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Pediatric paraduodenal dermoid cyst: Clinical presentation, minimally invasive management and literature review

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ABSTRACT

Dermoid cysts (mature cystic teratomas) are congenital masses composed of all three germ cell layers: commonly occurring in the head, neck, and gonads and rarely in the abdomen. We present the first documented case of a paraduodenal dermoid cyst in a child, and describe the minimally invasive surgical approach utilized for resection. The patient was an asymptomatic five-month old female diagnosed with a cystic lesion in the posterior mid-abdomen on a prenatal ultrasound, followed up by MRI at three months of age. We proceeded with a laparoscopic resection for both diagnosis and definitive management. Intraoperatively, the lesion was noted to be separate of the pancreas and the stomach, located between the superior mesenteric vessels and splenic vein. It was dissected out with histopathology confirming the diagnosis of a dermoid cyst. We believe that the use of a laparoscopic technique allowed for better post-operative pain control, less overall morbidity and a shorter hospital stay compared to an open approach. Given the significant recurrence rate and possibility of malignant degeneration with incomplete resection, it is imperative to perform a complete resection if this lesion is suspected.

Dermoid cysts (mature cystic teratomas) are slow-growing congenital masses composed of all three germ cell layers: endoderm, ectoderm and mesoderm. They commonly occur in the region of the head and neck as well as in the gonads, but rarely present in the abdomen as an extra-gonadal mass. In children, there are reported cases of ileocecal mesenteric dermoid cysts [1,2], pancreatic dermoid cysts [3], as well as a case report of a splenic dermoid cyst [4]. However, there are no previously reported cases of dermoid cysts associated with the duodenum. We report a case of a paraduodenal dermoid cyst, separate of the pancreas, ovary or intestines, and describe the minimally invasive surgical approach utilized for resection.

1. Case report

A five month-old female with a prenatal diagnosis of an abdominal cyst was brought in for surgical evaluation. The infant was in good overall health, feeding and developing appropriately without vomiting or anorexia. Physical exam was unremarkable, with no palpable abdominal mass, distension or ascites. Postnatal abdominal ultrasound completed at three months of age re-demonstrated the mass, and she underwent an abdominal magnetic resonance imaging (MRI) scan for further characterization. The MRI showed a 0.8 × 0.9 × 1.7 cm cystic structure in the posterior mid-abdomen consistent with a duodenal duplication cyst or a benign pancreatic cyst and no other intraabdominal pathology (Figs. 1 and 2). Given this differential diagnosis, no additional workup was performed and the decision made to perform a laparoscopic resection of the cyst.

In the operating room, the patient was placed in supine position and pneumoperitoneum was induced via a 4 mm umbilical trocar. Two additional 3 mm trocars were placed in the right and left upper

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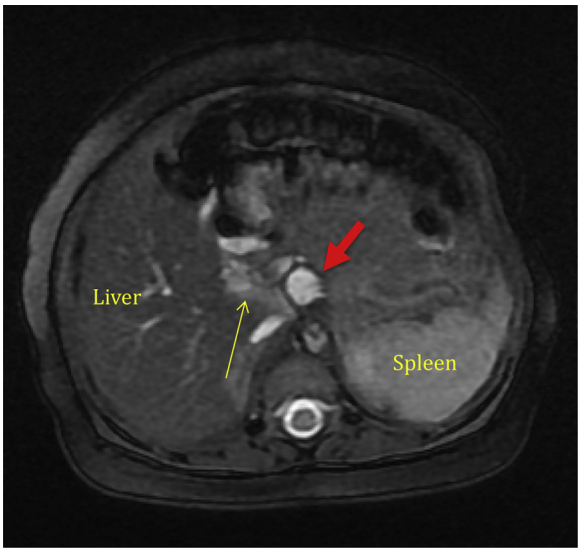

Fig. 1. Axial T2 weighted MRI of the upper abdomen. The red arrow points to the dermoid cyst, immediately posterior to the SMV and SMA. The yellow arrow points to the duodenum.

