

PEDIATRIC SURGERY Update © Vol 17 No 01 JULY 2001

Hidradenitis Suppurativa

Hidradenitis suppurativa refers to a chronic relapsing cutaneous inflammation of the apocrine glands of the body. Chronic hidradenitis suppurativa can affect, in order of preference, the axilla, perineum, scrotum, inquinal, scalp, palms of the hand, soles of the feet and the submammary region. Hidradenitis suppurativa usually manifests itself after puberty (androgen-dependent disorder). Females are more commonly affected than males. The inflammatory process begins as a local occluding spongiform infundibulo-folliculitis of a sweat gland with subsequent rupture and secondary bacterial infection. Symptoms include pain, swelling, purulent discharge, and pruritus of the affected region. Associated medical conditions include diabetes and obesity. Initial management consists of general hygienic measures with antibiotics, antiandrogens and estrogens. Surgery is needed when the condition is at an advanced stage with cellulitis and scarring. Nonoperative treatment is disappointing. Total excision of all apocrine-bearing axillary tissue with primary closure is the treatment of choice. Operative treatment can be safely accomplished even when draining sinuses are present. Recurrence results from inadequate excision or an unusually wide distribution of apocrine glands, but physical factors such as obesity, local pressure, and skin maceration play a role. Radical surgery gives good symptomatic control of severe hidradenitis suppurativa of the axilla, inguinoperineal, and perianal regions but is less satisfactory for submammary disease.

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Desmoid Tumors

Desmoid fibromatosis (DF) is a benign, locally aggressive tumor, with a strong propensity for infiltrative growth and local recurrence. More than 50% of the tumors develop within the first five years of life as an asymptomatic, firm, solid mass. DF can be found on the

head and neck, upper or lower extremities, the abdomen or the trunk. The lower extremities are the most frequent sites of manifestation. The most frequent types are the infantile fibromatosis (head, neck, shoulder, upper arm or thigh), extra-abdominal fibromatosis (chest wall, back and thigh) and fibromatosis colli (neck). Superficial lesions tend to be slow growing, small and rarely involve deep structures. Deep-seated DF tends to be faster growing, larger and involves deeper structures (extra-abdominal). Except fibromatosis colli that tends to regress spontaneously, infantile and extra-abdominal DF is best managed by gross total resection achieving negative margins unless tumor excision or chemotherapy is most often used with recurrent disease or as an alternative to mutilating surgery. Adjuvant radiation therapy improves local control. DF have no tendency to metastasize. It is believed they should be treated as low-grade malignancies with documentation of histologic margins and close clinical follow-up.

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Dieulafoy Lesion

First described by a French surgeon in 1897, Dieulafoy's lesion (DL) also known as cirsoid aneurysm, is a rare cause of massive upper gastrointestinal hemorrhage. Most DL occurs due to an abnormal submucosa artery in the stomach particularly along the lesser curvature in the region supplied by the left gastric artery and within six centimeters of the gastroesophageal junction. They have also been reported to occur in the jejunum and rectum. The problem with this lesion is that bleeding is intermittent and massive. With fiberoptic endoscopy the source of upper and lower gastrointestinal bleeding can be identified in 80 to 85% of children. The endoscopic appearance of DL may range from a pinpoint dot (two to 5 mm), clot, or tortuous vessel, to blood oozing or spurting from normal mucosa. There is no surrounding mucosal ulceration. Other times diagnosis is made at operation, as endoscopy and arteriography fail to identify the lesion. Repeated endoscopies might be needed. Management consists of endoscopic sclerotherapy injection and laser photocoagulation. If this fails, surgery follows. Simple ligation of the bleeding dot is all that is required to control the hemorraghe.

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