PIGMENTED VILLONODULAR SYNOVITIS (PVNS)

I@OP CASE OF THE WEEK

48-year-old female history of right knee OA and multiple loose bodies. RO loose bodies two months ago?



Fig.1. Coronal PD SPAIR of the knee with multi foci of high signal lobulated masses.

Fig.2. Sagittal PD SPAIR of the knee demonstrates masses causing erosion on the posterior surface of the tibial plateau with multi focal low signal from hemosiderin deposition.



Fig.3. Coronal PD SPAIR with heterogenous high signal lobulated masses in the inter condylar space & beneath the MCL Fig.4. Coronal illustration of PVNS in a knee with associated erosions

MRI Findings:

- Mass-like synovial proliferation with lobulated margins.
- Extensive in the diffuse form or limited to a well-defined single nodule in the localized form with low signal intensity due to hemosiderin deposition.
 - T1: low to intermediate signal
 - T1 C+ (Gd): variable enhancement
 - T2: low-intermediate signal
 - STIR: predominantly high signal 2
 - GE: low and may demonstrate blooming

Discussion:

- Overview
 - Rare an idiopathic monoarticular neoplastic synovial disease
 - \circ $\;$ Characterized by exuberant proliferation of synovial villi and nodules
 - o Affecting synovial membranes of joints, bursae or tendons resulting from probable
 - neoplastic synovial proliferation, villous & nodular projections and hemosiderin deposition.
- Epidemiology
 - o demographics
 - most commonly in adults age 30-40 but can occur at any age
 - equal incidence in men & women
- Classification:
 - localized (intra-articular or classic form)
 - knee is most common site (80%)
 - knee > hip > ankle > shoulder > elbow
 - o diffuse (extra-articular extension): behaves differently from localized
 - o giant cell tumour of tendon sheath: tendon sheaths of hands and feet
- Aetiology
 - thought to be neoplastic in origin
 - autonomous growth
 - malignant transformation potential
 - recently found constant cytogenetic aberrations
 - Previously thought to be chronic inflammation or trauma-induced haemorrhage.
- Genetics
 - 5q33 chromosomal rearrangement. Increased expression of CSF1 gene
 - Clinical
 - o History
 - 50% of patients will have a prior history of trauma to the area
 - o Symptoms
 - Pain & swelling, normally present for months before Dx
 - mechanical pain & limited ROM
 - recurrent atraumatic hemarthrosis is hallmark of disorder
 - Unusual in paediatric population, sometimes seen & more frequently poly-articular. Also been described in association with:
 - Cherubism, extremity lymphedema, mandibular lesions & Noonan syndrome
- Differential diagnosis
 - o Synovial chondromatosis, Haemophilia, Inflammatory synovitis
- Management
 - Nonoperative: observation
 - Operative
 - total synovectomy: knee anterior arthroscopic or posterior open
 - marginal excision
 - o intra-articular disease
 - arthroscopic partial synovectomy or open total synovectomy: extent & location
 - frequent recurrence is common due to incomplete synovectomy
 - o extra-articular
 - marginal excision is adequate for tendon giant cell tumour
 - external beam irradiation
 - combined with total synovectomy, reduces recurrence to 10-20%

Reference & Further reading:

Murphey MD, Rhee JH, Lewis RB et-al. Pigmented villonodular synovitis: radiologic-pathologic correlation. Radiographics. 28 (5): 1493-518. Masih S, Antebi A. Imaging of pigmented villonodular synovitis. Semin Musculoskelet Radiol. 2003;7 (3): 205-16.

Bravo SM, Winalski CS, Weissman BN. Pigmented villonodular synovitis. Radiol. Clin. North Am. 1996;34 (2): 311-26, x-xi.

Barile A, Sabatini M, Iannessi F et-al. Pigmented villonodular synovitis (PVNS) of the knee joint: magnetic resonance imaging (MRI) using standard and dynamic paramagnetic contrast media. Report of 52 cases surgically and histologically controlled. Radiol Med. 2004;107 (4): 356-66.

https://www.orthobullets.com/pathology/8050/pigmented-villonodular-synovitis