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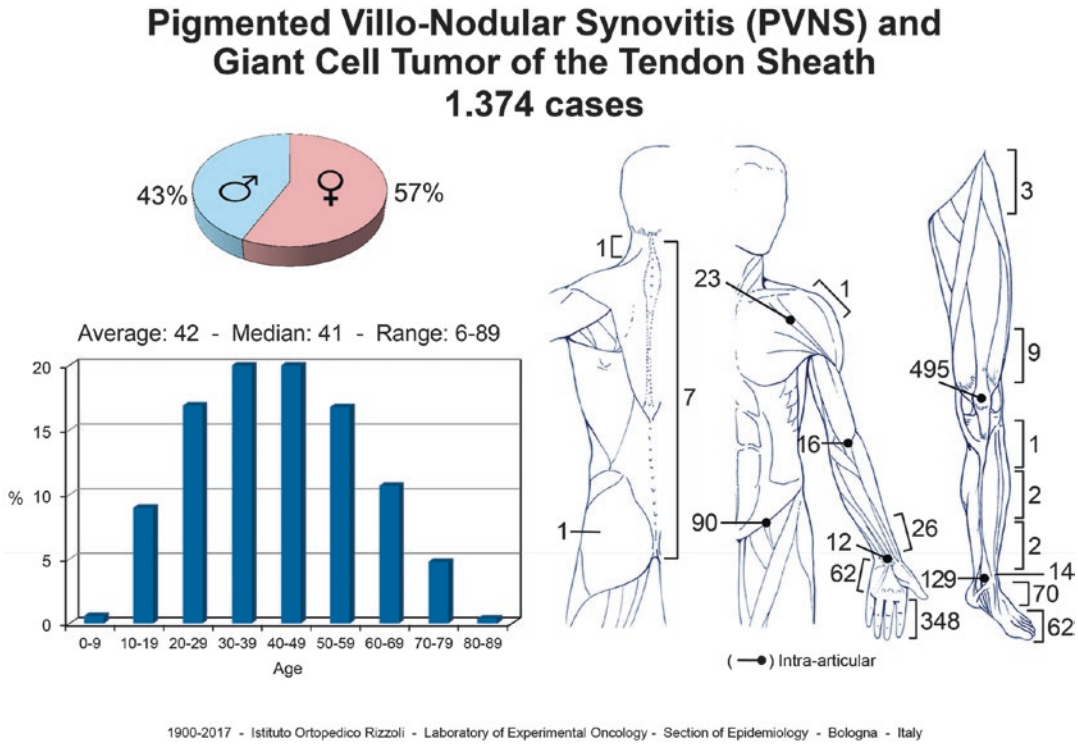
Pigmented Villonodular Synovitis and Giant Cell Tumor of the Tendon Sheaths (Tenosynovial Giant Cell Tumor)

Eric L. Staals

Definition: Benign neoplasm, composed of synovial-like mononuclear cells, creating a progressive inflammatory process in the joint, tendon sheaths, or bursae.

Epidemiology: No sex predilection. 20–40 years of age.



E. L. Staals (✉)
Orthopedic Oncology Department, IRCCS Istituto Ortopedico Rizzoli, Bologna, Italy
e-mail: ericlodewijk.staals@ior.it

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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, rare in the foot; (b) in the joint (rare): >75% in the knee, then in the hip, wrist, ankle, shoulder; (c) in the bursae (exceptional).

Clinical: The diffuse type, creating multiple nodules and usually involving the whole joint, often causes pain, swelling and stiffness. On the long term, degenerative changes and secondary osteoarthritis are frequently seen. The localized type is usually a single nodule and may cause blocking, clicking, and sometimes swelling of a joint. Pain is generally mild to moderate and if the nodule is small, this type can be (almost) asymptomatic.

