Original Research Paper

Volume - 14 | Issue - 06 | June - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Orthopaedics

# TENOSYNOVIAL GIANT CELL TUMOR (TGCT) OF HIP- A RARE CASE REPORT

| | |
|---|---|
| **Dr Ambareesh P** | Associate Professor- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |
| **Dr Supreeth D R** | Assistant Professor- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |
| **Dr Nishanth Reddy J*** | Post Graduate- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). *Corresponding Author |
| **Dr Vishwanath M S** | Professor- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre) |
| **Dr Varun GBS** | Professor- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |
| **Dr Shashank Sonth** | Assistant Professor- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |
| **Dr Harish DV** | Post Graduate- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |
| **Dr Chandan Kumar RV** | Post Graduate- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |
| **Dr Nabhay G** | Post Graduate- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |
| **Dr Mahadeva HN** | Post Graduate- Department Of Orthopaedics (Vydehi Institute Of Medical Sciences And Research Centre). |

**ABSTRACT** Tenosynovial giant cell tumor (TGCT) is a relatively rare inflammatory disease that affects the joint synovium, bursae, and tendon sheaths, leading to nonspecific, often insidious discomfort in the joints. Surgery, with or without adjuvant radiation, is the primary therapy option for tenosynovial giant cell tumor. Here is a report of a case of a 40 year old male presented with symptoms of pain and limp over right leg since 8month. Radiological assessment showed secondary arthritis of right hip with features of avascular necrosis (AVN). Considering clinical and radiological features the patient was planned for uncemented total hip replacement (THA).Intra operatively we observed unusual synovial hypertrophy with yellowish/gray discoloration. Total synovectomy was done and proceded with uncemented THA as per plan. The synovium was sent for histopathological assessment which showed partially encapsulated tumor cells. Post operatively course was uneventful and rehabilitation was done as per standard protocols. Patient is being followed up regularly till date and 6months follow up did not show any signs of recurrence.This observation left us with a conclusion that complete excision of lesion combined with THA can be an effective treatment for Giant cell tenosynovitis.

**KEYWORDS :**

## INTRODUCTION :

WHO Classification of Tumours (2020, 5th edition) categorized tenosynovial giant cell tumors (TGCT) as a group of typical benign lesions with similar pathogenesis and histological features. Synovial hyperplasia and hemosiderin deposition are remarked in tenosynovial giant cell tumors (TGCT) as it is derived from the synovium of joints, tendon sheaths and bursae. Localized-type TGCT (formerly giant cell tumor of tendon sheath/nodular tenosynovitis) and diffuse-type TCGT (formerly pigmented villonodular sclerosis/tenosynovitis/fibrous xanthoma of synovium) are the 2 subtypes of the tumor categorized according to their growth pattern (localized and diffuse) and location (intra- and extra-articular) along with varying clinical courses and symptoms.[1]

TGCT are neoplasms characterized by synovial cell proliferation. However, only a small percentage of cells (2-16%) in the tumor mass carry the neoplastic mutation, with the majority being non-neoplastic reactive cells. This has led to various theories regarding the etiology. TGCT is characterized by chromosomal abnormalities such as trisomy on chromosomes 5 and 7.[2]

In intra-articular forms, the large joints are the most affected- the knee >hip>ankle >shoulder and elbow. Hip is one of the largest and most important joints in the body, responsible for weight bearing and enabling mobility In this case, we will focus on a rare and unusual presentation of TGCT in hip joint.

## CASE DISCUSSION :

A 40 years male presented to orthopedicOPD with complain of pain in right hip joint for 8 months. Pain was insidious in onset, progressive, continuous and aggravated with walking. Pain was gradually progressive in nature. There was no h/o trauma, fever, weight loss and long term drug intake. Pain was associated with limp and difficulty in sitting cross legged and squatting affecting day to-day activities. Physical examination revealed restricted range of movementsin all planes and tenderness over the right hip joint. Plain radiograph of the right hip was advised which showed features of secondary arthritis of right hip with avascular necrosis which is evident by joint space narrowing: (due to the loss of cartilage and joint destruction), Subchondral bone erosion (destruction of the underlying bone beneath the cartilage), Osteophyte formation ( growth of new bone at the margins of the joint as a response to joint degeneration and inflammation),Changes in joint alignment (due to advanced arthritis). Considering clinical and radiological features the patient was planned for uncemented total hip arthroplasty (THA) after pre anesthesia check up. Using the standard posterior approach to access the hip joint and after dissection- intra operatively unusual synovial hypertrophy with yellowish/gray discoloration was observed. Total synovectomy was done and proceeded with uncemented THA as scheduled.

