Pigmented villonodular synovitis

Definition
A locally aggressive synovial tumour which affects both large joints and tendon sheaths (Bullough and Vigorita).
Same entity as giant cell tumour of tendon sheath.

Terminology
Other names for this disorder have included synovial xanthoma, synovial fibroendothelioma, synovial endothelioma, benign fibrous histiocytoma, xanthomatous giant cell tumour, myeloplaxoma, chronic haemorrhagic villous synovitis, etc

Epidemiology
1.8 per million.
No sex or racial predilection.
Most common in the 20s and 30s.

Aetiology
Essentially unknown.
Theories:
1. Hirohata – localised disturbance in the metabolism of lipids.
2. Trauma.
    a. However, low incidence in patients with haemorrhagic disorders, and has not been consistently reproduced in experimental animals.
3. Inflammation
    a. Widest held theory since 1941 (Jaffe).
    b. Trigger for inflammation has not been identified.

Pathology
Gross
1. Tan colour
2. In the knee usually consists of multiple nodules, often with dramatic associated hyperplastic villous changes in the synovium, giving a straggly beard appearance.
3. Extensive haemosiderin deposition.

Microscopic
1. Proliferating, collagen producing polyhedral cells
2. Often scattered multinucleated giant cells
3. Foam cells
4. Haemosiderin

Clinical presentation
1. Much more frequently found as a solitary nodule and more rarely as a diffuse multinodular condition
2. Most common sites are the knees and fingers
3. Can also occur in the wrist, hip, ankle and toes
4. Usually painless or only mildly painful. Onset is insidious
5. Approximately 50% can recall an episode of trauma

Signs
1. Local warmth
2. Swelling
3. Stiffness
4. Palpable mass
5. Point tenderness in 50%

Investigation

1. Aspiration produces a deep xanthochromic to brownish stained bloody fluid
2. Increase in WCC
3. Depressed level of serum cholesterol

Radiology

1. In the finger there is usually only soft tissue swelling or there may be cortical erosion
2. In the knee the major finding is soft tissue swelling which may be massive
3. Erosion may cause a lytic intramedullary lesion. Erosion is rare in the knee because a substantial bulk of tumour can be accommodated, but is more common in the hip.
4. Lucencies on either side of a joint are very characteristic of PVNS
5. Findings of extension outside the joint, calcification or cortical destruction suggest the diagnosis of synovial sarcoma
6. MRI findings are low to intermediate signal intensity on T1 and T2 weighted images.

Treatment

1. Excision
    a. Can be arthroscopic, or open using anterior and posterior approaches. In the posterior approach, a lazy S incision is made, the gastrocnemius heads are released and the capsule opened using two T shaped incisions, one medial and one lateral. The menisci are detached both anteriorly and posteriorly and are later repaired.
    b. Recurrence is common in the diffuse form of the disease
        E.g. 33% in a series of 18 patients with PVNS of the knee
    c. In the localised form recurrence is uncommon
2. Radiation synovectomy
    a. Radiocolloid yttrium-90 is injected into the knee
    b. Doesn't appear to affect the knee cartilage.
3. External beam irradiation may be used. One dosage that has been used is 35Gy. Tends to be used in recurrences.
4. End stage destruction, particularly in the ankle, often requires arthrodesis

Natural history

Metastatic disease may develop (e.g. to lungs in the Chin 2002 JBJSA article).

MRI is effective in detecting recurrence postoperatively.