Idiopathic isolated benign non-traumatic inflammatory stricture of the common hepatic duct successfully treated with endoscopic dilatation in a child

J. Ferreira a,*, H. Bostwick b, S. Berezin b, A. Bokhari c

aDepartment of Pediatrics, Maria Fareri Children's Hospital at Westchester Medical Center, USA
bDepartment of Pediatric Gastroenterology, Maria Fareri Children's Hospital at Westchester Medical Center, 503 Grasslands Rd., Ste 201, Valhalla, NY 10595, USA
cDepartment of Pathology, New York Medical College at Westchester Medical Center, 100 Woods Rd., Valhalla, NY 10595, USA

A R T I C L E   I N F O

Article history:
Received 6 August 2013
Received in revised form 26 November 2013
Accepted 3 December 2013

Key words:
Idiopathic isolated benign non-traumatic biliary stricture
Langerhans cell histiocytosis
Primary sclerosing cholangitis
Endoscopic retrograde cholangiopancreatography (ERCP)

A B S T R A C T

Idiopathic isolated benign non-traumatic biliary strictures are a rare cause of obstructive jaundice in children. The treatment has been surgical with resection of the stricture and biliary-enteric anastomosis. We report a case of an idiopathic isolated benign non-traumatic inflammatory biliary stricture successfully treated with endoscopic dilatation in a 26-month-old male with persistent obstructive liver disease after surgical resection of an adjacent lymph node.

1. Case report

A previously healthy 26-month-old male presented to the Emergency Department with a 9-day history of dark-colored urine, pale stools, jaundice, abdominal pain and pruritus. Physical examination was remarkable only for jaundice and a palpable liver edge 1 cm below the right costal margin. His laboratory evaluation revealed abnormal liver chemistries and elevated cholesterol (Table 1, Day 1). The erythrocyte sedimentation rate was 35 mm/h, serologies for Hepatitis A, B and C, Cytomegalovirus, Epstein–Barr virus and toxoplasmosis were negative and values for anti-smooth muscle antibody, liver-kidney microsomal antibody titers, alpha 1 antitrypsin level and ceruloplasmin were normal. An abdominal ultrasound (U/S) demonstrated dilatation of the intrahepatic ducts. This finding of intrahepatic ductal dilatation up to the level of the common bile duct was also identified on abdominal CT scan. Consequently, the patient underwent endoscopic retrograde cholangiopancreatography (ERCP), which revealed a stricture at the level of the common hepatic duct bifurcation with proximal dilatation. Brushings for cytology were negative.

MRI of the abdomen demonstrated a 1.8 cm mass at the site of the stricture (Fig. 1). On hospital day 5 he was started on ursodiol...
15 mg/kg/day (Liver Chemistries in Table 1, day 5). The decision was made to perform exploratory laparotomy on day 9. This revealed a periportal mass which was completely resected. The main resected specimen was 2.5 × 2 × 1 cm. Histological evaluation showed that the lesions were lymph nodes with reactive hyperplasia and sinus histiocytosis (Fig. 2).

One month after surgery the patient’s bilirubin decreased, but the liver enzymes remained persistently elevated (Table 1, 1 month after surgery). Liver biopsy revealed non-specific portal fibrosis.

In follow-up over 2 additional months the patient’s liver chemistries did not improve (Table 1, 3 months after surgery). Another abdominal U/S demonstrated dilatation of the intrahepatic ducts. MRCP was performed and showed a markedly distended gallbladder with mild dilation of the intrahepatic ducts. A repeat ERCP demonstrated persistence of the stricture seen during the initial ERCP. The CHD stricture was dilated using a 4 mm balloon catheter.

The patient’s liver enzymes improved and then normalized after 10 months (Table 1, 2 weeks, 2 months and 10 months after ERCP). Follow up of the patient for 11 years including physical exams, liver enzymes and abdominal U/S has not demonstrated recurrence of the disease.

2. Discussion

Our patient had obstructive jaundice associated with a biliary tract stricture and a benign inflammatory lymph node in the porta hepatis. Obstructive jaundice has been described in children secondary to extrinsic compression of the biliary tree caused by benign mass lesions of infectious, inflammatory or idiopathic etiologies [1–3]. The lymph node pathology of our patient reported reactive lymphoid follicular hyperplasia and reactive sinus histiocytosis (RSH) (Fig. 2). The entities that should be considered in the differential diagnoses while interpreting a lymph node containing a prominent proliferation of histiocytes within nodal sinuses include RSH, Langerhans cell histiocytosis (LCH), and Rosai–Dorfman disease (also known as sinus histiocytosis with massive lymphadenopathy, SHML).

RSH is a common and self-limiting finding of enlarged lymph nodes characterized by proliferating histiocytes within the nodal sinuses, and these lymph nodes express CD68, but lack S100 protein and CD1a [6]; findings found in our patient.

LCH, a condition with aberrant proliferation of the langerhans cells, can involve lymph nodes and has been described as a cause of obstructive jaundice [3,7,8]. Our patient had absence of effacement of lymph node architecture, absence of histiocytes with Langerhans cells cytomorphology, and negative staining of histiocytes for CD1a and S100, criteria required for the diagnosis of LCH [3,7,8]. SHML, a rare non-malignant histiocyte proliferative disorder that classically presents as massive lymphadenopathy and has complete and spontaneous resolution in most patients, is histologically characterized by nodal emperipolesis, capsular and pericapsular inflammation and fibrosis, and + S100 staining of histiocytes [9]; all absent in our patient.