The synovium was sent for histopathological assessment. Post operatively course was uneventful and rehabilitation was done as per standard protocols. Patient is being followed up regularly till date and

1year follow up did not show any signs of recurrence.

Macroscopic findingsof the lesion includedlarger grey white to grey brown tissue mass larger measuring 4 x 3 x 2.5cm withexternal surfaces being grey yellow to grey brown and cut section showing grey yellow to grey brown and smaller sample measuring 2.5 x 1.5 x 1.5cm withgrey yellow to grey brown external surfaces wheras cut section being grey yellow to grey brown.

On Hand E staining , the microscopic findings showed circumscribed and partially encapsulated tumor composed of sheets of polymorphic histiocytes mainly aggregated of foamy histocytes(xanthoma cells) surrounding fibroblasts with good number of lymphocytic infiltates.Few giant cells, Hemosedrin laden macrophages seen in the periphery of lesion with occasional focus of Hyalinization are also noticed. Included are areas of synovial tissue show hyperplastic epithelium with subepithelial chronic cell inflammatory infiltrate leaving an impression suggestive of Tenosynovial giant cell tumor - Right hip joint.

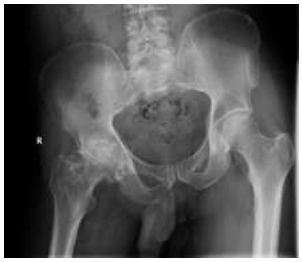

**Fig 1:**Pre-op Xray showing AVN of right Hip ;

**Fig 2:** Intra-op picture showing the sac.

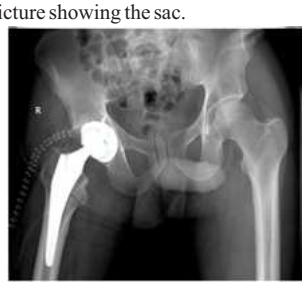

**Fig 3:** Post op Xray after Total Right hip Arthroplasty with excision of TGCT

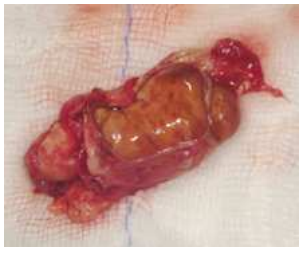

**Fig 4:** Gross specimen showing synovial sac

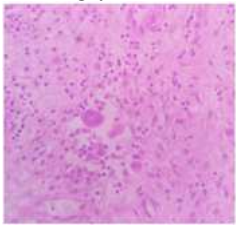

**Fig 5:** H and E 40x multinucleated giant cells with hemosiderin laden macrophages

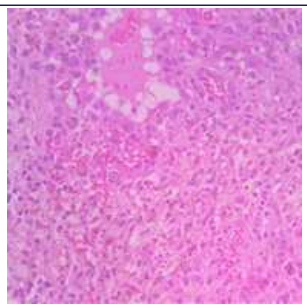

**Fig 6:** H and E 40 x foamy macrophage

**DISCUSSION:**

Tenosynovial giant cell tumors (TGCT), formerly known as pigmented villonodular synovitis, are locally aggressive neoplasms composed of synovial-like mononuclear cells mixed with multinuclear giant cells, foam cells, siderophages, and inflammatory cells. They can be intra- or extra-articular. [3]

A tenosynovial giant cell tumor (TGCT) is a neoplasm that develops from the connective tissue of the bone marrow's non-bone-forming region. The areas that are most prevalent are the distal end of the femur, the middle of the tibia, and the distal end of the radius. Patients who are practically skeletally mature and between the ages of 15 and 40 are most frequently affected by this lesion throughout their third decade of life as described by Jaiswal A et al which is in accordance to the patients age of the present study. [4]

TGCTs generally occur in the fingers, as well as in the ankles, wrists, and small joints of the lower limbs, but rarely in the spine as noticed by Kleinman GM et al [5], Whereas according to Mastboom MJ et al reported that median age at diagnosis was done at 33 years of which majority are female (79.8%), diffuse TGCT (70.3% ) and affected lower extremities (knee 70.9% and hip 9.5% ). [6]In our study pathology is noticed in unilateral hip. One joint is typically affected by TGCT; involvement of multiple joints is highly uncommon. The prevalence is slightly higher in female patients, according to several earlier investigations. However present study reported a male case.

