

# PEDIATRIC SURGERY Update 8 Vol. 33 No. 01 JULY 2009

## **Benign Mesothelioma**

Benign cystic mesothelioma (BCM) also known as cystic mesothelioma of the peritoneum is a rare abdominal tumor found in young women during their third decade of life. It originates from the pelvic retroperitoneum with a predilection for pelvic visceral serosal surfaces. It a neoplastic tumor with a high tendency for recurrence not associated to asbestos exposure. The tumor produce cysts that are usually intraperitoneal in location but also the chest and pericardium can be affected. Clinically the child presents with progressive painless abdominal distension. Most reliable method of preoperative diagnosis is with aspiration and cellular analysis of peritoneal washing showing abundant mesothelial cells. The role of immunohistochemistry and electron microscopy in diagnosis can be important. CT-Scan provides an idea of the extension of the benign tumor. The differential diagnosis includes a cystic lymphangioma. BCM shows multiplicity, extensive involvement of the peritoneal and serosal surfaces without significant organ invasion. Management of BCM is surgical excision. The cysts appear almost avascular. Multiple resections are common due to the recurrence rate of the tumor. Prognosis depends on the rate of recurrence and postoperative complications from multiple abdominal procedures.

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# **Anaplastic Thyroid Cancer**

Anaplastic or undifferentiated thyroid cancer occurs very rarely during young adults and children. Most cases of thyroid malignancy are well differentiated papillary and follicular variants. Other times medullary thyroid carcinoma associated with multiple endocrine neoplasia syndromes. Nevertheless it-s good to learn what is the characteristic of this unusual disease in young adults and children. Poorly-differentiated thyroid carcinoma is a stage in the development of anaplastic carcinoma from well-differentiated neoplastic transformation of follicular epithelium. The BRAFT1799A mutation has been identified. In patients under 40 years of age, encapsulated poorly-differentiated tumors are more

frequent with a trabecular histological pattern alone with a much smaller size (30 mm). Pathologic criteria used in the diagnosis of poorly or undifferentiated thyroid carcinoma includes (1) presence of a solid/trabecular/insular pattern of growth, (2) absence of the conventional nuclear features of papillary carcinoma, and (3) presence of at least one of the following features: convoluted nuclei; mitotic activity >or =3 x 10 HPF; and tumor necrosis. Among papillary carcinomas, the frequency of a solid growth pattern, a criterion for classifying a tumor as poorly differentiated, was higher in the Belarus region of Russia than that in Japan. Management of anaplastic thyroid malignancy is total thyroidectomy, since chemotherapy, radiotherapy and radioiodine are of no avail.

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### **Genital Tumors**

Tumors occurring in the vulva and external vaginal orifice are rare to find in female children. In the area of the vulva the most common tumor is either and hemangioma, lymphangioma, lipomas, neurofibromatosis and vulvar intraepithelial neoplasia. Hemangiomas resolve with conservative therapy, while lymphangiomas or other type of tumor will require surgical excision. Vulva intraepithelial neoplasia are associated with cases of sexual abuse and human papilloma viral infections. In the external vaginal orifice bleeding hemangiomas have been previously reported in children. Cavernous hemangiomas with brisk and continuous bleeding will require some form of therapy such as cryosurgery, excision or steroid injection. Interferon has also been used effectively. It is always important to study with imaging (MRI) the extension of the perineal hemangioma. Another lesion of importance in this privilege area is the vaginal rhabdomyosarcoma which presents with protrusion and a bleeding mass. Management consists of a biopsy, chemotherapy followed by surgery if necessary.

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