Parul University Journal of Health Sciences and Research
ISSN:2583-990X (E)
https://doi.org/10.62373/PUJHSR.2023.127

Giant Cell Tumor of Soft Tissue Masquerading as a Fibroma - A Rare Clinical Entity

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ABSTRACT

Giant cell tumor of the soft tissue (GCT-ST) is a rare and distinctive entity, separate from its bony counterpart, giant cell tumor of bone. This primary soft tissue tumor is often encountered in middle-aged individuals but rarely in children, with common sites including superficial soft tissue of the extremities. GCT-ST typically presents as a painless, slowly growing mass, emphasizing the importance of complete excision to prevent recurrences and metastasis.
We present a case of GCT-ST in a 7-year-old boy with a nodular swelling on the right hip, initially suspected to be a fibroma. Histopathological examination revealed characteristic features of GCT-ST, including spindle and stellate cells, multinucleated giant cells, and haemosiderin-laden macrophages with metaplastic osseous tissue. This case adds to the limited literature on GCT-ST, highlighting its diverse clinical presentations and challenging diagnostic nature.
The discussion includes a review of reported cases of GCT-ST, which have shown varied clinical presentations and outcomes. Histologically, GCT-ST resembles a giant cell tumor of bone, with positive staining for CD68, SMA, Vimentin, and p63. Imaging studies typically reveal a solid hemorrhagic mass. Differential diagnoses include benign and malignant soft tissue tumors rich in giant cells.
Keywords: Giant Cell Tumor of Soft Tissue (GCT-ST), Rare, Fibro-Histiocytic Tumor Of Intermediate Malignancy, Soft Tissue Tumor, Rare Tumors In Children.

INTRODUCTION:

Giant cell tumor (GCT) of the soft tissue (GCT-ST) is a rarely encountered tumor, not commonly seen in clinical practice. It is a rare primary soft tissue tumor utterly distinct from other giant cell tumors of bone or soft tissue. It is the soft tissue doppelganger of the Giant cell tumor of bone. At present, it is grouped with other *fibro-histiocytic tumors of intermediate malignancy*.

It is a rare tumor, seen usually in people of middle age (usually 40 years and above) and very rarely in children. Common sites of its occurrence include superficial soft tissue of extremities (2/3rds), rarely breast, thigh, trunk, groin, and surgical scars.

It usually presents as a painless, slowly growing mass. However, incomplete excision can lead to recurrences and metastasis, warranting close clinical observation.

History and clinical presentation: A 7-year-old boy presented to the outpatient department of a tertiary care hospital with a history of mildly painful nodular swelling of the right hip noticed incidentally while bathing. As per history, it slowly increased in size during the four months of its existence. Recently, it had started to cause persistent, dull, aching pain. On palpation, there was a small, firm. well-circumscribed, noninflamed nodule approximately 3 x 2 cm in size over the right hip (Fig: 1), which was nontender.

There were no apparent signs of inflammation, and clinical examination ruled out vascular malformations. A working diagnosis of ***fibroma*** was made, and the child was taken up for excision of the nodular lesion as a daycare surgical procedure since it was symptomatic. The specimen was sent for histopathology review.

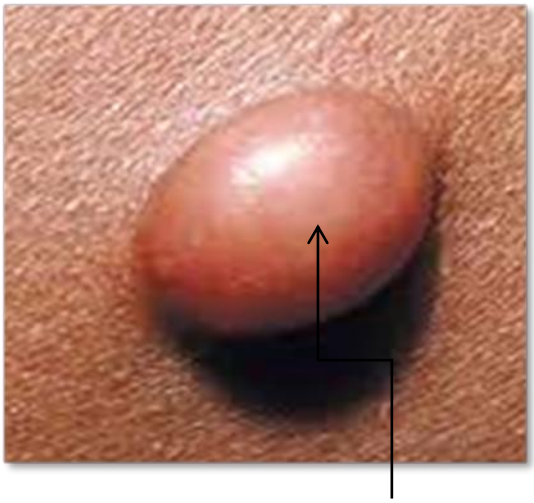

Fig 1: A smooth pinkish, non inflamed nodule over the right hip of approx 3x2 cm size resembling a fibroma.

RESULTS

Histopathology, however, was not suggestive of a fibroma. On the contrary, it showed a well-circumscribed lesion in the deeper layers of the dermis with loose clusters of spindle and stellate cells, a lot of multi-nucleated giant cells in a hemorrhagic background along with a lot of haemosiderin laden macrophages, with vesicular nuclei. Metaplastic osseous tissue was seen to encase the tumor. A pathological diagnosis of Giant cell tumor of the Soft tissue (GCT-ST) was made. (Fig: 2)