quadrants. The transverse colon was reflected anteriorly and the cyst was visualized at the root of the mesentery, adjacent to the duodenum. A Kocher maneuver was then performed and it was determined that the cyst was not originating from the first or second portion of the duodenum. The lesser sac was accessed and the retroperitoneum was exposed. The cyst was identified to be separate from the duodenum, pancreas and the stomach, located between the superior mesenteric vessels and splenic vein. It was dissected free from surrounding structures using a 3 mm curved tip electrosurgical sealing device and removed through the umbilical

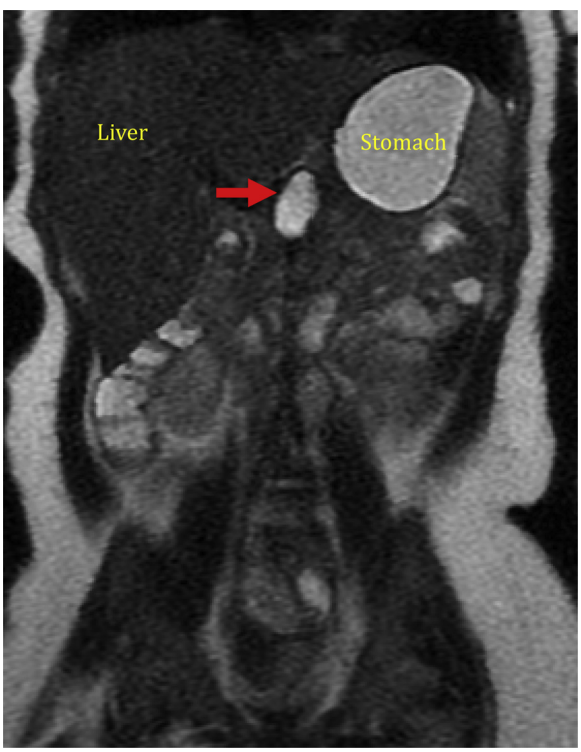

Fig. 2. Coronal T2 weighted MRI. The red arrow points to the dermoid cyst.

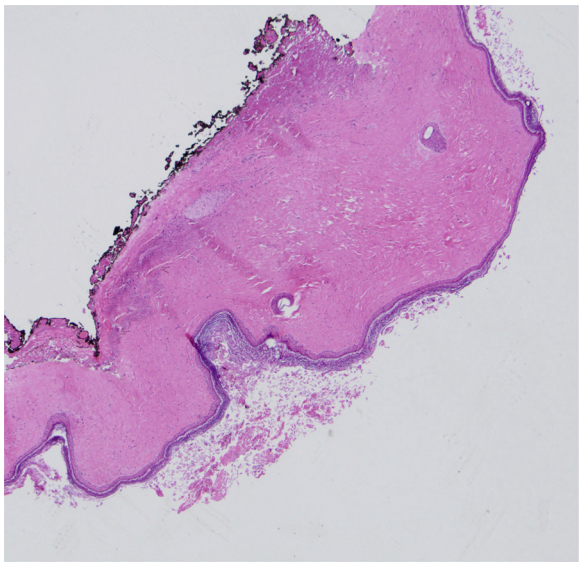

Fig. 3. Low magnification photomicrograph of dermoid cyst. Two circular skin appendages noted within cyst wall.

port site incision intact and without difficulty. The port sites were closed in standard fashion with absorbable suture. The infant recovered well from the procedure and was discharged home two days later.

Pathologic examination of the cyst revealed a pale pink cyst measuring 2.3 × 1.4 × 0.8 cm (Fig. 3). The specimen was bisected to reveal white cheesy material. Sections of the specimen showed a cyst with squamous epithelium and overlying keratin with fibrotic subepithelial tissue and hair appendages (Fig. 4). These histopathological findings were consistent with a dermoid cyst.