The localised TGCT (TGCT) contained previously reported giant cell tumors in the tendon sheath. Chromosomal translocation occurs when a segment or section of a chromosome separates and reorganizes, causing genetic information to shift and modifying the chromosome set. The translocation responsible for these malignancies is linked to specific locations on chromosomes 1 and 2. This translocation leads cells to overproduce a protein called colony-stimulating factor-1, or CSF-1. The TGCT cells use CSF-1 to recruit white blood cells to incorporate into the tumor, which most likely produces the inflammatory changes seen in these tumors. [4]

Although several symptoms emerged as obvious markers of the disease, there was evidence of diversity in the symptom experience. Not all patients suffer the same symptoms. There is significant variation in how patients perceive symptoms within and between days. The evidence of heterogeneity based on patient reports highlights the importance of using limited recall intervals when measuring these symptoms. According to the findings of a study conducted by Gelhorn HL et al, the most prevalent and relevant symptoms reported by TGCT patients are pain, edema, and stiffness. Reduced range of motion, instability, giving out, and catching were all frequently mentioned. [7]

Clinical measurement of swelling is likely to be more objective; also, numerous patients claimed that swelling as a symptom was important to them because of the resulting discomfort, stiffness, or impairment in physical functioning[7] which is similar to that of our patient who suffered from pain and limp over hip since 8 months and difficulty and impairment of physical activity.

The different types of TGCTs share the same morphological features on microscopy, mainly consisting of large synovial-like monocytes, small mononuclear histiocytes, and osteoclast-like giant cells. Although there is no conclusive evidence regarding the cell of origin of TGCTs, most authors agree that TGCTs originate from fibroblasts and histiocytes of the synovium. West et al determined that the colony-stimulating factor 1 (CSF1) gene encoding the CSF1 receptor ligand is translocated in only 2% to 16% of tumor cells, suggesting that only a

minority of TGCT cells are tumor cells. By reviewing 81 cases of TGCT, Rao et al demonstrated that the process of diffuse TGCT (also known as PVNS) formation is neoplastic.[8,9]

Although many treatments have been reported, the extensively accepted treatment for TGCT of the knees is surgical resection. Tumor recurrence is the most common complication related to tumor resection, and recurrence rates are reported to be as high as 50% for some patients. Complete resection is an important treatment option for TGCT resection but is difficult to perform in some patients with TGCT in the knees because of the cruciate ligaments and without sacrificing other tissues. Maintaining a balance between retaining the knee structure and removing the TGCT with negative margins is difficult. Because the tumor is rare and nonlifethreatening, clinical evidence regarding TGCT in the knees is lacking. [9]

Patients undergoing one-stage synovectomies achieved an equal range of motion postoperatively, but stayed shorter in the hospital and had fewer complications. Thus, one-stage synovectomies are preferred over two-stage synovectomies if feasible.[10]

Prior trials have shown that synovectomy with THA produces better results than synovectomy alone. Yoo et al. reported no recurrent TGCT in eight instances treated with cementless THA after an average follow-up of 8.9 years.They concluded that THA combined with synovectomy is an appropriate therapy option for TGCT patients with end-stage joint deterioration, as it appears to considerably improve clinical outcomes and prevent disease recurrence. [8,9,10]

The histopathology of Pigmented Villonodular Synovitis (TGCT) of the hip joint often demonstrates synovial proliferation, or aberrant synovial tissue growth. This tissue might manifest as a diffuse, thickenend lining or as nodular lumps within the joint. The synovial tissue is formed from out of various cell types, such as macrophages, fibroblasts, and multinucleate giant cells. The synovial tissue in TGCT is distinguished by an excess of hemosiderin-laden macrophages, which give the tissue a brownish appearance and account for the term "pigmented." This hemosiderin accumulation is caused by bleeding within the synovium, a frequent characteristic of TGCT.[11,12,13]

Under a microscope, the nodular masses in TGCT may appear as well-defined lesions made up of synovial lining cells, histiocytes, and multinucleated giant cells. These nodules have the potential to infect and destroy the surrounding joint structures, resulting in joint pain and discomfort. [14]

## CONCLUSION:

The non-specific symptoms and uncommon occurrence of TGCT of the hip joint may render diagnosis challenging. This case report emphasizes the necessity of including TGCT in the differential diagnosis of hip pain and stiffness in young adults. THA combined with total synovectomy is a successful therapeutic option for hip TGCT, resulting in symptom relief and improved joint function. Long-term follow-up is required to watch for recurrence of the disease.

