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RMS

Rhabdomyosarcoma (RMS) the most common soft tissue sarcoma in infants and children represents about 5-15% of all malignant solid lesions. It has a peak incidence before the age of five years, and a second surge during early adolescence. Head, neck and pelvic malignancies are more prevalent in infancy and early childhood, while trunk, extremity and paratesticular RMS is largely a disease of adolescents. RMS arises from a primitive cell type and occurs in mesenchymal tissue at almost any body site (excluding brain & bone). Predominant histologic type in infants and small children is embryonal. Botryoid RMS is a subtype of the embryonal variety, which ordinarily extends into body cavities such as bladder, nasopharynx, vagina, or bile duct. The alveolar cell type, named for a superficial similarity to the pulmonary alveoli, is the most common form found on the muscle masses of the trunk and extremities, and is seen more frequently in older children. Clinical findings, diagnostic evaluation and therapy depend upon location of the primary tumor and is beyond the scope of this review. Head and neck RMS are most common and occur in the orbit, nasopharynx, paranasal sinuses, cheek, neck, middle ear, and larynx. Most are treated by simple biopsy followed by combined therapy or preop chemotherapy and radiation followed by conservative resection. Operations for extremity lesions include wide local excision to remove as much of gross tumor as possible. The trend in management is more chemotherapy with conservative surgical therapy. Survival depends on primary site, stage of disease, and treatment given.

Hepatic Cysts

Hepatic cysts (HC) can be either parasitic (echinococcal) following infestation in endemic regions, acquired (after trauma or inflammatory processes), or nonparasitic (congenital) in nature. Congenital nonparasitic HC are uncommon, solitary, benign lesions that arise from aberrant development of intrahepatic biliary radicals after ischemic thrombo-embolic phenomena (vascular disruption theory). The cyst is lined with cuboidal or squamous epithelium, and there is a female and white children predominance. Although generally asymptomatic, children may manifest increased abdominal girth,

vague abdominal discomfort, infection, or obstructive jaundice. Ultrasound and CT-Scan are diagnostic tools. Management may consist of: simple unroofing, complete removal by enucleation or hepatic lobectomy, internal roux-en-Y drainage, or percutaneous aspiration and sclerosis (alcohol, minocycline). The surgical alternative to use will depend on size, location (central, peripheral or dumbbell), and presence of communication with biliary system of the cyst (see figure). Some cases diagnosed prenatally or during the neonatal period have undergone slow spontaneous regression.

HSP

Henoch-Schönlein Purpura (HSP) is a generalized autoimmune vasculitis characterized by non-thrombocytopenic purpuric rash, arthritis, glomerulonephritis, and gastrointestinal symptoms thought to occur as diffuse IgA hypersensitivity response. Main clinical features are: age between 3 and 7 years, skin rashes followed by GI symptoms, joint symptoms, soft tissue edema, and renal involvement. GI symptoms (pain, vomiting and bleeding) occurring in almost 2/3 of cases may need prompt surgical evaluation during crisis. Some complication leading to surgical therapy includes intussusception (most common, involves small bowel alone), perforation, infarction, and massive GI bleeding. Ultrasound can show the thickened hemorrhagic infiltration of the intestinal wall, follow evolution of these lesions and identify potential surgical complications Obstructed children need sequential Ba enema and UGIS. Steroids can mask symptoms of fistula and abscess formation. Suspicion and early diagnosis of HSP after clinical, imaging and lab findings avoid unnecessary operations.

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