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A Rare Case of Pigmented Villonodular Synovitis of the Wrist Joint

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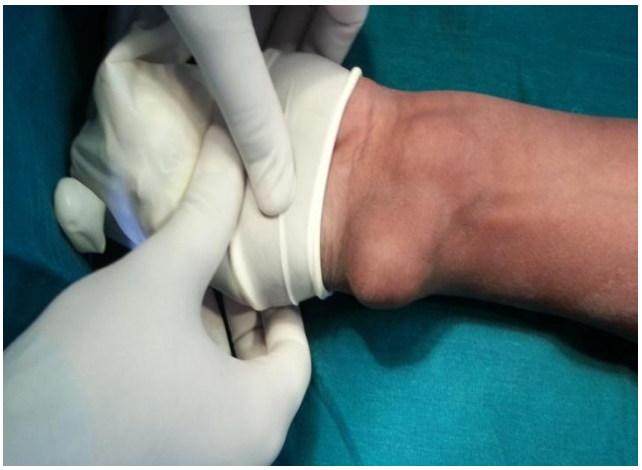
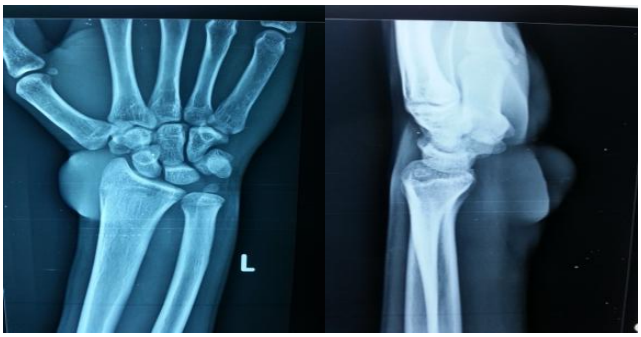
Abstract: ***Introduction:*** ***Pigmented villonodular synovitis is a benign lesion which may be located in joints, tendon sheaths or bursae. The most common sites are the knee and fingers. These account for about 75% of cases. A review of medical literature reveals that PVNS of the wrist with the involvement of carpal bones is rare.[4]*** ***Case Report History:*** ***17 year old, right hand dominant male, presented with chief complaints of swelling over left wrist since 3 years. The swelling was spontaneous and insidious in onset, initially the size of a peanut, gradually progressing to the present size of a big lemon, not associated with pain or any discharge from the swelling. There was no h/o trauma. He denied any constitutional symptoms.*** ***Examination:*** ***Single swelling oval shaped around 7 x 4 cm over volar aspect of left wrist and distal forearm, lobulated surface, without local rise of temperature, non tender, well defined margins, fluctuant, weakly transilluminant, in the subcutaneous plane, transversely mobile with the overlying skin being normal. Radial artery pulsation felt feeble compared to other side. Range of motion at wrist was full and painless. No neurological deficits in median or ulnar nerve distribution.***

Keywords: Pigmented villonodular synovitis; benign lesion; volar aspect of left wrist; hyerintense lesion; surgical or arthroscopic synovectomy

1. Intraoperative Images



After Draping



X ray showed no bony involvement

Figure 1: Preoperative X-ray

2. MRI

Showed a hyerintense lesion seen extending from subcutaneous fascia to the space posterior to the flexor tendons, medially upto lower end of radius and laterally forming a bulge beyond the radial shaft, inferiorly upto first metacapo-phalangeal joint. No extension into carpal tunnel with subtle erosion of anteroinferior margin of radius.

3. Surgery

With the provisional diagnosis of tubercular tenosynovitis/pigmented villo nodular synovitis, the patient was taken up for surgery.

- Using volar approach, skin flaps were raised.
- Sheath of the swelling was identified.
- Palmaris longus was found free.

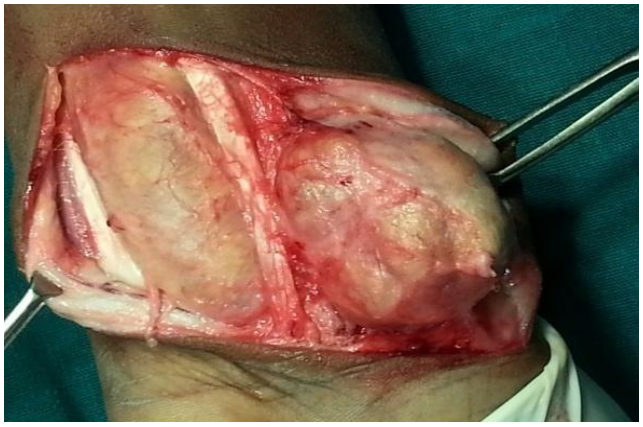


Figure 2: Exposure of the swelling

- Using blunt dissection, margins of swelling were isolated.
- The swelling was found to encompass flexor carpi radialis, flexor digitorum superficialis and profundus.