Although our patient’s liver chemistries did not improve, the jaundice resolved after resection of the mass; this suggests that the lymph node was a factor in the obstructive process, but was not the sole cause. The obstructive liver disease did not resolve until dilatation of the biliary stricture. The etiology of this stricture remains unclear. It does seem possible that the stricture occurred on the basis of inflammation or scarring due to a localized inflammatory process related to the enlarged lymph node. However, the lymph node may have been secondary to a primary inflammatory process of the biliary tract.

Acquired benign biliary strictures in children can be caused by cholelithiasis, choledocholithiasis, chronic pancreatitis, abdominal trauma andiatrogenic injury after cholecystectomy or biliary tract procedures [4]; none of which were present in our patient. Other

---

**Table 1 Clinical data.**

<table>
<thead>
<tr>
<th></th>
<th>Day 1</th>
<th>Day 5</th>
<th>1 month after surgery</th>
<th>3 months after surgery</th>
<th>2 weeks after ERCP</th>
<th>2 months after ERCP</th>
<th>10 months after ERCP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Bilirubin (Mg/dl)</td>
<td>8.4</td>
<td>10</td>
<td>1.5</td>
<td>1.7</td>
<td>1.5</td>
<td>0.1</td>
<td>0.2</td>
</tr>
<tr>
<td>Direct Bilirubin (Mg/dl)</td>
<td>5.1</td>
<td>6.3</td>
<td>0.8</td>
<td>1.2</td>
<td>0.9</td>
<td>&lt;0.1</td>
<td>&lt;0.1</td>
</tr>
<tr>
<td>AST (U/L)</td>
<td>498</td>
<td>440</td>
<td>368</td>
<td>382</td>
<td>144</td>
<td>82</td>
<td>28</td>
</tr>
<tr>
<td>ALT (U/L)</td>
<td>506</td>
<td>502</td>
<td>362</td>
<td>338</td>
<td>142</td>
<td>66</td>
<td>29</td>
</tr>
<tr>
<td>GGT (U/L)</td>
<td>602</td>
<td>643</td>
<td>872</td>
<td>814</td>
<td>420</td>
<td>95</td>
<td>11</td>
</tr>
<tr>
<td>ALP (U/L)</td>
<td>2200</td>
<td>&gt;1980</td>
<td>&gt;1980</td>
<td>&gt;1980</td>
<td>1866</td>
<td>775</td>
<td>239</td>
</tr>
<tr>
<td>Cholesterol (Mg/dl)</td>
<td>779</td>
<td>934</td>
<td>1059</td>
<td>998</td>
<td>428</td>
<td>188</td>
<td>183</td>
</tr>
</tbody>
</table>

AST, aspartate aminotransferase; ALT, alanine aminotransferase; GGT, gamma-glutamyl transpeptidase; ALP, alkaline phosphatase.

---

**Fig. 1.** Preoperative MRI (A) precoronal cut demonstrating a 1.8 cm soft tissue mass at the porta hepatis area (arrow). (B) Intrahepatic bile duct dilation (arrows).
causes such as primary sclerosing cholangitis and idiopathic isolated benign inflammatory strictures must be considered [3–5].

Primary sclerosing cholangitis (PSC) is characterized by inflammation and obliterative fibrosis of the intra- and extrahepatic bile ducts. No effective medical therapy aimed at preventing disease progression is currently available and the disease usually leads to biliary cirrhosis and liver failure [10,11]. Strictureing and diffuse irregularities (“beaded” appearance) of the extrahepatic biliary system are found on imaging studies [12,13], and the pathognomonic finding of concentric periductular fibrosis on liver biopsy are diagnostic [14]. In a long-term follow up study of 52 pediatric patients with PSC demonstrated that the median (50%) survival free of liver transplantation was 12.7 years [10].

We do not believe that our patient had PSC. Even though he was found to have a stricture involving the CBD, the fact that this patient has no recurrence of liver disease for over 10 years argues against this condition.

Previous reports demonstrate that isolated, benign, non-traumatic inflammatory bile duct strictures of unknown etiology can occur in young children. In these reports the management of all patients involved laparotomy with biliary-enteric anastomosis or biliary reconstruction with resection of the abnormality of the extrahepatic biliary tree. The diagnosis of inflammatory stricture was based on histopathologic examination findings of partial or complete biliary epithelial denudation, chronic inflammation and mural fibrosis [3–5]. Our patient had an excellent outcome with ERCP balloon catheter dilation and therefore did not require surgery. As such we could not make a pathologic diagnosis. An alternative approach to the management of this patient could have been surgical excision of the periportal mass followed by an intraoperative cholangiogram and excision of the strictured segment with a Roux-en-Y hepaticojejunostomy in the same procedure.

Advantages of this approach over our patient’s approach include single exposure to anesthesia and definitive histological diagnosis, however surgical hepaticojejunosotmy does expose the patient to postoperative complications such as persistence of elevated liver enzymes [4], bile leak, cholangitis, anastomotic stricture, adhesive intestinal obstruction and need for a second intervention [15,16]. While ERCP does have complications such as pancreatitis, cholecystitis, cholangitis, perforation and gastrointestinal bleeding; the majority of untoward events are of mild-to-moderate severity and generally experience in the short term [17].

3. Conclusion

This case report demonstrates that a 26-month-old patient with an idiopathic isolated benign non-traumatic inflammatory stricture can be treated successfully with ERCP balloon dilation alone. This should only be done after any progressive, neoplastic or inflammatory disorder has definitely been excluded. Previous reports describe treatment to be extensive surgery. We conclude that ERCP balloon dilatation should be considered as the initial treatment for benign biliary strictures.

Conflict of interest
No authors have any disclosure.

Sources of funding
There was no source of funding for this publication.

References