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Boerhaave's Syndrome

Spontaneous rupture of the esophagus after a sudden increase in intraluminal pressure is known as Boerhaave's syndrome. The sudden increase in intraluminal pressure is most commonly caused by vomiting, but other causes of barogenic rupture include coughing, childbirth, defecation, seizures and blunt abdominal trauma. Spontaneous transmural esophageal rupture can also be seen as a primary manifestation of eosinophilic esophagitis preceded by an episode of food impaction with induced vomiting. In adults, the esophageal rupture is usually longitudinal, from 2 to 6 cm long, involves the distal thoracic esophagus usually toward the left into the left pleura. Rupture of the cervical, middle third or abdominal segment of the esophagus is rare. In most children the rupture in the distal esophagus is on the right side into the right pleural cavity. Sudden chest pain after the exerting pressure is the most common symptom. Half cases have the triad of forceful vomiting, mild hematemesis and substernal chest pain. With esophageal rupture and mediastinitis the child develops tachycardia, diaphoresis, fever and hypotension. Simple chest films reveal effusion, pneumothorax, hydropneumothorax, and subcutaneous emphysema. The diagnosis of Boerhaave's syndrome is confirmed with a simple water-soluble oral contrast study of the esophagus or CT-Scan of the chest. The esophageal tear may be diagnosed with esophagoscopy, but insufflation with high pressure may worsen pneumothorax or pneumomediastinum and cause life-threatening tension pneumothorax. Delaying the diagnosis of Boerhaave's syndrome increases the mortality due to mediastinitis. Initial management consist of antibiotics and chest tube drainage until the diagnosis is confirmed. Conservative management may be applied to children with small defects, contamination limited to the mediastinum and late diagnosis (> 24 hours after symptoms). Most patient will need an operation as treatment of choice which may consist of primary closure of the perforation, partial resection of the esophagus, drainage, or intraluminal stent. Primary closure can be covered with a pleural patch or fundoplication over the defect.

References:

1- Ramsook C: Boerhaave's syndrome: a pediatric case. J Clin Gastroenterol. 33(1):77-8, 2001
2- Korn O, Onate JC, Lopez R: Anatomy of the Boerhaave syndrome. Surgery. 141(2):222-8, 2007
3- Antonis JH, Poeze M, Van Heurn LW: Boerhaave's syndrome in children: a case report and review of the literature. J Pediatr Surg. 41(9):1620-3, 2006
4- Vernon N, Mohananey D, Ghetmiri E, Ghaffari G: Esophageal rupture as a primary manifestation in eosinophilic esophagitis. Case Rep Med. 2014;2014:673189. doi: 10.1155/2014/673189. Epub 2014 May 11.
5- Sanka S, Gomez A, Heuschkel R, Krishnamurthy K: Boerhaave's syndrome: a differential diagnosis of acute chest pain following a vomiting illness. West Indian Med J. 62(2):152-3, 2013
6- Donnet C, Destombe S, Lachaux A: Esophageal perforation in eosinophilic esophagitis: five cases in children. Endosc Int Open. 8(7):E830-E833, 2020

Laparoscopic Excision Urachal Remnant

Urachal remnants are rare congenital anomalies pertaining to the development and involution of the urachus. The urachus is the embryological remnant of the allantois. A vestigial fibrous cord forms that obliterates in normal development to form a cord lying between the peritoneum and transversalis fascia and connecting the umbilicus to the bladder dome. Failure of obliteration at birth in the connection of the bladder to the umbilicus results in a urachal anomaly. Five urachal remnants are recognized, and they include: patent urachus with urine coming from the umbilicus (fistula), vesicourachal diverticulum, urachal sinus, urachal cyst and alternating fistula which drain either to the umbilicus or bladder. Urachal cyst is the most common anomaly of the urachus occurring in approximately one of 5000 births. Though clinically asymptomatic, the child can present with abdominal suprapubic pain, infraumbilical swelling with erythema, urinary symptoms (dysuria), infection of the cyst, umbilical drainage, umbilical mass, omphalitis and incidental finding during surgery. Persistent urachal anomalies can lead to recurring infection, stone formation and development of adenocarcinoma in the epithelium of the urachus. Urachal anomalies are associated with vesicoureteral reflux, hypospadia, meatal stenosis and ureteropelvic obstruction. Infection is the most common complication of urachal remnants. When infected it might need percutaneous drainage and systemic antibiotics followed by excision. Diagnosis can be established with ultrasound, CT-Scan, MRI or VCUG (less sensitivity; not part of standard evaluation). Managements of urachal anomalies include resection of the urachus throughout its entire length from the navel to include a cuff of normal bladder to avoid leaving urachal epithelium behind. There is controversy whether to do bladder cuff excision to all cases. In patent urachus and diverticulum remnant bladder cuff resection is indicated. Without bladder cuff excision postoperative Foley time, recurrence and complications are significantly less. Traditionally this urachal resection has been performed using an open hypogastric transverse or midline vertical incision. During the past 20 years the laparoscopic technique has been utilized to manage urachal anomalies in children and adults. Three trocar technique is usually utilized. The bladder cuff resection can be performed with double endoloops sutures as using a mechanical stapler machine can bring problems of future stone formation within the suture line or bleeding from the staplers. The laparoscopic approach confirms the presence of the urachus, enables magnified dissection along the extraperitoneal plane until the dome of the bladder in the space of Retzius. The laparoscopic approach is associated with minimal postoperative pain, rapid recovery and return to normal activities with the added advantage of better cosmetic results.

