



PEDIATRIC SURGERY Update © **Vol. 47 No. 01 JULY 2016**

Paratesticular Rhabdomyosarcoma

Paratesticular rhabdomyosarcoma (Pt-RMS) comprise 10% of all genitourinary RMS tumors and third most common after prostate and bladder in children. Presents before the age of five years or in adolescents as a painless scrotal mass, trauma or bruising. Testicular ultrasound should be the initial diagnostic imaging demonstrating a solid heterogenous extratesticular mass. In Pt-RMS levels of beta-HCG and alpha fetoprotein are not elevated, while LDH might be elevated if there is significant metastatic disease. Thin-cut (5 mm) CT-Scan with IV/po contrast is needed once the diagnosis is established for clinical staging regarding pulmonary, mediastinal and retroperitoneal metastasis. The most common histologic variant is embryonal RMS (80%). TNM staging system is used to stage these tumors based on tumor size, invasiveness, nodal status and presence of distant metastasis. Tumor location also decides a favorable or unfavorable prognosis. Pt-RMS can be either stage I or IV given its location as a favorable primary site. RMS staging is multifactorial and outcome depends on three different classifications: Stage (determine by location, size, presence of regional nodes or metastasis), Group (based on tumor status after resection or biopsy, tumor margin and lymph node disease) and Risk (combination of stage, group and histology). FDG-PET/CT is more sensitive tool in staging an restaging patients with RMS, and also in the assessment of chemotherapy response. Evaluation should include bone marrow, bone scan and lumbar puncture. Multimodal therapy with surgery, chemotherapy and radiotherapy is used to maximize tumor control. Tissue diagnosis is the initial step and suspected Pt-RMS should undergo radical orchiectomy through an inguinal incision with high ligation of the spermatic cord. Scrotal approach is inadequate due to microscopic residual disease needing wide local re-excision of the scrotal scar. Ipsilateral lymph node dissection is controversial; recommended with evidence of enlarged lymph nodes imaging. Survival at 3-years is 95% with multimodal therapy.

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Wilms Tumor: Intracaval Extension

Wilms tumor also known as nephroblastoma is the most common malignant renal tumor in children. The survival rate of Wilms tumor has improved significantly over the years to better than 90% long-term survival with the use of surgery, chemotherapy and in a few instances radiotherapy. Large size of tumor, involvement of adjacent vital structures and intracaval tumor thrombus are universal accepted inoperable criteria. Wilms tumor has a strong tendency to invade blood vessels in the form of tumor thrombus into the renal veins, inferior vena cava and right atrium. Extension of tumor thrombus along to the renal veins into the inferior vena cava occurs in 4-10% of all children, while intraatrial extension occurs in 1-3%. Intracaval extension has been reported to be more common in the right kidney due to a short anatomic renal vein. Most cases with intracaval extension of tumor are asymptomatic and the diagnosis is made during imaging workup (Ultrasound with Doppler, CT-Scan or MRI). Preop identification of intracaval extension is important for surgery, since removal of the tumor can cause significant bleeding and/or tumor embolization with acute cardiac decompensation or arrest after manipulation of the thrombus if it's not adhered to the vessel wall. Staging the extension includes: Level 1-infrahepatic extension < 5 cm, Level 2 - intrahepatic extension > 5 cm and Level 3 - suprahepatic or Level 4 - atrial extension. Identification of intracaval tumor extension should ideally be managed with preoperative chemotherapy prior to resection to reduce the anticipated surgical risks. Preop chemotherapy reduces size of the tumor, dissolute the thrombus and provides easy surgical removal. This approach can avoid but nor eliminate the need of cardio-pulmonary bypass and cavotomy during removal of the tumor. Failure of regression, failure to tolerate chemotherapy or acute tumor rupture may need early resection. Single best predictor of survival is the histologic subtype.

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Urticaria Pigmentosa

Urticaria pigmentosa (UP) falls into the classification of disorders known as pediatric maculopapular cutaneous mastocytosis. Most cases occur in white patients with lesions occurring before the age of two years in the majority of cases. Urticaria pigmentosa is associated with mutation in the signaling receptor molecule c-KIT. UP can appear as a generalized maculopapular rash in the trunk and proximal extremities or as a mastocytoma single lesion large tan-orange plaque or nodule. With mechanical irritation of the plaque or nodule histamine, leukotriene and prostaglandin is release from mast cells causing the symptoms of urticaria (Darier's sign). The diffuse form has an indolent benign course. Diagnosis of urticaria pigmentosa is clinical. Biopsy is rarely necessary but is definitive and may be performed in cases where the diagnosis is not certain based on clinical features or the child is having constant irritation of the lesion with frequent signs of urticaria and pain. Histopathological diagnosis is made by observing mast cells showing metachromasia with toluidin blue in full-thickness skin biopsy. Main management of the systemic form of the disease consist of long acting oral H1-antihistamines. Persistently symptomatic mastocytomas or blistered or ulcerated lesions may be treated with high-potency topical glucocorticoids under occlusion or surgical excision if deemed necessary. The prognosis in most cases is excellent.

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