Rare case of gallbladder papiloma in children

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**Abstract**

The incidence of villous papilloma of the gallbladder is rarely found amongst children. We report on a 9-year-old girl known case of metachromatic leukodystrophy presented with abdominal distension. A radiological examination revealed massive ascites and a papillary mass branching and projecting through the gallbladder fundus. Furthermore, a histopathological examination of the gallbladder showed villous papilloma with focal high-grade dysplasia and a single gallbladder stone. This report describes the rare presentation of a child with metachromatic leukodystrophy in the form of gallbladder polyp, intractable ascites, and a gallbladder stone.

1. Background

The etiology and pathogenesis of gallbladder papilloma in children are not thoroughly addressed in the literature due to a limited number of reported cases [1]. To our knowledge, there are only three pediatric cases reported in the literature [1–3] that are similar to the current case. Consequently, there is very little experience in managing this disease in children. Gallbladder polyps are classified as tumorous and non-tumorous. The tumorous group may be adenomas, adenomyomas, and early gallbladder carcinoma. Non-tumorous polyps are either inflammatory or of the cholesterol type [4], with cholesterol lesion accounting for the majority of the gallbladder polyps [5].

2. Case report

We present a nine-year-old girl who showed normal development until the age of five, when she began to experience deterioration in her developmental milestones characterized by impaired speech that further progressed to an inability to speak, weakness in her extremities and seizures resulting in a spastic quadriplegia state. She presented to our institute with marked abdominal distension, on examination, she was vitally stable, bedridden and spastic quadriplegic with massive ascites.

Laboratory results revealed WBCS 8.2, HB% 8.1, PROTEIN 60 g/l, ALBUMINE 34 g/l, BILIRUBIN 2.6 umol/l, ALP 245 u/l, AMYLASE 87 u/l, lipase 163 u/l, CRP 19.1 mg/dl, CA 19-9 1433.7 iu/ml, LDH 259 μl, CEA 3.85 mg/l. An abdominal ultrasound (U/S) showed a gallbladder fossa mass measuring 4.11 × 4.24 cm with massive ascites. An abdominal MRI revealed a dilated gallbladder with a heterogeneous mass that had papillary like projections measuring 4 × 4.4 × 4.1 cm. The MRI also confirmed the presence of massive ascites with no evidence of hepatic invasion or biliary dilatation. The gallbladder mass was suspected to have breeched the fundus (Fig. 1), which was highly suggestive of malignancy (rhabdomyosarcoma).

Doppler U/S showed that the hepatic and portal veins were patent with a good blood flow, and there was no evidence of thrombosis or invasion. Due to the high index of malignant suspicion, we elected to biopsy the mass rather than perform other definitive procedures (i.e., mass excision).

Under U/S guidance, the mass was biopsied and a pigtail was inserted for ascetic taping. In addition, the patient received furosemide and spironolactone until the ascites subsided. However, ascites reoccurred within days upon removal of the pigtail and discontinuance of diuretic medication.

The ascetic fluid analysis revealed a yellow color fluid, hazy in appearance with WBCs 108 cells/ul, RBCs 4 cells/ul, neutrophil 12%, lymphocytes 12%, macrophages 76%; bilirubin 5.06 umol/l, amylase 22 u/l, PH 8 and specific gravity 1.010. The result of calculating serum-ascites albumin gradient (SAAG) was 14 g/l. Results of analysis of the ascetic fluids for organisms were negative, and Acid Fast Bacilli (AFP)

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at 6 week incubation revealed no growth. Peritoneal fluid cytology with immunohistochemical stains verified that there were reactive mesothelial cells, and there were no carcinoma or lymphoma cells. Gallbladder mass FNA results showed benign glandular epithelial cells, and no evidence of malignant cells.

We decided to proceed for laparoscopic exploration, as the findings of the investigations were inconclusive; and there was no clear histological confirmation, as well as no other probable cause for ascites other than the gallbladder lesion. Intraoperatively, the abdomen was found full of off-white gelatinous fluid with patches of gelatinous material on the peritoneal surface. Moreover, the gallbladder was perforated near the fundus with multiple delicate, tendril-like fronds of tan tissue protruding through the wall (Fig. 2). Cholecystectomy with gallbladder bed resection was successfully completed, with an uneventful postoperative course.

Gross pathological examination showed the gallbladder and part of the liver measuring $7 \times 6 \times 3$ cm with a polypoid papillary mass measuring $4.5 \times 4 \times 2$ cm emanating from the neck and body of the gallbladder, and protruding through its wall. In addition, a green stone measuring 0.8 cm in diameter was discovered. The histological diagnosis was villous papilloma with focal high-grade dysplasia (Fig. 3), and there was no evidence of invasive carcinoma. The gallbladder bed liver tissues were normal. During a one-year follow up, the patient remained symptoms-free without reappearance of ascites or abdominal complaints.

3. Discussion

Papillomatosis of the gallbladder may be a primary disease without clear or specific underlying causes; or it may be secondary to a systemic disease such as metachromatic leukodystrophy (MLD), Peuts-Jegher syndrome or pancreatobiliary malunion [6]. Metachromatic leukodystrophy is an autosomal recessively inherited lysosomal storage disorder caused by a deficiency of the enzyme arylsulphatase that leads to an accumulation of sulphatide in the nervous system and in certain visceral organs including the gallbladder [7]. An increased level of sulphatide excreted into the bile induces a proliferative and metaplastic epithelial response within the gallbladder. A gallbladder involved in MLD is described as shrunken and fibrotic [1,8], as well as various other forms such as thickening of gallbladder wall, sludge, gallstone, hemobilia, cholecystitis, polyposis and Papillomatosis [2,7]. In our patient, MLD involving the gallbladder manifested in the form of villous papilloma with focal high-grade dysplasia and a gallbladder stone.

CA 19-9 is a known tumor marker in a variety of malignancies in adult patients, predominantly in pancreatic cancer. Although CA 19-9 is not normally elevated in nonmalignant diseases,
Elevated levels of CA 19-9 has been found in several benign conditions such as inflammation or proliferation of noncancerous gastrointestinal, hepatobiliary and splenic tissue [9]. This may explain the elevated level of carbohydrate antigen 19-9 (CA 19-9) found in our patient.

In many underdeveloped, and some developing countries, infections such as TB are the most frequent causes of ascites after the newborn period, whereas hepatobiliary disease (e.g. storage disease) is a common cause in developed countries. Our patient’s presenting symptom was abdominal distension due to massive ascites. As the cause of the ascites was not evident and the diagnosis of the gallbladder mass was uncertain, two low-risk procedures (i.e., U/S guided needle biopsy and paracentesis) [10] were performed to obtain diagnostic information, as well as, to provide immediate relief of the patient symptoms related to massive ascites. Investigations showed neither indication of infectious, nutritional, cardiac, renal, nor liver diseases. According to the findings of previous studies when the calculating SAAG >1.1 g/dl, signified that the cause ascites were due at least in part to an increase in portal pressure with 97% accuracy [11]. However, in our patient this is was not the case, as the Doppler U/S study showed no evidence of portal hypertension. Consequently, the most probable cause of ascites was peritoneal irritation due to the accumulation of glandular epithelial cells and metachromatic material excreted from the gallbladder lesion into the peritoneum cavity.

A review of the literature revealed that this is the first case reporting metachromatic leukodystrophy with gallbladder villous papilloma presenting with massive ascites in children.

References