominal ultrasound was requested, which demonstrated a mass arising from the liver (Fig. 1). Following the ultrasound, surgical consultation was initiated. A magnetic resonance imaging (MRI) was then ordered to further assess the mass. The MRI did not include a magnetic resonance cholangiopancreatography (MRCP) to assess biliary anatomy. In view of the pattern of enhancement of the lesion hemangioma/hemangioendothelioma was initially suggested. The differential diagnosis included an atypical hepatoblastoma or a mesenchymal hamartoma (Fig. 2). A laboratory workup was ordered concomitantly which included liver function test exam, lactate dehydrogenase (LDH) and Alpha Fetoprotein (INR: 1.02, Albumin: 44 g/L, ALT: 17 IU/L, AST: 48 IU/L, Total Bilirubin: 14.5 μmol/L, LDH: 266 IU/L, Alpha Fetoprotein: 64.7 μg/L). After the initial workup, the working diagnosis was a hemangioma. Consultation with a pediatric radiologist with expertise in vascular anomalies was obtained and his opinion was that the mass was an atypical hepatoblastoma. The patient was presented at tumor board where further recommendations consisted of performing a PET scan. The hepatic mass was not metabolically active in the fluorodeoxyglucose (FDG)-PET. Given the uncertainty of the diagnosis, and the inability to completely rule out hepatoblastoma, as well as the patient’s failure to thrive, decision was made to proceed with an open left hepatectomy (Fig. 3). At this point our working diagnosis was a hemangioma, but an atypical hepatoblastoma and an atypical hamartoma could not be completely excluded.

The left hepatectomy was uneventful and post-operatively the patient was monitored in the pediatric intensive care unit (PICU).
She was transferred to the ward on post-operative day 1. Post-op course was complicated by a suspected pneumonia which responded to antibiotics. She was discharged home on post op day 10, tolerating a regular diet with her Jackson Pratt (JP) still in place with minimal non-bilious drainage. Pathology of the mass demonstrated a mesenchymal hamartoma of the liver.

At time of first post-operative clinic visit, one week post discharge, there was leakage noted around the JP and parents reported bilious output of approximately 100 ml/day. An ultrasound was ordered to evaluate for possible collection. Imaging showed post surgical changes with minimal fluid around the surgical bed and no significant collection.

At follow up 48 h later, the patients reported continued bilious drainage from the JP with decreased oral intake and some weight loss. The patient was admitted and an MRCP was ordered to assess the biliary tree and evaluate the source of the bile leak. The MRCP showed dilation of the right hepatic duct with abrupt diameter change at the level of the common bile duct (CBD) suspicious for obstruction. She then underwent insertion of a percutaneous catheter to drain the obstructed segment in interventional radiology. During cholangiography, a contrast leak was noted which drained out into the JP. The common bile duct could not be cannulated from the dilated segment. Interestingly, the patient’s stools were colored and the bilirubin not significantly elevated.

A cholangiogram was repeated and further attempts were made to enter the CBD, which were unsuccessful. Given the bile leak and her inability to tolerate oral intake, it was decided to take back the patient to the OR to explore the biliary tree. In the operating room the extra-hepatic biliary tree was carefully dissected free. The left duct had been divided during the first surgery, and the right duct was found to be intact and connected to the CBD. An on-table cholangiogram was performed with the assistance of an interventional radiologist. It was then discovered that the patient had an anatomical abnormality where the posterior right duct drained into the left hepatic duct. The dilated intrahepatic duct seen on US and MRI was actually the right posterior duct, which had inadvertently become obstructed when the left hepatic duct was divided at the original surgery. Patient underwent a hepatico-jejunostomy to the right posterior duct.

Post-operative course was uneventful. She was discharged home when tolerating a regular diet, with her trans-hepatic drain in place. It was eventually removed when a cholangiogram showed good drainage of contrast into the bowel with the drain having been clamped.
2. Discussion

2.1. Mesenchymal hamartoma

Hepatic mesenchymal hamartoma corresponds to hamartomatous growth of mesenchymal tissue, bile ducts and blood vessels of the liver, of uncertain etiology [2]. Mesenchymal hamartomas represent one third of benign hepatic neoplasms in children [1]. Eighty percent are discovered during the first two years of age and the remainder by age of 5 [3]. There is a slight male: female predominance of 1.5:1 [4].

Mesenchymal hamartomas usually present as an asymptomatic mass lesion. However, as it grows it has the potential to compress adjacent organs, resulting in various symptoms and complications, which include a wide spectrum ranging from respiratory distress to death. Approximately 75% of liver hamartomas occur in the right lobe of the liver [5]. Liver function tests and Alpha Fetoprotein (AFP) usually remain within normal range or can be mildly elevated [6]. Imaging studies will demonstrate a large, well circumscribed mass which may contain cyst of various sizes. On MRI, the solid part of the lesion will appear hypointense in both T1 and T2 phase compared to the adjacent normal liver, while the cystic part(s) will have a signal intensity similar to water on T2-weighted images [7].

Treatment consists of a complete surgical excision with clear margins. On pathologic evaluation gross cut sections will demonstrate multiple cysts containing yellow, serous fluid. Microscopically, hamartomas show mesenchymal and epithelial components. The myxomatous stroma contains epithelial and non-epithelial-lined cysts [8].

2.2. Common anatomic biliary anomalies

Anatomical variants of the biliary tract are not uncommon in the population and they can have important consequences if missed during a surgical procedure. Therefore, awareness of those variants is important when performing procedures involving the biliary tree in order to minimize complications [9].

The right hepatic duct drains the anterior (V and VIII) and posterior (VI and VII) segments of the right lobe of the liver through the right anterior, right posterior and the left hepatic ducts into the common hepatic duct, 3) A right anterior duct draining into the left hepatic duct, 4) A right posterior duct draining into the cystic duct and 5) Accessory hepatic ducts originating from the right or left ductal system.

3. Conclusion

3.1. Key learning points

- Hepatic hamartomas represent approximately one third of benign liver neoplasms in children. Their diagnosis can be misleading given the variable appearance of the mass on imaging studies. The treatment of hamartoma is complete resection.
- Anatomical variation of the right hepatic duct is common, and a good knowledge of the possible variants is important in pre-operative planning and to avoid potential post-operative complications.
- MRCP should be considered before any major hepatic resection given that normal anatomy is present only in 58–66% of the time.
- It is important to remember that hepatic hamartomas do not always fulfill their radiological and biochemical characteristics as described in our case report.

References