- Radial artery was found to enter the substance of the swelling through and through, which was dissected free from the swelling and was protected.

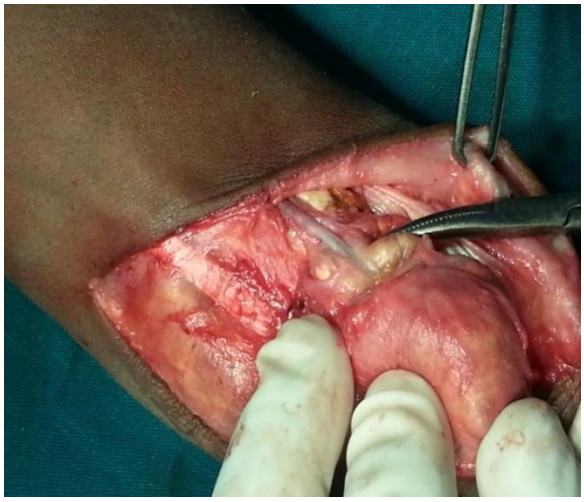


Figure 3: Radial Artery Isolated

- Pronator quadratus muscle was involved and the swelling was adhered to periosteum of radius.
- Medially, ulnar artery and nerve was found free.
- Median nerve was found adhered to the lesion deep to it.
- The nerve was freed and was protected.
- Swelling was excised in situ.

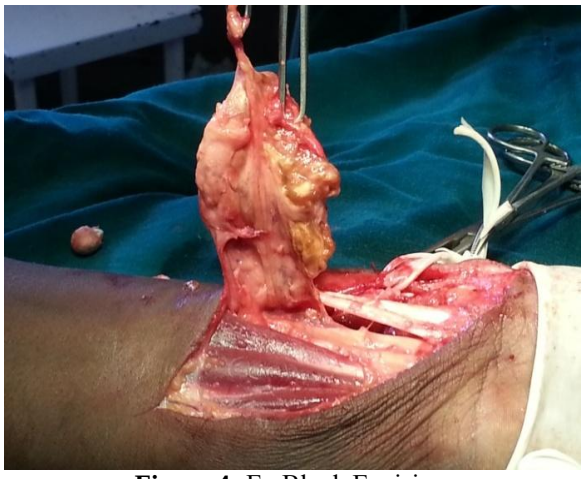


Figure 4: En Block Excision

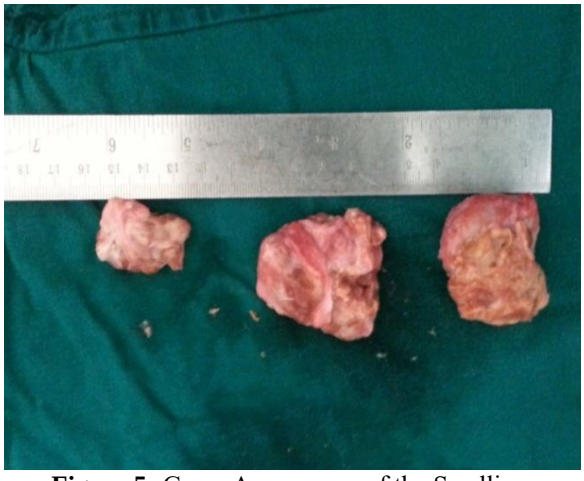


Figure 5: Gross Appearance of the Swelling

Grossly: reddish brown, nodular mass

- The lesion was sent for histopathology and culture.
- PCR for tuberculosis complex : negative
- Histopathology revealed features suggestive of pigmented villonodular synovitis.
- Culture for AFB: no growth after 6 wks.