References:

1- Navarrete S, Sanchez Ismayel A, Sanchez Salas, et al: Treatment of urachal anomalies: a minimally invasive surgery technique. JSLS. 9(4):422-5, 2005

2- Kim TW, Chung H, Yang SK, et al: Laparoscopic management of complicated urachal remnant in a child. J Korean Med Sci. 21(2):361-4, 2006

3- Kurtz M, Masiakos PT: Laparoscopic resection of a urachal remnant. J Pediatr Surg. 43(9):1753-4, 2008

4- Sukhotnik I, Aranovich I, Mansur B, et al: Laparoscopic Surgery of Urachal Anomalies: A Single-Center Experience. Isr Med Assoc J. 18(11):673-676, 2016

5- Chiarenza SF, Bleve C: Laparoscopic management of urachal cysts. Transl Pediatr. 5(4):275-281, 2016

6- Stopak JK, Azarow KS, Abdessalam SF, et al: Trends in surgical management of urachal anomalies. J Pediatr Surg 50: 1334-1337, 2015
7- Tanaka K, Misawa T, Baba Y, et al: Surgical management of urachal remnants in children: open versus laparoscopic approach: A STROBE-compliant retrospective study. Medicine (Baltimore). 2019 Oct;98(40):e17480. doi: 10.1097/MD.0000000000017480.

Angiosarcoma

Angiosarcomas are extremely rare malignant vascular tumors originating from endothelial cells differentiation found almost anywhere in the body. They account for less than 1%of all sarcomas in children. Angiosarcomas predominantly arise from skin and subcutaneous tissue of the head and neck region, but they may also arise from deep soft tissue and other organs such as liver, spleen, kidney, heart, breast, thyroid gland and bone. They are associated with chronic lymphedema, radiation, arteriovenous fistulas and chronically immunosuppressed patients. Some authors have described angiosarcoma in children developing as a result of exposure to environmental factors such as radiation or arsenic. Histologically angiosarcomas can be well-differentiated to high-grade, stroma poor epithelioid neoplasms categorized as papillary, spindled, epithelioid or plasmacytoid. More than 50% of angiosarcomas demonstrated epithelioid characteristics. Epithelioid angiosarcomas tumor cells often forms sheets tubules or cluster of epithelioid malignant cells. In the liver, angiosarcoma presents as an abdominal mass. Associated symptoms can be jaundice, abdominal pain, vomiting, fever, tachypnea, dyspnea and anemia. High output cardiac failure, ascites, disseminated intravascular coagulation, bleeding and Kasabach-Merritt syndrome has also been reported in hepatic angiosarcoma in children. Diagnosis is made with US, CT-Scan and MRI. Adequate representative tissue is needed to establish a histologic diagnosis usually obtained through laparotomy. Overall prognosis of hepatic angiosarcoma is very poor regardless of therapy. Combination of chemotherapy, radiotherapy and surgical resection seldom provides a long-term disease-free survival in children. Liver transplantation also carries a high recurrence rate and poor posttransplant survival. Splenic angiosarcoma is extremely rare, aggressive malignancy that is also uniformly fatal. Only children with localized disease amenable to surgical resection can achieve long-term survival. They present with abdominal pain, pancytopenia and splenomegaly. Splenic angiosarcomas proliferate rapidly, recur locally, spread widely and have a propensity to lymph node dissemination. Small tumor size (< 5 cm) is associated with better prognosis. Overall prognosis is grim.

References:

1- Geller RL, Hookim K, Sullivan HC, et al: Cytologic features of angiosarcoma: A review of 26 cases diagnosed on FNA. Cancer Cytopathol. 124(9):659-68, 2016
2- Geramizadeh B, Safari A, Bahador A, et al: Hepatic angiosarcoma of childhood: a case report and review of literature. J Pediatr Surg. 46(1):e9-11, 2011
3- Serrano OK, Knapp E, Huang K, et al: Pediatric primary splenic angiosarcoma: an aggressive multidisciplinary approach to the oncologic management of a rare malignancy. World J Surg Oncol. 12:379, 2014
4- Lee KC, Chuang SK, Philipone EM, Peters SM: Characteristics and Prognosis of Primary Head and Neck Angiosarcomas: A Surveillance, Epidemiology, and End Results Program (SEER) Analysis of 1250 Cases. Head Neck Pathol. 13(3):378-385, 2019

5- Sparber-Sauer M, Koscielniak E, Vokuhl C, et al: Endothelial cell malignancies in infants, children and adolescents: Treatment results of three Cooperative Weichteilsarkom Studiengruppe (CWS) trials and one registry. Pediatr Blood Cancer. 2020 Mar;67(3):e28095. doi: 10.1002/pbc.28095. Epub 2019 Dec 8.
6- Pariury H, Golden C, Huh WW, et al: Pediatric ovarian angiosarcoma treated with systemic chemotherapy and cytoreductive surgery with heated intraperitoneal chemotherapy: Case report and review of therapy. Pediatr Blood Cancer. 2019 Jul;66(7):e27753. doi: 10.1002/pbc.27753. Epub 2019 Apr 11.

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