2. Discussion

Dermoid cysts are rarely found in the extra-gonadal abdomen and this is the first documented case of a pediatric paraduodenal dermoid cyst. Dermoid cysts are slow growing, benign tumors that present as an asymptomatic palpable mass or with a myriad of symptoms when they impinge on surrounding structures. They derive from remnants of totipotent cells, although the exact pathogenesis remains unknown. Histologically, they appear similar to

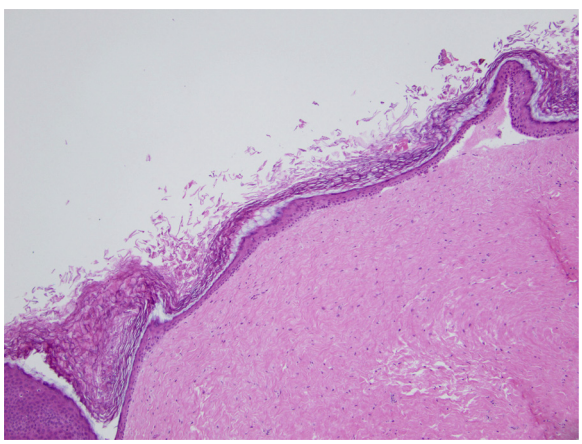

Fig. 4. Higher magnification photomicrograph demonstrating fibrotic cyst wall surrounded by squamous epithelium and overlying keratin.

epidermal inclusion cysts because they contain keratinized, stratified squamous epithelium, but are distinguished by the presence of skin appendages/dermal structures, including sebaceous glands, sweat glands and hair follicles.

Unlike previously documented cysts in the upper abdomen, this cyst did not involve the pancreas or the spleen. Given the location adjacent to the duodenum, the preoperative differential diagnosis in this pediatric age group included duplication cysts and mesenteric cysts. As our case demonstrated, these lesions may be indistinguishable despite appropriate workup with preoperative ultrasound and MRI. In adults with pancreatic dermoid cysts, endoscopic ultrasound (EUS) has been used in some cases to aid in the diagnosis of these lesions [5]. While the location of this dermoid cyst may have been amenable to EUS, in children, these lesions are more likely to be misdiagnosed as duplication cysts rather than malignant cystic neoplasms that are found in adults. Similar to duplication cysts, the definitive treatment for dermoid cysts is complete surgical resection, thus EUS would be unlikely to change the course of treatment.

Minimally invasive approaches have been used successfully in the excision of other pediatric mesenteric dermoid cysts [1]. We believe that the use of laparoscopy allowed for better post-operative pain control, less overall morbidity and a shorter inpatient hospital stay compared to an open approach. In this case, the dermoid cyst did not involve the second portion of the duodenum and was able to be dissected free from nearby major vessels, obviating the need for a more extensive surgery.

It is difficult to ascertain the recurrence rate of abdominal dermoid cysts in children due to the rarity of the lesions. One case report documented a recurrent pancreatic dermoid cyst fifteen months after initial resection, although this may have been due to incomplete resection at the primary surgery due to its large size and location in the body of the pancreas [3]. In a retrospective review of pediatric ovarian dermoid cysts, 11% of patients were found to have recurrent or persistent cysts within five years [6]. Additionally, there have been a few reported cases of malignant degeneration of dermoid cysts in adults [7–9], although none in the pediatric literature.

As this was the first case of a paraduodenal dermoid cyst in the pediatric literature, there are no established guidelines for post-surgical follow up imaging. As the cyst was resected intact in its entirety and was not associated with any abdominal structures such as the pancreas, ovary or intestines, theoretically there is minimal risk of recurrence. Given the slow-growing nature of the tumor, we plan to repeat an abdominal ultrasound at one year to reassess for recurrence and continue routine wellness visits with her pediatrician thereafter.

3. Conclusion

Dermoid cysts in the upper abdomen are rare and this is the first documented case of a pediatric paraduodenal dermoid cyst. Clinical presentation and preoperative imaging studies may be nonspecific for this lesion. Given the significant recurrence rate and possibility of malignant degeneration if incompletely resected, it is important for surgeons to perform a complete resection if this lesion is suspected.

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