Chapter 58
Pigmented Villonodular Synovitis (PVNS) and Giant-Cell Tumor of Tendon Sheath

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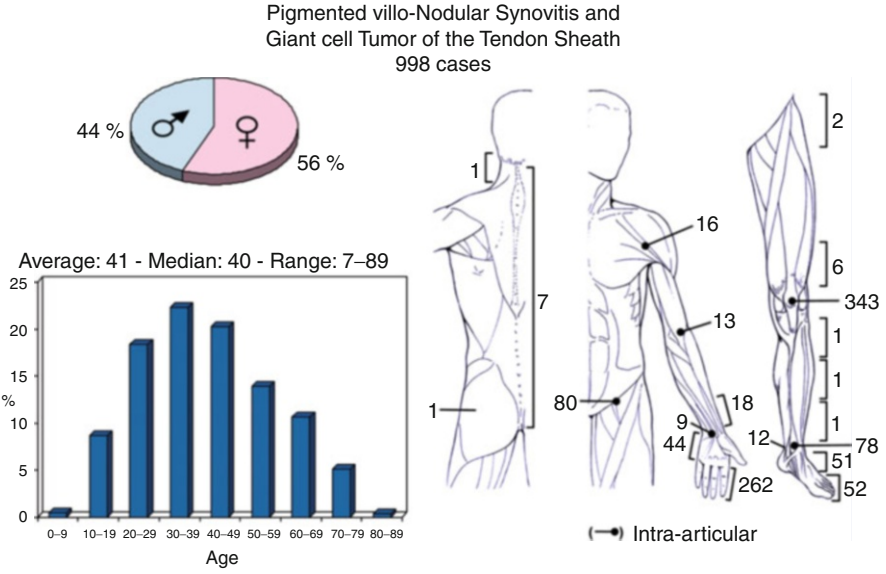
Definition: Benign, composed of synovial-like mononuclear cells of the joint, the tendon sheaths, and the mucous bursae.
Epidemiology: No sex predilection. 20–40 years of age.



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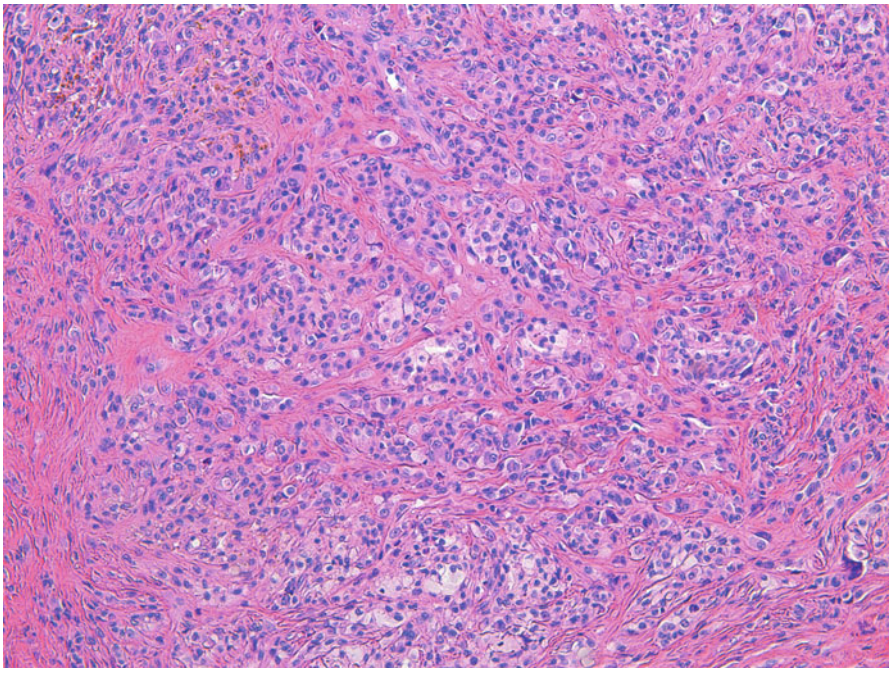
Location: (a) Paratendinous (frequent): in the sheath of a flexor tendon of the fingers, in the palm of the hand close to a metacarpophalangeal joint, in the wrist, on the dorsum of a finger adjacent to an extensor tendon, and rare in the foot; (b) in the joint (rare): >75 % in the knee, then in the hip, wrist, ankle, and shoulder; and (c) in the bursae (exceptional).
Clinical: Pain, swelling, effusion, or completely asymptomatic.
Diagnosis: On x-ray thickening of the synovial membrane with the same density of the soft tissue and without calcification. Skeletal erosions due to compression are frequent as rounded or multilobulated osteolytic lesions with well-defined margins, on the perimeter of the joint. Erosions are multiple and superficial, with sclerotic edges. On CT—lobulated newly formed tissue in the joint with considerable uptake of contrast dye. On bone scan—bone uptake and increased flow and blood pool in the mass. On MRI—heterogeneous, mostly low signal on T1 and T2 is characteristic. Intra- and peritumoral enhancing curvilinear regions on contrast T1.
Histopathology: Roughly lobulated, single, soft, yellow-white to pale-brown nodules with smooth surface. In advanced stages it matures in a fibrous scar. It becomes hard, compact, and white with some yellow or brown bands and adheres to the surrounding tissues, to bone, and to tendon. The synovial membrane appears thickened, leathery yellow, and matted by long large villi like a "ruffled beard," with multiple, soft, yellow-brown, lobulated nodules of varying size. Fibrin membranes cover the villi surface. Soft, pasty, friable, and yellow-brown tissue fills joint space in more advanced lesions. Pathologic tissue may be easily enucleated from the bone lesions that have a smooth bony wall. PVNS may invade the joint capsule and expand into the muscles, between the tendons, but it never infiltrates them.
Course and Staging: Slowly growing, it may be an aggressive lesion. Usually, stage 2, but also stage 3 in late cases of enormous size.
Treatment: Marginal excision is easy and adequate. In PVNS of the joint, surgery is difficult and sometimes it is impossible to eradicate the whole lesion. A wide synovectomy is indicated, but often local recurrence occurs. Radiotherapy is useful as adjuvant treatment when complete excision is not feasible. When the disease destroys the joint cartilage, an arthrodesis or prosthesis may be required.

Radiograph and T2 coronal MR image. Hip lesion with multiple erosions of the bone of both parts of the joint. Lesions have a low signal on T2 image due to iron deposits



Sheets of synovial mononuclear cells with abundant hemosiderin pigment and scattered groups of foam cells. Vaguely nodular pattern on panoramic view

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