



# **PEDIATRIC SURGERY UPDATE ©**

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### **Infantile Myofibromatosis**

The fibromatosis are a heterogeneous collection of tumors distinguished by proliferating fibroblast, minimal mitotic and inflammatory activity, limited local fat and muscle invasion that affect all age groups. They originate in the fascial sheath and musculo-aponeurotic tissues, and represent about 12% of soft-tissue tumors in children. Infantile myofibromatosis represent the most common fibrous tumor of infancy. It is characterized by either a localized or multicentric form, appearance during the first year of life, and myofibroblast cells as main pathological participants. A benign, self limited disease process demonstrating spontaneous regression. It includes the formation of palpable masses in skin, muscle, viscera, bone, and subcutaneous tissue. The solitary (localized) form is more commonly found in the head, neck and trunk region affecting mainly boys later during infancy. Is twice as common as the multicentric form. Lesions are usually found in skin, muscle and subcutaneous tissue. Prognosis is excellent with a very low recurrence rate after surgical excision. Positive surgical margins within the tumor predict a high probability of recurrence, especially if large number of slit-like blood vessels and undifferentiated mesenchymal cells are identified. The multicentric form favor lesions in soft tissue, muscle, bone and viscera. Infants are born with the lesions and the prognosis depends on the degree of visceral involvement. Excision is reserved for lesions that jeopardize vital structures.

### **Lipoblastoma**

Tumors of adipose tissue origin are more commonly found in adult than children. They include lipoma, lipoblastoma and angiomyolipoma, to mention a few. Lipoblastoma is a benign tumor of fetal-embryonal fat tissue. Age at presentation is usually less than five years (mean 2.5 years) with a slight male predominance. The main clinical picture is that of a rapidly growing mass in a peripheral location, mainly the extremity (70%). It can arise within soft tissue. Other places identified are: shoulder, back, omentum, retroperitoneum, mediastinum and intrascrotal. Lipoblastomas are wellcircumscribed masses made up of immature fat cells. They have the capacity for differentiation. Microscopy will show lipoblasts with vacuolated cytoplasm present along with

primitive mesenchymal cells in a myxoid stroma with plexiform capillaries. Skeletal muscle is sometimes involved. Mitotic figures are normal and rare. The lesion produces a bright signal in both the T1 and T2 weighted magnetic resonance images. A breakdown in the long arm of chromosome eight (8q11-13) is a consistent finding in this tumor. Management consist of complete surgical removal to prevent local recurrence, avoiding radical mutilating resections whenever possible. The tendency to recur is approximately 15%. The prognosis is usually excellent.

### **Breast**

Most breast disorders in children of either sex are benign. Congenital lesions are: absent or multiple breast. Transplacental hormonal influence in neonates may cause hyperplasia of breast tissue with predisposition to infection (Mastitis neonatorum). Premature hyperplasia (thelarche) in females is the most common breast lesion in children. It occurs before the age of eight as a disk-shaped concentric asymptomatic subareolar mass. Remains static until changes occur in the opposite breast 6-12 mo later. It can regress spontaneously or stay until puberty arrives. Biopsy may mutilate future breast development. On the contrary, discrete breast masses in males cause concern and excision is warranted. Gynecomastia is breast enlargement cause by hormonal imbalance, usually in obese pre-adolescent boys. If spontaneous regression does not occur, it can be managed by simple mastectomy. Virginal hypertrophy is rapid breast enlargement after puberty due to estrogen sensitivity. If symptomatic, management is reduction mammoplasty.

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