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Urachal Remnants

Urachal remnants are uncommon anomalies that present symptoms in infancy or early childhood. The urachus is formed in early embryonic life as a tubular connection between the dome of the bladder and the allantoic stalk. This fistulous tract obliterates into a medial ligament. Rare persistence of this remnant after birth can lead to a completely patent urachus, a sinus (opening to the navel), a diverticulum (opening to the bladder), a cyst (both end close but central portion remains open) or alternating sinus (cyst-like structure can drain to either the bladder or umbilicus). Neonatal patent urachus can undergo spontaneous involution. Otherwise, the transitional epithelium is replaced by granulation tissue and the main symptom of a patent urachus is urine discharge from the umbilicus associated with cellulitis, pain and swelling. A cyst or diverticulum can present as a midline, localized, painful abdominal mass associated with fever and leucocytosis. The diagnosis can be done with ultrasound (cystic process is shown), cystography (the ventral position of urachal diverticulum and fistula is seen, and a bladder-neck obstruction diagnosed) or fistulography (a connection with the bladder is demonstrated). If infected initial management consists of antibiotics, otherwise cure is accomplished with excision of the urachal remnant with a cuff of bladder (to avoid recurrent infection, stone formation and later development of carcinoma), double layer closure and drainage.

References

1- Zieger B, Sokol B, Rohrschneider WK, Darge K, Troger J: Sonomorphology and involution of the normal urachus in asymptomatic newborns. Pediatr Radiol 28(3):156-61, 1998
2- Suita S, Nagasaki A: Urachal remnants. Semin Pediatr Surg 5(2):107-15, 1996
3- Nagasaki A, Handa N; Kawanami T: Diagnosis of urachal anomalies in infancy and childhood by contrast fistulography, ultrasound and CT. Pediatr Radiol 21(5):321-3, 1991
4- Goldman IL, Caldamone AA, Gauderer M, Hampel N, Wesselhoeft CW, Elder JS: Infected urachal cysts: a review of 10 cases. J Urol 140(2):375-8, 1988
5- Newman BM; Karp MP; Jewett TC; Cooney DR: Advances in the management of infected urachal cysts. J Pediatr Surg 21(12):1051-4, 1986

Medullary Thyroid Carcinoma

Medullary thyroid carcinoma (MTC) is a rare, solid, thyroid neoplasm with amyloid stroma that arise from the parafollicular C-cells, is inherited as autosomal dominant, and may develop in childhood sporadically or associated with a multiple endocrine neoplasia (MEN) or Familial syndrome. In sporadic cases the presentation occurs in adolescence as a thyroid nodule. Production of thyrocalcitonin by MTC and its precursor (C-cell hyperplasia) permits diagnosis and follow-up, though a significant number of these children are not

cured by surgery due to extensive disease at diagnosis. DNA testing has found that the RET proto-oncogene mutation is associated with MCT development in kindreds of sporadic cases, MEN or familial MTC syndromes. This has permitted early (prophylactic) gland removal in infancy (MEN IIB) or early childhood (MEN IIA) before biochemical or clinical MCT develops. MCT metastasizes to local lymph nodes followed by lungs, bone and liver. Management consists of total thyroidectomy and central lymph node sampling with general sampling if enlarged.

References

1- Telander RL, Moir CR: Medullary thyroid carcinoma in children. Semin Pediatr Surg 3(3):188-93, 1994
2- Wells SA Jr, Chi DD, Toshima K, Dehner LP, Coffin CM, Dowton SB, Ivanovich JL, DeBenedetti MK, Dilley WG, Moley JF, et al: Predictive DNA testing and prophylactic thyroidectomy in patients at risk for multiple endocrine neoplasia type 2A. Ann Surg 220(3):237-47, 1994
3- Skinner MA, DeBenedetti MK, Moley JF, Norton JA, Wells SA Jr: Medullary thyroid carcinoma in children with multiple endocrine neoplasia types 2A and 2B. J Pediatr Surg 31(1):177-81, 1996
4- La Quaglia MP, Telander RL: Differentiated and medullary thyroid cancer in childhood and adolescence. Semin Pediatr Surg 6(1):42-9, 1997
5- Skinner MA, Wells SA Jr: Medullary carcinoma of the thyroid gland and the MEN 2 syndromes. Semin Pediatr Surg 6(3):134-40, 1997
6- Lallier M, St-Vil D, Giroux M, Huot C, Gaboury L: Prophylactic thyroidectomy for medullary thyroid carcinoma in gene carriers of MEN2 syndrome. J Pediatr Surg 33(6):846-8, 1998
7- van Heurn LW, Schaap C, Sie G, Haagen AA, Gerver WJ, Freling G, van Amstel HK, Heineman E: Predictive DNA testing for multiple endocrine neoplasia 2: a therapeutic challenge of prophylactic thyroidectomy in very young children. J Pediatr Surg 34(4):568-71, 1999

Gastric Perforation

Gastric perforation (GP) is a rare abdominal catastrophe seen chiefly in premature infants. GP occurs primarily (spontaneously) after selective ischemia with blood shunting during periods of neonatal asphyxia. Secondary GP is caused by mechanical disruption (excessive distension or instrumentation) as observed in situations such as: aggressive mask resuscitation, duodenal atresia, esophageal atresia with TE fistula, volvulus of the stomach, esophageal intubation, mechanical ventilation and nasogastric tubes (iatrogenic). Boys are more commonly affected than girls. Sudden onset of abdominal distension, feeding intolerance, respiratory distress, metabolic acidosis, shock and hypoactivity within the first week of life is characteristic. Pneumoperitoneum is seen in plain abdominal films. Most GP occurs along the greater curvature of the stomach between the smooth muscle layers. With severely sick infants, temporary peritoneal drainage with lavage removes gas and acid, decompresses the abdomen improving ventilation (abdominal compartment syndrome), and grants time to stabilized the sick infant (improve acidosis, shock and coagulopathy) in preparation for surgical closure of the perforation. GP carries a high mortality rate.

References

1- St-Vil D, LeBouthillier G, Luks FI, Bensoussan AL, Blanchard H, Youssef S: Neonatal gastrointestinal perforations. J Pediatr Surg 27(10):1340-2, 1992
2- Shashikumar VL, Bassuk A, Pilling GP IV, Cresson SL: Spontaneous gastric rupture in the newborn: a clinical

review of nineteen cases. Ann Surg 182(1):22-5, 1975
3- Rosser SB, Clark CH, Elechi EN: Spontaneous neonatal gastric perforation. J Pediatr Surg 17(4):390-4, 1982
4- Tan CE, Kiely EM, Agrawal M, Brereton RJ, Spitz L: Neonatal gastrointestinal perforation. J Pediatr Surg 24(9):888-92, 1989
5- Houck WS Jr, Griffin JA 3d: Spontaneous linear tears of the stomach in the newborn infant. Ann Surg 193(6):763-8, 1981
6- Aviles T, Lugo-Vicente H, Ocasio MT, Guiven A, Pagán V, Lliteras O, Vázquez H: Perforated NEC: The role of percutaneous peritoneal drainage (submitted for publication).

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