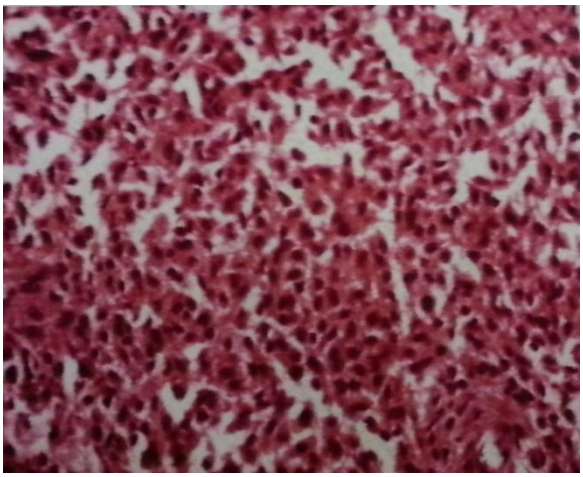


Figure 6: Histopathology

4. Gross

Received two vials: specimen sent in formalin: Unlabelled vials. 1. Received irregular golden brown membranous tissue measuring 5*4*2.5 cms partially covered by fascia. Cut section golden brown spongy with focal ? papillary projections. (3P) 2. Received nodular golden brown firm tissue measuring 4.5*4*3.5 cms, cut sectiongolden brown spongy separated by grey white septae. (3P) (6).

5. Microscopy

Multiple sections studied show a lesion with closely packed medium sized polyhedral cells arranged in sheets and papillary projections. These papillary projections are made up of foamy cells and haemosiderin containing phagocytes. Large clefts and pseudoglandular spaces lined by synovial cells are also present. Also seen are few mixed inflammatory cell infiltrate, focal areas of hyalinization with congested and proliferating blood vessels. Sections are negative for granulomas and malignancy.

6. Impression

Pigmented Villonodular Synovitis.

7. Discussion

PVNS was first described by Jaffe, Lichtenstein, and Sutro in 194.[2]

Pigmented villonodular synovitis (PVNS) consists of proliferating synovial tissue containing histiocytes, fibroblasts, multinucleated giant cells, and capillaries that can destroy dense fibrous tissue, form soft tissue masses, and invade bone.

Like GCT of tendon sheath, PVNS may represent a reactive inflammatory process or a benign neoplasm. It occurs most commonly in adolescents and young adults in large synovial joints including the knee, hip and ankle, although it also occurs in smaller synovial joints, tendon sheaths, and bursae.[1]

It can occur in a focal or diffuse form. When the entire synovium of the joint is affected, and when there is a major villous component, the condition is referred to as diffuse pigmented villonodular synovitis. When a discrete intraarticular mass is present, the condition is called localized pigmented villonodular synovitis.[2] Invasion and erosion of bone in the nodular form is common. In the diffuse form bone and intra articular involvement is less common.[5, 7]

The incidence of PVNS involving joints other than knee and hip is too small to indicate specific principles of clinical diagnosis.[6]
Both the diffuse and the localized form of villonodular synovitis usually occur as a single lesion, mainly in young and middle-aged individuals of either sex. One of the most characteristic findings in PVNS is the ability of the hyperplastic synovium to invade the subchondral bone, producing cysts and erosions. Although the cause is unknown and is often controversial, some investigators have suggested an autoimmune pathogenesis. Trauma is also a suspected cause, because similar effects have been produced experimentally in animals by repeated injections of blood into the knee joint.[2]

Most patients present with a swollen joint and give a history of recurrent effusions. Clinically, PVNS is a slowly progressive process that manifests as mild pain and joint swelling with limitation of motion. Occasionally, increased skin temperature is noted over the affected joint. The knee joint is most commonly affected and 66% of patients present with a bloody joint effusion. In fact, the presence of a serosanguinous synovial fluid in the absence of a history of recent trauma should strongly suggest the diagnosis of PVNS. The synovial fluid contains elevated levels of cholesterol, and fluid reaccumulates rapidly after aspiration. Other joints may be affected, including the hip, ankle, wrist, elbow, and shoulder.[2]

Plain radiographs initially show erosions on both sides of a joint. There is periarticular bone destruction and degenerative joint disease in the later stages.

The MRI appearance of PVNS is a low signal on both T1- and T2-weighted images. Occasionally it presents as solitary or multiple soft tissue nodules near a joint and can resemble GCT of tendon sheath.[1]

Treatment usually consists of surgical or arthroscopic synovectomy.

Most authorities agree that nodular lesions of the upper extremity should be treated by local excision, despite the recurrence rate of upto 48%.

Radiation therapy has been utilised both alone and in combination with synovectomy.[3]

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