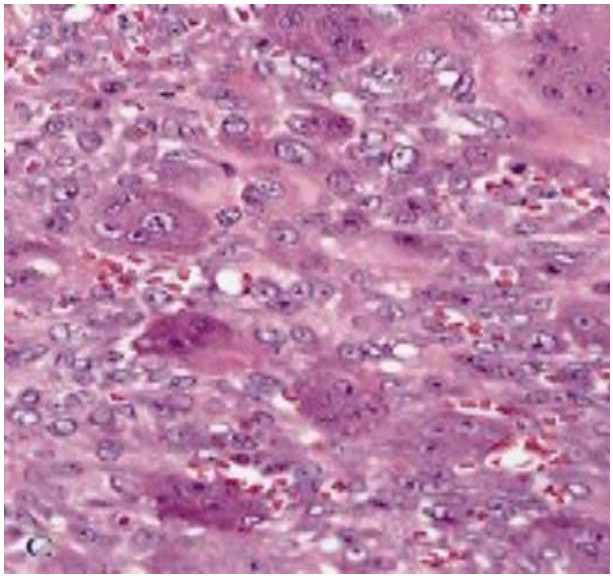

Fig: 2

Osteoclast-like metaplastic giant cells, haemosiderin-laden macrophages, and mild pleomorphism are seen. Cells with multiple nuclei are seen forming a syncytium.

DISCUSSION

Giant cell tumor of the soft tissue (GCT-ST) is such an unusual and rare tumor that the following are the only reported reports in the world. (arranged chronologically as per date of reporting):

1.A 74 year old male with a 20 cm mass in the right buttock[1]
2.A 58 year old lady with a 1.5 cm lesion over the left ankle[2]
3.A 73 year old man with metastatic tumor associated with post-transplant immunosuppression [3]
4.A 60 years old woman with a giant cell breast tumor and a fatal outcome [4]
5.A 54 year old man with GCT of finger [5]
6.A 19 year old lady with a 2.5 cm neck lesion[6]
7.A tumor of 46 years duration[7]
8.2 cases of GCT arising from post-operative surgical scars[8]
9.A 53 year old man with a GCT masquerading as mediastinal mass[9]
10. A 30 year old man with a 2.5 cm soft tissue mass on the thigh[10]
11. A **60-year-old** lady with a nodular thigh lesion. [11]
12. A 33-year-old female with a 3 cm tumor on the right thigh. [12]
13. A 30-year-old male with a nodule of unstated size over the right middle finger.[13]

Primary giant cell tumor of soft tissue (GCT-ST) is a rare soft tissue tumor with borderline malignant potential. It was first described in the same year, i.e. 1972, in two separate case series by Salm and Sissons (4 cases) [14] and Guccion and Enzinger [15], respectively (32 cases).

The lesion commonly affects patients above 40 years of age, is highly uncommon in children, and has no gender predilection [16]. It commonly involves the superficial soft tissues of the extremities and rarely the mediastinum, breast, trunk, thigh, and surgical scars.

The histologic features are similar to Giant cell tumor of bone, which is its bony counterpart so to say.

Histologically, it is usually a well-circumscribed, diffusely infiltrative multinodular, non-encapsulated lesion composed of round or spindle-shaped cells arranged in a storiform or fascicular pattern along with multiple scattered osteoclast-like giant cells. The giant cells often show multiple vesicular nuclei. [17]. Osteoid, chondroid, or mature metaplastic bone may also be seen, usually at the tumor periphery, sometimes extending to the center. Significant atypia excludes this diagnosis; however, frequent mitotic activity may be seen. Necrosis is rare. Hemorrhage is fairly common [18].

On immune-histochemical staining, these tissues are strongly positive for CD68, SMA, Vimentin, and p63 and negative for CD34, CD117, smooth muscle antibody, and Ki-67. [16]. Osteoclast-type giant cells stain positively for Tartrate Resistant Acid Phosphatase (TRAP), TRAIL, RANKL, and osteoprotegerin.

These tumors lack somatic mutations of the H3F3A gene, thereby differentiating GCT-ST from their bony counterpart.

Regarding imaging studies, CT Scans, and MR imaging usually reveal a solid, in-homogeneous hemorrhagic mass.

Differential diagnoses of GCT-ST include benign soft tissue tumors that are rich in giant cells, like pigmented villonodular synovitis, nodular tenosynovitis, tenosynovial giant cell tumor and malignant giant cell tumors of soft tissues like giant cell malignant fibrous histiocytoma, osteoclast-like giant cell-rich leiomyosarcoma, malignant melanoma with osteoclasts like giant cells and extra-skeletal osteosarcoma.

When compared to Giant cell tumor of bone. GCT-ST has a lower local recurrence but higher metastatic potential and death rate. The treatment and excision margins of GCT-ST are not clearly defined. It has been seen that iincomplete surgical excision is usually followed by aggressive local recurrence with poor overall prognosis.

Conclusion

Giant cell tumor of the soft tissue (GCT-ST) is a rare, unusual primary soft tissue tumor that might be confused with a variety of other soft tissue lesions without thorough workup and knowledge about its existence as one of the rare differentials.

Early reports reported a poor prognosis of GCT-ST, based on the histologic appearance, reports of local recurrence and metastasis, and aggressive clinical course [15]. However, recent papers have established that GCT-STs have a better clinical course than giant cell tumors of bone and other malignant differentials, and the depth of the primary lesion does not appear to be a significant prognostic factor [16, 19] in the eventual outcome.

However, the propensity for aggressive local recurrence, borderline malignant potential, and the chances of metastasis warrant a close clinical follow-up.

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Article information
Manuscript Submitted: 04-01-2024
Manuscript Revised: 19-01-2024
Manuscript Accepted: 26-01-2024
Manuscript published: 19-02-2024

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