

Joint Tumors and Tumor-Like Conditions

Reactive tumor-like lesions, such as ganglions, synovial cysts, and osteochondral loose bodies commonly involve joints and tendon sheaths. They usually result from trauma or degenerative processes and are much more common than neoplasms. Primary neoplasms are rare, usually benign and tend to recapitulate the cells and tissue types (synovial membrane, fat, blood vessels, fibrous tissue, and cartilage) native to joints and related structures. Malignant tumors are rare; these are discussed later with soft tissue tumors.

Ganglion and Synovial Cysts

A *ganglion* is a small (1 to 1.5 cm) cyst that is almost always located near a joint capsule or tendon sheath. A common location is around the joints of the wrist, where it appears as a firm, fluctuant, pea-sized translucent nodule. It arises as a result of cystic or myxoid degeneration of connective tissue; hence the cyst wall lacks a cell lining. The lesion may be multilocular and enlarges through coalescence of adjacent areas of myxoid change. The fluid that fills the cyst is similar to synovial fluid; however, there is no communication with the joint space. Despite the name, the lesion is unrelated to ganglia of the nervous system.

Herniation of synovium through a joint capsule or massive enlargement of a bursa may produce a *synovial cyst*. A well-recognized example is the synovial cyst that forms in the popliteal space in the setting of rheumatoid arthritis (*Baker cyst*). The synovial lining may be hyperplastic and contain inflammatory cells and fibrin.

Tenosynovial Giant Cell Tumor

Tenosynovial giant cell tumor is the term for several closely related benign neoplasms that develop in the synovial lining of joints, tendon sheaths, and bursae. Clinical variants of tenosynovial giant cell tumor include the *diffuse type* (previously known as *pigmented villonodular synovitis*), and the *localized type* (also known as *giant cell tumor of tendon*

sheath). Whereas the diffuse form tends to involve large joints, the localized type usually occurs as a discrete nodule attached to a tendon sheath, commonly in the hand. Both variants usually are diagnosed in the 20s to 40s and affect the sexes equally.

Pathogenesis. These tumors harbor a reciprocal somatic chromosomal translocation, $t(1;2)(p13;q37)$, resulting in fusion of the type VI collagen α -3 promoter upstream of the coding sequence of the monocyte colony-stimulating factor (M-CSF) gene. As a result, the tumor cells overexpress M-CSF, which, through autocrine and paracrine effects, stimulates proliferation of macrophages, in a manner similar to giant cell tumor of bone (described previously).

MORPHOLOGY

Tenosynovial giant cell tumors are red-brown to orange-yellow. In diffuse tumors the normally smooth joint synovium is converted into a tangled mat by red-brown folds, finger-like projections, and nodules (Fig. 26-49A). In contrast, localized tumors are well circumscribed. The neoplastic cells, which account for only 2% to 16% of the cells in the mass, are polygonal, moderately sized, and resemble synoviocytes (Fig. 26-49B). In the diffuse variant they spread along the surface and infiltrate the subsynovial issue. In nodular tumors, the cells grow in a solid aggregate that may be attached to the synovium by a pedicle. Both variants are heavily infiltrated by macrophages, and may contain hemosiderin or foamy lipid. Scattered multinucleated giant cells and patchy fibrosis are commonly present.

Clinical Features. Diffuse tenosynovial giant cell tumor presents in the knee in 80% of cases, followed in frequency by the hip, ankle, and calcaneocuboid joints. Affected individuals typically complain of pain, locking, and recurrent swelling similar to monoarticular arthritis. Tumor progression limits the range of movement of the joint and causes it to become stiff and firm. Sometimes a palpable mass is appreciated. Aggressive tumors erode into adjacent bones and soft tissues, causing confusion with other types of

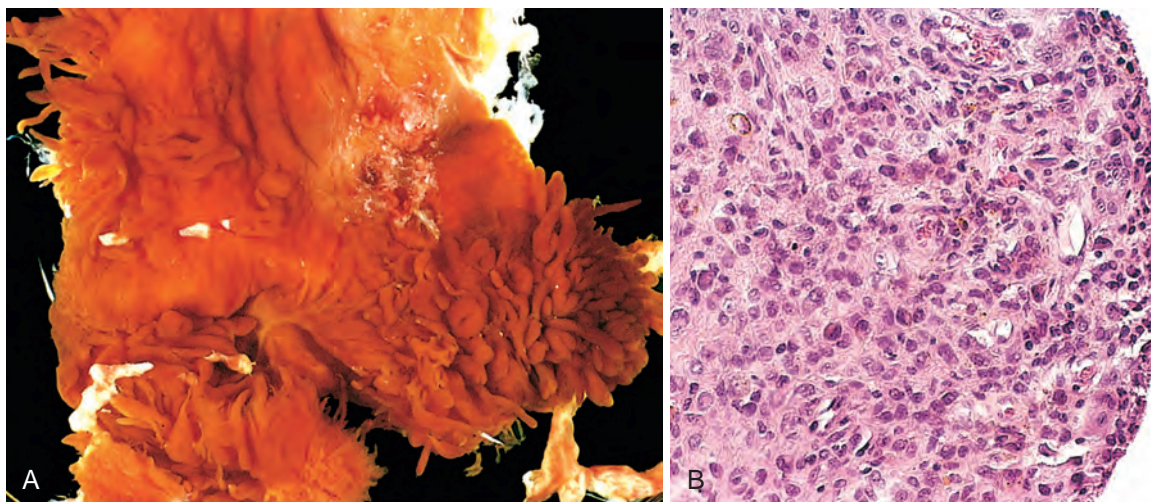


Figure 26-49 Tenosynovial giant cell tumor, diffuse type. **A**, Excised synovium with fronds and nodules typical of pigmented villonodular synovitis (arrow). **B**, Sheets of proliferating cells in tenosynovial giant cell tumor bulging the synovial lining.