33 Other Rare Conditions of Pseudotumoral and Benign Lesions of Soft Tissues

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33.1 Myxoma

The myxomas are a group of relatively common, entirely benign probably unrelated lesions, which most commonly involve large muscles (intramuscular myxoma) but may also occur around large joints (juxta-articular myxoma) or in the skin (cutaneous myxoma). All cases are characterized by abundant myxoid matrix, bland stellate to spindled cells, and hypovascularity. These three morphological features are fundamental to differentiate myxoma to myxoid nodular fasciitis and to malignant myxoid neoplasms, such as myxofibrosarcoma and myxoid liposarcoma [1]. Local, complete excision is adequate treatment, but juxta-articular myxoma may recur locally in 30% of cases, particularly if incompletely excised [1].

33.2 Angioleiomyoma

Angioleiomyoma, also called angiomyoma and vascular leiomyoma, is a benign dermal or subcutaneous tumor composed of well-differentiated smooth muscle cells arranged around many vascular channels. It represents 4–5% of benign soft tissue tumors, occurring at all ages but commonest between the fourth and sixth decades of life [2]. Angioleiomyoma can occur anywhere in the body, but is most often seen in the extremities followed by the head and trunk and it typically presents as a small, slowly growing firm nodule measuring <2 cm in diameter associated with pain in half of patients. Incomplete excision or a deeply situated lesion may exceptionally result in local recurrence [2].

33.3 Glomus Tumor

Glomus tumor (also called glomangioma and glomangiomyoma) is mesenchymal neoplasms composed of cells resembling the modified smooth muscle cells of the normal glomus body. This tumor is rare accounting for less than 2% of soft tissue tumor with a similar gender distribution and with a predilection for young adults. The vast majority occur in skin or superficial soft tissue in the distal extremities associated with a long history of pain. Rarely (fewer than 40 cases reported in literature), glomus tumors are morphologically considered malignant when the tumor show marked nuclear atypia and any level of mitotic activity or atypical mitotic figures [3]. "Typical" glomus tumors are benign neoplasms that require only simple excision. Malignant cases are highly aggressive with metastases and death from disease in up to 40% patients.

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33.4 Nodular Fasciitis

Nodular fasciitis is a common, benign, self-limiting, pseudo-sarcomatous reactive process that is mainly composed of fibroblasts and myofibroblasts. It is a solitary, small (<3 cm), sometimes painful subcutaneous nodule that occurs in young and middle-aged adults (20–50 years), with no sex predilection and that develops rapidly (often <1 month). It can be seen anywhere in the body but most common in the upper extremities (50% of cases), especially in the forearm [4]. Morphologically, nodular fasciitis is a highly proliferative lesion and it is commonly mistaken for a sarcoma. Important histological clues to the diagnosis of nodular fasciitis include the short, randomly arranged fascicles, the absence of a well-developed thick-walled vasculature, and the absence of nuclear pleomorphism or hyperchromatism. Simple excision is the treatment of choice with a percentage of local recurrences of less than 2% of cases [4].

33.5 Proliferative Fasciitis and Proliferative Myositis

Proliferative fasciitis and proliferative myositis are morphologically similar to nodular fasciitis but contain ganglion-like myofibroblastic cells, which are usually negative for muscle markers. Proliferative fasciitis is usually seen in the subcutaneous tissue of the upper limbs of middle-aged adults (40–60 years), whereas proliferative fasciitis mainly affects the muscles of the trunk and shoulder girdle. In children, proliferative fasciitis may be cellular and mitotically active, mimicking rhabdomyosarcoma or epithelioid sarcoma [4]. Both these lesions are benign, self-limiting, and reactive process. Simple excision is the treatment of choice. Local recurrences are exceptional.

33.6 Pseudotumoral Calcinosis

Pseudotumoral calcinosis (also called tumoral calcinosis, calcifying collagenolysis, calcifying bursitis, tumoral lipocalcinosis, and hip stone disease) is the designation for an extraskeletal soft tissue hydroxyapatite calcification with a granulomatous response that develops in patients with secondary hyperparathyroidism or hypercalcemia, usually idiopathic or because of end-stage kidney disease [5].

33.7 Hibernoma

Hibernoma is a benign adipocytic tumor characterized by a brown fat cell component variably intermingled with mature white adipose tissue. This tumor, which usually presents as a painless, slow-growing mass, tends to occur in the subcutis of young adult with a predilection for the thigh, followed by the trunk, the upper limbs, and the head and neck area [6]. The main differential diagnosis is with atypical lipomatous tumor with hibernoma-like features: the absence of MDM2 amplification by FISH associated with an immunohistochemical negativity for MDM2/CDK4 in hibernoma can help to differentiate these entities. Local excision is curative.

References

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