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Giant-Cell Tumour of The Tendon Sheath: A Rare Case Report

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ABSTRACT

Giant-cell tumour of the tendon sheath, also called pigmented villonodular synovitis, is a benign tumour with a high incidence of recurrence. The GCTTS is the most common benign neoplasm in the hand after the ganglion cyst. Etiology of the disease is unknown. It is a relatively rare soft tissue tumour, an overall incidence is 1 in 50,000 individuals. There is no certain treatment protocol but complete local excision with or without radiotherapy is the treatment of choice. Local recurrence after excision is approximately 10 % to 20 %. We hereby report a rare case of GCT of tendon sheath over wrist in a 25 year old male patient which was managed successfully by local excision.

KEYWORDS Giant Cell Tumor, Tendon Sheath, Pigmented villonodular synovitis, Extensor Tendon, Local Excision.

Introduction:
Giant-cell tumour of the tendon sheath is a solitary, firm, extra-articular localized, benign, soft tissue tumour that usually presents with painless swelling for several years. It arises from the tendon sheath mostly of small joints of the hands and feet. It is unusual for giant cell tumours to involve larger joints but it can be found around the ankle, knee joints, elbow or hip. Aetiology of the disease is unknown. It is a relatively rare soft tissue tumour, an overall incidence is 1 in 50,000 individuals and usually affects people between 30 to 50 years. It is more often seen in women, the female to male ratio is 3:2. We report here a case of GCT of tendon sheath over wrist which was managed successfully by local excision.

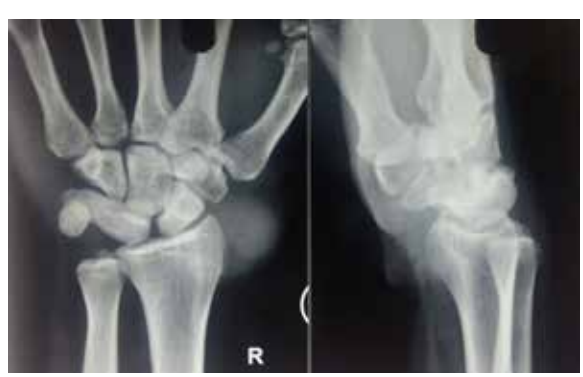
Case report:
A 25-year-old male patient, shopkeeper by occupation presented with complaints of swelling over the dorsal aspect of the right wrist since 2 years and mild pain in swelling since 2 months. The Swelling was insidious in onset, initial size being that of pea nut which gradually progressed to present size. Patient did not complain of swelling in any other part of the body. There was no history of fever, loss of appetite or loss of weight.

The past medical history was insignificant; there was no history of previous surgery, trauma or radiation exposure. The family, occupational, recreational and drug histories were insignificant. The general physical and systemic examinations were within normal limits.

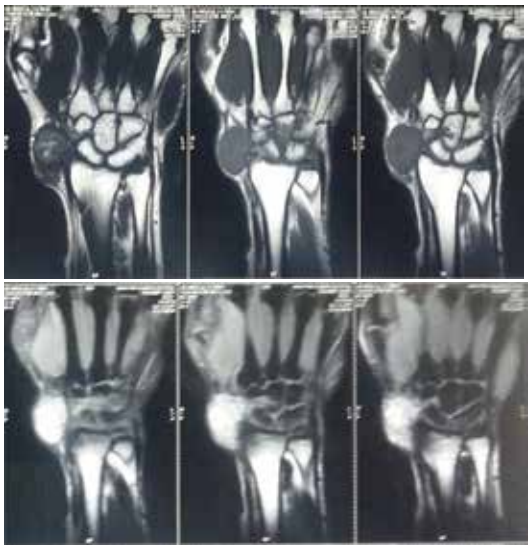
On local examination proper, there was a solitary, swelling measuring 5 x 3 x 4 cm, extending from the lower 1/3rd of forearm to upper half of right hand on the dorsal aspect. Swelling was globular in shape with the overlying skin being normal. Swelling was well defined, firm in consistency with no tenderness and no local rise of temperature. It was non-reducible, non-translucent, and mobile. Movements of wrist were normal with slight limitation of dorsiflexion. Movements of MCP and IP joints were normal. Regional lymph nodes were not enlarged.

Anteroposterior and lateral radiographs of wrist revealed soft tissue swelling over radius on dorsal surface without any evidence of bone involvement.

MRI scan revealed lobulated lesion on radial aspect of wrist joint. Lesion was heterogeneously hypointense on T1W while Hyperintense on T2W and STIR sequences without any evidence of invasion of bone and neuro-vascular structures.



X-Ray of right wrist joint showing soft tissue swelling over bone without any bone involvement.



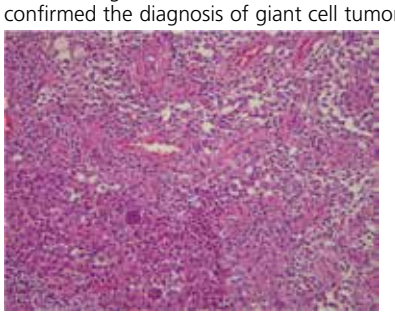
MRI scan showing lobulated lesion which is heterogeneously hypointense on T1W while Hyperintense on T2W and STIR sequences without any evidence of invasion of bone and neuro-vascular structures.

