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Median Arcuate Ligament Syndrome

Median arcuate ligament syndrome (MALS) also known as celiac artery compression is a rare condition that occurs when the fibrous portion of the diaphragmatic crura known as the median arcuate ligament compresses the proximal celiac artery trunk. This compression can also occur with an excessive amount of sympathetic nerves or ganglions. The compression usually occurs during expiration. MALS is a diagnosis of exclusion characterized by weight loss, postprandial abdominal pain, diarrhea, nausea, vomiting and epigastric bruit. Most cases are female. The diagnosis can be established using invasive arteriography or non-invasive with CT angiography, magnetic resonance angiography and Doppler ultrasound studies. CT-angio demonstrates a focal hook appearance narrowing in the proximal celiac artery. Doppler studies shows variations in peak systolic velocity with a marked increase during expiration and functional geometric changes such as celiac trunk deflection. Treatment options for MALS include surgical or laparoscopic division of the median arcuate ligament, celiac ganglion destruction, endovascular stenting or bypass surgery. Advantages of the laparoscopic approach are the imaging magnification, fewer postoperative adhesions, smaller incisions, better cosmesis and shorter hospital stay. Besides relieving the mechanical effect of the arcuate ligament, the interruption of somatic or sympathetic fibers in the course of division may be responsible for the improvement in abdominal pain due to relieve of vasospasm.

References:

1- Alehan D, Dogan OF: Pediatric surgical image. A rare case: celiac artery compression syndrome in an asymptomatic child. J Pediatr Surg. 39(4):645-7, 2004
2- Gander S, Mulder DJ, Jones S, Ricketts JD, Soboleski DA, Justinich CJ: Recurrent abdominal pain and weight loss in an adolescent: celiac artery compression syndrome. Can J Gastroenterol. 24(2):91-3, 2010
3- Said SM, Zarroug AE, Gloviczki P, Shields RC: Pediatric median arcuate ligament syndrome: first report of familial pattern and transperitoneal laparoscopic release. J Pediatr Surg. 45(12):e17-20, 2010
4- Aschenbach R, Basche S, Vogl TJ: Compression of the celiac trunk caused by median arcuate ligament in children and adolescent subjects: evaluation with contrast-enhanced MR angiography and comparison with Doppler US evaluation. J Vasc Interv Radiol. 22(4):556-61, 2011
5- Ozel A, Toksoy G, Ozdogan O, et al: Ultrasonographic diagnosis of median arcuate ligament syndrome: a report of two cases. Medical Ultrasonography 14(2): 154-157, 2012
6- Wani S, Wakde V, Patel R, et al: Laparoscopic release of median arcuate ligament. J Minim Access Surg 8(1): 16-18, 2012

Giant Omphalocele

A giant omphalocele is defined as a defect larger than 10 cm in length that harbors the liver. Prenatally diagnosed giant omphaloceles will need cesarean section as route of

birth to avoid fetal liver rupture. Management of giant omphalocele has a high morbidity and mortality due to the defect size, visceroabdominal disproportion, and the associated congenital and genetic malformations. The large size of the defect and small abdominal cavity creates a situation where primary closure is almost impossible unless some sort of stage reduction is tailored. Pulmonary hypoplasia, genetic defects and cardiac malformation are the source of early mortality in these babies. The pendulum of management of giant omphalocele has moved toward a more conservative initial management using topical coverage creams to create granulation tissue and skin on top of the membrane followed by repair of the ventral hernia much later in life when the medical condition of the child permits. Silver sulfadiazine (Silvadene) provides a moist wound healing environment that promotes epithelization and simultaneously minimizes the risk of invasive infection including antifungal coverage. Silver toxicity, though rare can include seizures, peripheral neuropathy, ocular pathology, nephrotic syndrome, raised liver enzymes, leukopenia and algiria. For smaller size defects the use of silo, tissue expanders, biologic mesh, vacuum-assisted closure or component separation technique closure is indicated. Giant omphalocele is associated with deficits in developmental achievements in most of the affected infants ranging from mild to profound delays.

References:

1- Pacilli M, Spitz L, Kiely EM, Curry J, Pierro A: Staged repair of giant omphalocele in the neonatal period. J Pediatr Surg. 40(5):785-8, 2005

2- Kilbride KE, Cooney DR, Custer MD: Vacuum-assisted closure: a new method for treating patients with giant omphalocele. J Pediatr Surg. 41(1):212-5, 2006

3- Lee SL, Beyer TD, Kim SS, Waldhausen JH, Healey PJ, Sawin RS, Ledbetter DJ: Initial nonoperative management and delayed closure for treatment of giant omphaloceles.J Pediatr Surg. 41(11):1846-9, 2006

4- van Eijck FC, de Blaauw I, Bleichrodt RP, Rieu PN, van der Staak FH, Wijnen MH, Wijnen RM: Closure of giant omphaloceles by the abdominal wall component separation technique in infants.J Pediatr Surg. 43(1):246-50, 2008

5- Danzer E, Gerdes M, D'Agostino JA, Bernbaum J, Siegle J, Hoffman C, Rintoul NE, Liechty KW, Flake AW, Adzick NS, Hedrick HL: Prospective, interdisciplinary follow-up of children with prenatally diagnosed giant omphalocele: short-term neurodevelopmental outcome. J Pediatr Surg. 45(4):718-23, 2010

6- Ein SH, Langer JC: Delayed management of giant omphalocele using silver sulfadiazine cream: an 18-year experience. J Pediatr Surg. 47(3):494-500, 2012

Congenital Solid Tumors

Congenital solid tumors refer to masses of tissue that have grown in location diverge from the normal pattern of development and do not duplicate any normal structure of the body. Though rare prenatal sonography has increased the rate of diagnosis. Abdominal tumors are the most common prenatally diagnosed fetal tumors. Half of congenital solid tumors are diagnosed at birth and two-third during the first week of life. The most common congenital tumors are extracranial teratoma, neuroblastoma, soft tissue tumors and brain tumors. The most frequent benign tumor is teratoma, while the most frequent malignant is neuroblastoma. Although easier to detect, cervical and mediastinal tumors have a worse prognosis. Abdominal masses are more difficult to

detect but have a better prognosis. Management should be conservative as possible with surgery playing a major role due to the relative benign behavior of congenital cancer and the potential long term effect of chemotherapy and radiotherapy.

References:

1-Parkes SE, Muir KR, Southern L, Cameron AH, Darbyshire PJ, Stevens MC: Neonatal tumours: a thirty-year population-based study. Med Pediatr Oncol. 22(5):309-17, 1994

2- Sbragia L, Paek BW, Feldstein VA, Farrell JA, Harrison MR, Albanese CT, Farmer DL: Outcome of prenatally diagnosed solid fetal tumors. J Pediatr Surg. 36(8):1244-7, 2001

3- Albert A, Cruz O, Montaner A, Vela A, et al: [Congenital solid tumors. A thirteen-year review].Cir Pediatr. 17(3):133-6, 2004

4- Mahony R, McParland P: Approaches to the management of antenatally diagnosed congenital tumours.Pediatr Radiol. 39(11):1173-8, 2009

5- Lopez Almaraz R, Villafruela Alvarez C, Rodriguez Luis J, Domenech Martinez E: [Neonatal neoplasms: a single-centre experience]. An Pediatr (Barc). 65(6):529-35, 2006

6- Crocoli A, Bagolan P, Boldrini R, et al: Congenital Askin tumor with favorable outcome: case report and review of the literature. J Pediatr Surg 47(7): 1440-44, 2012

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