Gastrinoma

Gastrinomas, the cause of the Zollinger-Ellison syndrome, are gastrin producing tumors that result in fulminant peptic ulceration and diarrhea. In children, half the cases are associated with malignancy, metastasis and multicentricity. Epidemiologically 75% gastrinomas are sporadic and 25% associated with MEN type 1. Diagnosis of gastrinoma is confirmed after documenting fasting serum hypergastrinemia (> 200 pg/ml), markedly elevated gastric output, a positive secretin-stimulated gastrin analysis or histologic confirmation of a neuroendocrine tumor. Most tumors are found in the head, body and tail of the pancreas, followed by duodenum, liver and lymph nodes. Localization studies include CT-scan, MRCP, octreotide scan (specially indium-labeled pentetreotide), endoscopic US, and Imamura test (selective arterial secretin injection test). Radical resection of the gastrinoma to achieve biochemical cure even when tumor is extrapancreatic and in lymph nodes is the cornerstone of therapy. Unresectable or metastatic cases are managed with pharmacologic therapy using proton pump inhibitors and octreotide. Total gastrectomy is reserved for absolute failure of both medical and surgical management. Gastrinomas in children are slow growing, indolent, and compatible with long life. Prognostic factors correlating with long-term survival are small tumor size, absence of metastatic disease and non-pancreatic location of primary tumors. Factors that do not affect survival include age at diagnosis, sex, presence of lymph node metastases, associated multiple endocrine neoplasia, and method of ulcer treatment.

References:

Epigastric Hernias

Congenital epigastric defects occur anywhere in the linea alba from the navel to the xiphoid process. They represent almost 5% of all hernias defect that presents in children. Most epigastric hernias occur in the midline, are small (15-25 mm), asymptomatic and
reducible. Multiple fascial defects can also be present in 20% of all cases. The defect might arise congenitally from an abnormally wide orifice of a blood vessel during development of the linea alba. The bump is the result of a piece of preperitoneal fat stuck through the fascial defect. Tenderness is an unusual symptom while growth of the defect occurs with time. Most surgeons recommend repair of the defect at the time of presentation. Repair is an outpatient procedure done under general anesthesia with low morbidity and risk of recurrence. Voluminous epigastric hernia (5-10 cm) with a sac that contains epiploic appendages or viscera (ileum loops, stomach) has also been rarely reported in infants.

References:

Necrotizing Fasciitis

Necrotizing fasciitis (NF) is a rare, severe, rapidly progressive soft tissue infection involving the skin, subcutaneous tissue and superficial fascia. Most cases occur during the newborn period. In newborns NF is most frequently associated with omphalitis, necrotizing enterocolitis, balanitis, urachal anomalies and fetal monitoring. In older children NF can be seen as a postoperative complication (appendectomy, gastrostomy), in neutropenia after chemotherapy and secondarily to varicella infestation. Most frequent sites of initial involvement are the abdominal wall, followed by thorax, back, scalp, and extremity. Infection is usually polymicrobial containing both gram positive (beta-hemolytic Streptococcus and Staphylococcus aureus) and gram negative bacteria (E. Coli and Pseudomonas). Important clinical findings in infants with NF are tachycardia, abnormal white blood cell count, systemic toxicity, severe edema, and, in older children pain out of proportion to the apparent degree of infection. The child can also develop wound crepitation and radiographic evidence of subcutaneous gas. An ominous sign indicative of the need for immediate radical debridement is the appearance of a patch of dusky or gangrenous skin. Initial management consists of antibiotics, intravenously afluids, blood transfusions, calcium replacement (due to saponification) and general patient support. Neutropenic patients benefit from granulocyte transfusions. Surgical procedures include extensive debridement as soon as possible and as needed for continued necrosis, secondary closure and skin grafting. Mortality ranges from 10 to 60%. Improved survival requires early diagnosis followed by prompt aggressive surgical debridement.

References:
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