## REFERENCES


1. Kager M, Kager R, Fałek P, Fałek A, Szczypiór G, Niemunis-Sawicka J, Rzepecka-Wejs L, Starosławska E, Burdan F. Tenosynovial giant cell tumor. Folia Med Cracov. 2022;62(2):93-107. doi: 10.24425/fmc.2022.141702. PMID: 36256897.
2. Cupp JS, Miller MA, Montgomery KD, Nielsen TO, O'Connell JX, Huntsman D, van de Rijn M, Gilks CB, West RB. Translocation and expression of CSF1 in pigmented villonodular synovitis, tenosynovial giant cell tumor, rheumatoid arthritis and other reactive synovitides. Am J Surg Pathol. 2007 ;31:970-6. doi: 10.1097/PAS.0b013e31802b86f8. PMID: 17527089.
3. Report of the World Health Organization technical consultation on prevention and control of iron deficiency in infants and young children in malaria-endemic areas, Lyon, France, 12–14 June 2006. Food Nutr Bull, 2007; 28: 489–S631
4. Jaiswal A, Ambade R. Tenosynovial giant cell tumour of the finger: a case report. Pan Afr Med J. 2023 19;45:49. doi: 10.11604/pamj.2023.45.49.37714. PMID: 37575525; PMCID: PMC10422033.
5. Kleinman GM, Dagi TF, Poletti CE. Villonodular synovitis in the spinal canal: case report. J Neurosurg. 1980;52:846–848.
6. Mastboom MJ, Planje R, van de Sande MA. The Patient Perspective on the Impact of Tenosynovial Giant Cell Tumors on Daily Living: Crowdsourcing Study on Physical Function and Quality of Life. Interact J Med Res. 2018;7:e4. doi: 10.2196/ijmr.9325. PMID: 29475829; PMCID: PMC5845102.
7. Gelhorn HL, Tong S, McQuarrie K, Vernon C, Hanlon J, Maclaine G, Lenderking W, Ye X, Speck RM, Lackman RD, Bukata SV, Healey JH, Keedy VL, Anthony SP, Wagner AJ, Von Hoff DD, Singh AS, Becerra CR, Hsu HH, Lin PS, Tap WD. Patient-reported Symptoms of Tenosynovial Giant Cell Tumors. Clin Ther. 2016 ;38:778-93. doi: 10.1016/j.clinthera.2016.03.008. Epub 2016 Apr 1. PMID: 27041409; PMCID: PMC5469507
8. Rao A, Vigorita V J, "Pigmented villonodular synovitis (giant-cell tumorof the tendon sheath and synovial membrane). A review of eighty-one cases," J Bne Jnt Surg 1984; 66: 76–94.
9. Zheng K, Yu XC, Hu YC, Xu M, Zhang JY. A New Simple and Practical Clinical Classification for Tenosynovial Giant Cell Tumors of the Knee. Orthop Surg. 2022 ;14(2):290-297. doi: 10.1111/os.13179. Epub 2021 Dec 16. PMID: 34914180; PMCID: PMC8867407.
10. Siegel M, Bode L, Südkamp N, Kühle J, Zwingmann J, Schmal H, Herget GW. Treatment, recurrence rates and follow-up of Tenosynovial Giant Cell Tumor (TGCT) of the foot and ankle-A systematic review and meta-analysis. PLoS One. 2021;16(12):e0260795. doi: 10.1371/journal.pone.0260795. PMID: 34855875; PMCID: PMC8638888.
11. Fecek C, Carter KR. Pigmented Villonodular Synovitis. [Updated 2023 Aug 7]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK549850/
12. Sugandhi A, Kondaveeti SKS, Sunder A. A Case of Pigmented Villonodular Synovitis. Cureus. 2022 Jun 15;14(6):e25957. doi: 10.7759/cureus.25957. PMID: 35855244; PMCID: PMC9286003.
13. Choi WS, Lee SK, Kim J-Y, Kim Y. Diffuse-Type Tenosynovial Giant Cell Tumor: What Are the Important Findings on the Initial and Follow-Up MRI? *Cancers*. 2024; 16(2):402. https://doi.org/10.3390/cancers16020402
14. Chen EL, de Castro CM 4th, Hendzel KD, Iwaz S, Kim MA, Valeshabad AK, Shokouh-Amiri M, Xie KL. Histologically benign metastasizing tenosynovial giant cell tumor mimicking metastatic malignancy: A case report and review of literature. Radiol Case Rep. 2019 May 24;14(8):934-940. doi: 10.1016/j.radcr.2019.05.013. PMID: 31193787; PMCID: PMC6542375.