Pigmented Villonodular Synovitis

The etiology of pigmented villonodular synovitis (PVNS) remains controversial. It presents as an inflammatory synovial disease, usually involving only one joint, but histologically the disease demonstrates histiocytic proliferation in the subsynovial tissue that takes on characteristics of a neoplastic condition similar to a giant cell tumor. PVNS occurs typically in the subsynovial tissue about major joints of the lower extremity in patients between the ages of 20 and 40 years. The knee joint is the most common site, followed next by the hip, ankle and foot. It is rare to see this disease in the upper extremity. The histiocytic proliferation in subsynovial tissues is similar to that seen in giant cell tumor of tendon sheathes in the hand and foot. The clinical picture in the knee joint is that of spontaneous swelling associated with pain and synovial hypertrophy. Hemarthrosis can result in massive swelling about the knee joint and can occasionally result in juxta-articular erosion of bone, similar to what is seen in rheumatoid synovitis. Clinical conditions with a similar presentation include hemophilia and coccidioidomycosis. In fewer than ten per cent of cases this condition will present as a localized focal mass in the suprapatellar pouch of the knee or high in the popliteal space posteriorly and can masquerade as a neoplastic condition such as a synovial sarcoma.

Treatment for the more generalized synovial involvement of the knee joint or other lower extremity joints consists of a subtotal synovectomy often performed through an arthroscope. In more extensive cases an open procedure may be necessary. The recurrence rate is fairly high, in the range of 30 per cent. In cases where multiple recurrences result, treatment with external beam radiation therapy in the neighborhood of 1500 – 3000 rads is used. Injectable isotopes have also been used for radiation treatment of recurrent cases. Secondary arthritic changes, especially in the knee joint, can occur as a late complication of this disease and these changes could lead to a total joint replacement at the age of 50 or 60 years. On very rare occasions, this disease can convert to a neoplastic sarcoma with a high degree of giant cell activity. This is similar to the conversion of a giant cell tumor to a malignant sarcoma.