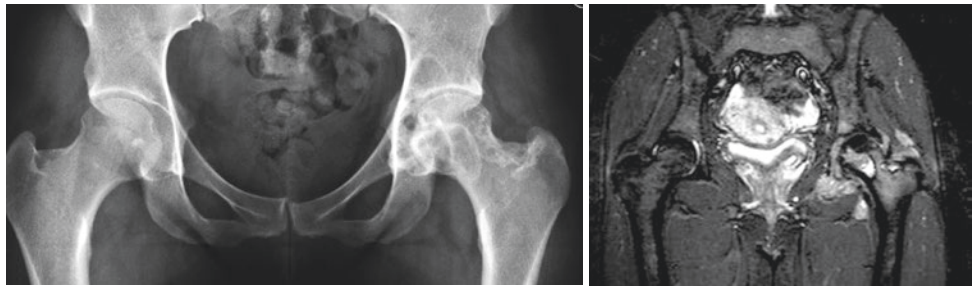
Diagnosis: On X-ray joint effusion may be detectable, with thickening of the synovial tissue. Skeletal erosions due to long standing nodular lesions creating osteolytic lesions with well-defined sclerotic margins, on the perimeter of the joint. On CT—lobulated newly formed tissue in the joint with considerable contrast enhancement. On bone scan—uptake may be increased due to bone compression/erosion. On MRI—heterogeneous, mostly low signal both in T1 and T2 is characteristic. Intra and peritumoral enhancing curvilinear regions on contrast T1. A PET-scan typically shows very high SUVmax values in TGCT, mimicking a malignant process.

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, white with some yellow or brown bands and adheres to the surrounding tissues, to bone, to tendon. The synovial membrane appears thickened, leathery-yellow, 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. Histologically, sheets of mononuclear cells with plasmacytoid features, dense intercellular collagen, multinucleated giant cells, hemosiderin pigment, and scattered groups of foam cells.

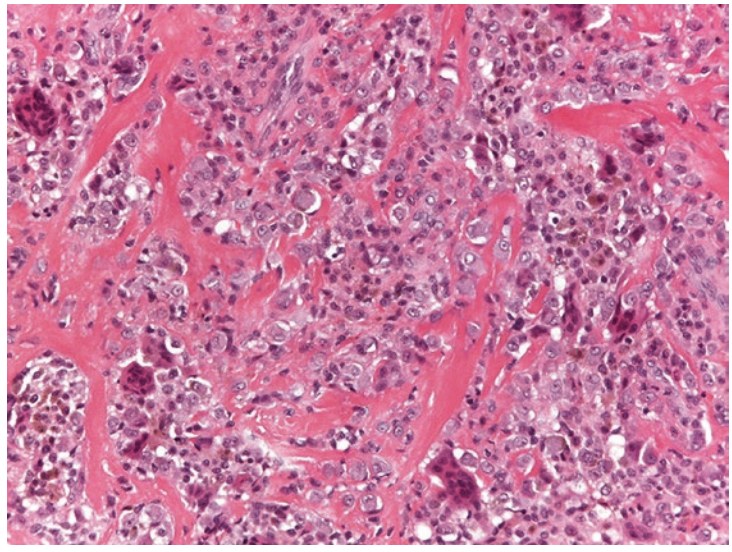
Soft, pasty, friable, and yellow-brown tissue fills joint space in more advanced lesions. Pathologic tissue may be easily enucleated from the bone lesions that have a smooth bony wall. The diffuse type TGCT may invade the joint capsule and expand into the muscles, between the tendons, but it never infiltrates them. It may dislocate or encase the neurovascular structures.

Course and Staging: The clinical course is rather unpredictable. TGCT is usually slowly growing, and clinical presentation may remain stable for many years. However, recurrent disease is very frequent in the diffuse type and may lead to significant functional loss and impaired quality of life. Malignant TGCT has been reported in exceptional cases.

Treatment: Excision of the nodular type is usually curative and may be performed either through an open excision or arthroscopically. In diffuse type TGCT surgery is difficult and often it is impossible to eradicate the whole lesion. Complete synovectomy is indicated, but postoperative complications are frequent and local recurrence rates are as high as 40%. When the disease destroys the joint cartilage an arthrodesis or prosthesis may be required, and in rare cases amputation is necessary. External or intrarticular radiotherapy has been suggested as adjuvant treatment when complete excision is not feasible. Recent studies have underlined the important role of the CSF1-CSF1R pathway in the pathogenesis of TGCT and several target therapies have been proposed with promising short-term results.



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, hemosiderin pigment, scattered multinucleated giant cells, and foam cells. Vaguely nodular pattern on panoramic view

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