A Histopathological detailing was done with Fine Needle Aspiration Cytology (FNAC) which reported features suggestive of giant cell tumor of tendon sheath and excision biopsy was planned for confirmative diagnosis and definitive treatment.

Intraop course was uneventful and specimen sent for Histopathological examination. Gross examination of the biopsy specimen revealed single nodular grey white mass of tissue measuring 6×3×1.5 cm, external surface nodular, cut section shows homogenous grey white nodular areas.

Intra-operative photograph showing tumour on dorsum of wrist.

Microscopic examination smears showed admixture of osteoclast like giant cells and stromal cells in the background of dense collagenous stroma with few mitotic figures. Findings confirmed the diagnosis of giant cell tumor of tendon sheath.



HPE section showing osteoclasts like giant cells and collagenous stroma with few mitotic figures.

The post operative course was uneventful. The patient was asymptomatic after the surgery and the scar healed well with primary intention. At 12 months follow up, there was no recurrence of growth at the operative site and patient was pain free.

Disscussion :

GCTTS is a relatively rare soft tissue tumour. The most frequent tumour location is a hand (exceeded in that location only by ganglion cyst), especially the fingers [1]. Jaffe, Lichtenstein and Sutro regarded the synovium of the tendon sheath, bursa and joint as an anatomical unit in which giant-cell tumour of the tendon sheath may occur [2].

GCTTS is an extra-articular localized mostly painless soft tissue mass. Patients usually present with painless swelling for several years [3]. The intra-articular form of such tumour is commonly described as pigmented villonodular synovitis and share similar histological characteristics [4].

GCTTS can be divided into localized nodular type (common in hand) and diffuse type (common in joints). Diffuse form is hypercellular with several giant cells, while localized form is relatively hypocellular [5]. Another classification proposed by Al-Qattan classified GCTTS into Type I (single tumour, round and multi-lobulated) and Type II (two or more distinct tumours, not joined together). Type II is more often related with recurrence as satellite lesions when microscopic excision is not done [6, 7].

Etiology of the disease, indeed, is unknown so the tumour is generally considered idiopathic. Some patients have history of trauma, but the reason cannot be determined in most of the cases. There are also some other risk factors such as infection , disorder in the immune system, osteoclastic proliferation, vascular abnormality, localised lipid metabolism disorder, etc [8,9].

Recent studies showed the presence of clonal chromosomal translocation, t(1;2)(p13;q37), which fuses colony stimulating factor (CSF1) coding sequences to the promoter of the collagen type VI alpha-3 gene. As a result, the tumour cells overexpress CSF1, a chemoattractant for macrophages, which infiltrate the tumour in large numbers [10].

According to the World Health Organization classification system for bone and soft tissue tumours, it is classified as a "fibriohistiocytic tumour" [11,12].

If a tumour is preceded by trauma, the patients can have consistent pain at the injury site or gradually growing nodules at those sites. Due to pressure effects, this tumour can cause bone erosions. They have been observed in 9% to 25% of cases and they usually manifest as small cortical defects. Neurological symptoms are not a common feature of GCT-TS. So, if there is pain or neurogenic symptoms with a solitary lesion of the limb, a nerve-sheath tumour or soft-tissue sarcoma should be more suspected in the differential diagnosis [13].

Giant cell tumours are superficial lesions usually located within the few centimeter under skin, first method to diagnose GCTTS could be ultrasound due to its high resolution for small structures such as tendons and ligaments. It can detect whether tumour is solid or cystic and provide information about the tumour vascularity, size and its relation to the surrounding tissue.Close contact with the tendon sheath or joint does make this tumour a primary consideration when diagnosing a soft tissue mass near a tendon sheath or a joint [14].

Radiographs are not so important in making this diagnosis but it will tell us whether there is cortical compression, intraoseal involvement or soft tissue swelling. MRI is of course the most precise procedure where GCTTS is seen as low signal intensity on both T1- and T2-weighted images and it can accurately assess the tumour size and degree of tumour extent which can affect the type of surgical approach. To reach a diagnosis pre-

operatively, fine needle aspiration biopsy is applied [15].

Differential diagnosis include Giant cell tumour of bone, fibroma of tendon sheath, ganglion cyst, desmoid tumour, lipoma, synovial sarcoma, and other soft tissue tumours. There is no certain treatment protocol but complete local excision with or without radiotherapy is the treatment of choice. The tumour should be dissected gently without allowing any seedling. Local recurrence after excision is approximately 10 % to 20 % and according to some authors even 45 %, especially in the hand.

In case of possible incomplete excision, presence of mitotic figures and bone involvement , Kotwal et al. recommended postoperative radiotherapy of 20 Gy in divided daily doses of 2 G y. In their study, recurrence rate was 0% (0 out of 14 patients) [17].

A gene present in normal cells that is responsible for infiltration - nm23 can be used as a prognostic marker for the risk of recurrence. Grover et al. has found that mutation of gene nm23 is associated with increased rate of recurrence [18]. There are also some other complications of treatment such as numbness, joint stiffness, painful scar and skin necrosis [19,20].

Conclusion:

GCTTS is a rare benign soft tissue lesion that arises from the tendon sheath and that is situated extra-articular. It should be considered as a differential diagnosis if the mass is found next to the joint. The most helpful diagnostic procedure is MRI, but to make definite diagnosis histopathological examination is required. Complete excision is the treatment of choice,but tumour has quite high recurrence around 10-20 %.

Conflict of interests:None.

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