A rare case of wrist diffuse type of tenosynovial giant cell tumour

Clinical History:

A 66-year-old male patient presented with a 10-month history of insidious onset of right wrist swelling and intermittent pain. He had no history of bleeding disorder or trauma. Physical examination revealed right dorsal wrist swelling with mild impaired extensor function of ring and little fingers.

Imaging Findings:

Plain radiographs demonstrated erosion of distal radioulnar joint and ulnocarpal joint. Marked soft tissue swelling was seen at the dorsal side of wrist at carpal level.

Ultrasound revealed increase in soft tissues with low to immediate echo in the ulnocarpal joint, extending to dorsal aspect with involvement of overlying extensor tendons. This soft tissue showed mild hypervascularity with Doppler study. There was also extension to the dorsal aspect with encasement of extensor tendons at wrist and carpal bones levels, as demonstrated by ultrasound scan.

MRI scan revealed joint effusion, mainly involving ulnocarpal and distal radioulnar joints. There is T1-weighted and T2-weighted low-signal-intensity rim with nodular thickening seen along the distended joint capsule, suggestive of haemosiderin deposits. The post contrast scan with fat saturation revealed thickened and enhancing synovium.

Overall findings were suggestive of right wrist tenosynovial giant cell tumour, diffuse type (pigmented villonodular synovitis, PVNS) with involvement of overlying extensor tendons.

Discussion:

Tenosynovial giant cell tumour, diffuse type, also called pigmented villonodular synovitis (PVNS), represents a rare benign neoplastic process affecting synovial membranes of joints and bursa. It occurs predominantly in patients aged between the 2nd to the 5th decade. The classical presentation of disease is joint swelling, pain and joint dysfunction secondary to destruction. Joint effusion commonly co-exists. Malignant transformation is very rare.

Tenosynovial giant cell tumour is currently classified into localised type and diffuse type according to WHO.
classification of Tumours of Soft Tissue and Bone[1]. The site of involvement of PVNS can be intra-or extraarticular.
The classical PVNS mainly involves intraarticularly, usually mono-articular involvement.
The common sites of involvement are large joints such as knee (66-80%) and hip joints (4-16%). Intraarticular
involvement of wrist in PVNS is very rare. [2]

PVNS can also be extraarticular in location, mainly involving the hand and foot regions. It has a slight female
predominance. Most of them are periarticular in location but purely intramuscular or subcutaneous lesions were also
reported. Other nomenclatures to describe extraarticular lesions had been used based on the location of
involvement. For example, involvement of bursa is called pigmented villonodular bursitis (PVNB) while involvement
to tendon sheath was called pigmented villonodular tumour of tendon sheath (PVNTS) or giant cell tumour of the
tendon sheath (GCTTS). These could not well be distinguished from the localised type of tenosynovial giant cell
tumour which was also called giant cell tumour of tendon sheath or nodular tenosynovitis both clinically or
radiologically.

This case is particularly rare in terms of predominant intraarticular involvement with bone erosions in a rare location
of the wrist, with extraarticular encasement of adjacent tendons leading to dysfunction. There are only reported case
reports of the disease at flexor tendon sheath causing an unusual cause of carpal tunnel syndrome [3] and bone
erosion [4].

Radiographs of PVNS can show joint effusion and articular erosions without calcification. Ultrasound can show joint
effusion and thickened hypervascular synovium. It can also show heterogeneous lesion with variable echogenicity
depending of amount of haemosiderin present.

MRI can reveal joint effusion and synovial thickening. Key to diagnosis is articular erosion with presence of
haemosiderin (T1 and T2-weighted hypointense). Additional sequence with gradient echo with blooming can further
aid the diagnosis. Variable degree of enhancement is present in post-contrast-images.

Patient underwent complete synovectomy with tendon grafting. The diagnosis is confirmed histologically.
Recurrence rates after total synovectomy are reported to be ~15% (range 7-20%)[2].

**Differential Diagnosis List:** PVNS of wrist with unusual both intra-and extraarticular involvement, Haemophilic
arthropathy, Haemorrhagic synovitis, Rheumatoid arthritis, Hypertrophic synovitis

**Final Diagnosis:** PVNS of wrist with unusual both intra-and extraarticular involvement

**References:**

Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F (Eds) (2013) WHO Classification of Tumours of Soft Tissue
and Bone. IARC: Lyon 2013: 100-103 Fourth Edition
Figure 1

Description: Plain radiograph shows articular erosion of distal ulna and distal radioulnar joint. Origin: Department of Radiology, Tuen Mun Hospital
**Description:** Heterogeneous hypoechoic soft tissue with increased vascularity in Doppler study, showing encasement of extensor tendons. **Origin:** Department of Radiology, Tuen Mun Hospital
Description: T1-weighted intermediate signal involving the ulnocarpal joint with encasement of adjacent extensor tendon dorsally. Origin: Department of Radiology, Tuen Mun Hospital
Description: T2-weighted hyperintense joint fluid with haemosiderin deposit. (white open arrow) and erosion of distal radioulnar joint (white solid arrow) Origin: Department of Radiology, Tuen Mun Hospital
Description: Intra-operative photo revealing dark intra-articular soft tissue (arrow), which is a classical finding in diffuse type of tenosynovial giant cell tumour (PVNS). Origin: Department of Orthopaedics & Traumatology, Tuen Mun Hospital
Description: Histology shows both villous change and haemosiderin-laden macrophages, suggestive of a diffuse type of tenosynovial giant cell tumour (PVNS). Origin: Courtesy to Dr. Loo, Department of Pathology, Tuen Mun Hospital