Case Report

Giant cell tumour of soft tissue arising from intra muscular region of thigh - A rare case report

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Abstract

Giant cell tumour arising from soft tissue is a rare condition, which is of low malignant potential resembling giant cell tumour of bone histologically, Also known as osteoclastoma of soft tissue. We report a case of 29 year old male patient who has progressively increasing painfull swelling in his right thigh since 3 months, This patient has been referred to our center for an M.R.I scan of thigh.

Key Words: Giant Cell Tumor, Vastus Intermedius, Giant Cells, MRI Sequences, GCTST (Giant Cell Tumor of Soft Tissue).

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INTRODUCTION

Giant cell tumour of soft tissue which is histologically similar to giant cell tumor of bone (Ref 4) is an extremly rare disorder, First described by by Salm and Sissons, followed by Guccion and Enginger in early 1970's.It is described as a neoplasm with low malignant potential, low recurrence rate and very low metastasingcapacity. This neoplasm noted araising commenly in extremities, more predominantly in thigh and trunk followed by the upper limb and the head and neck region (Ref 1and2).The term malignant giant cell tumor has been reserved to only histological high grade lesions. There are only 200 reported cases noted of this rare condition in the literature(Ref 3). The main objective of reporting this case is to brief the clinical and more importantly radiological features of this rare condition and the histopathological correlation which is all ways an adjunct to radiological investigations to take the case to final conclusive diagnosis. The role of histopathology is very important as there were few close differential diagnosis offered to this particular case while studying M.R.I. So this case report helps radiologists to add Giant cell tumour as close differentials in soft tissue tumours as it is overlooked being very rare condition. A detailed clinical history with detailed radiological work up and histological correlation helps in diagnosing this rare condition which helps clinician for a precise treatment.

CASE REPORT

A 29 yr old male patient has been referred to our center for an M.R.I scan who came with chief complaints of pain and progressively increasing swelling in right thigh since 3 months. M.R.I of right thigh with hip joint has been performed on PHILIPS INGENIA 1.5 TESLA MRI machine with T1, T2, STIR sequences in axial, coronalandsaggital sections, DWI and ADC in axial sections.

M.R.I FINDINGS: MRI of right thigh with hip joint showing mixed signal intensity in right thigh involving vastus intermedius muscle which is predominantly hyperintense on T2 and STIR, predominantly hypointense on T1 with multiple fluid measuring 8.6cms in coronal 8.1 in saggitaland 4.5 cms in axial sections, fluid levels showing restriction on diffusion and reversal on ADC with a hyperintensity at proximal end of the right femur.

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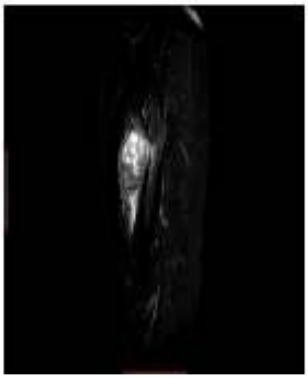
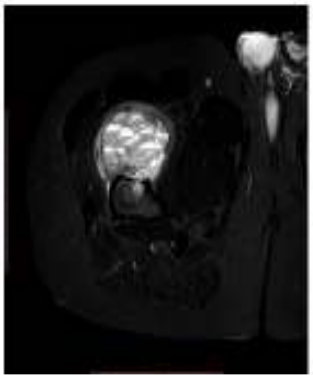


Figure 1



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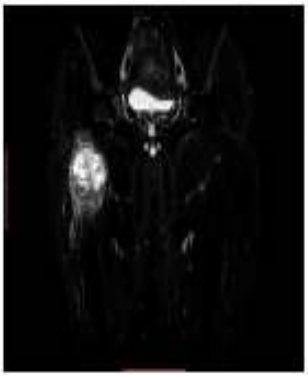

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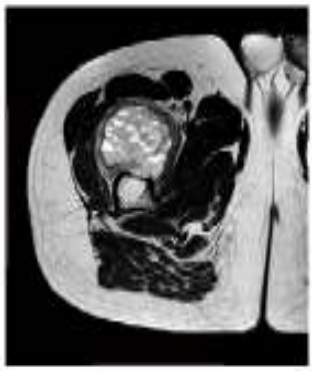

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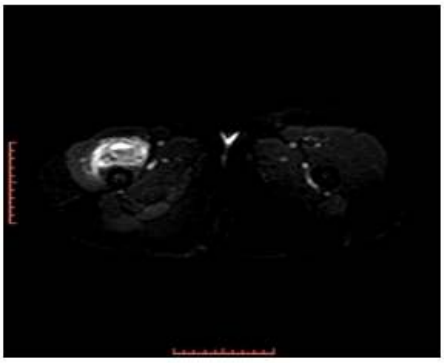

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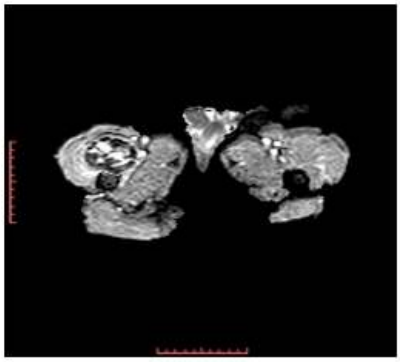

Figure 6

HISTOPATHOLOGY FINDINGS: After performing M.R.I to this patient gun biopsy has been done which showed large number of multi nucleated giant cells on back ground of fibrous tissue.

BIOPSY REPORT OF THE PATIENT:

BIOPSY

(Method: Leica Automated Tissue Processor and Stainer)

Biopsy No:

H17 - 5357.

Specimen :

Gun biopsy from right thigh for HPE.

Clinical Details :

(Method: Leica Automated Tissue Processor and Stainer)

Complaints of pain in right thigh since 3 months.
Complaints of swelling over right thigh since 1 month.
No history of fever / cough.
MRI - Suggestive of extra osseous ABC or soft tissue GCT.
? Soft tissue sarcoma right thigh.

Macroscopic Findings :

(Method: Leica Automated Tissue Processor and Stainer)

Type of specimen - Not mentioned : Received multiple grey white tissue bits altogether measuring 0.2 cms.
1 Cap - All embedded.

Microscopic Findings :

(Method: Leica Automated Tissue Processor and Stainer)

Sections studied show features of a giant cell tumor . Large number of evenly distributed multinucleated giant cells on a background of fibrous tissue are seen . Few foamy macrophages are noted . Focally, reactive/metaplastic bone formation is also noted. No evidence of sarcomatoid transformation / atypical mitosis . Negative for malignancy in the sections studied.

DISCUSSION

As stated earlier soft tissue giant cell tumour was described first in early 1970' s by Salm and Sissons who coined the term “Giant-cell tumors of soft tissues”. In the course, Guccion and Enzinger reported a more aggressive form of this same entity through the report of 32 cases considering that they are a related form of malignant fibrous histiocytoma (Ref 5and6).. In 1999, Folpe *et al*. proposed a histological sub-classification into two distinct forms according to nuclear atypia, pleomorphism and mitotic activity of the neoplastic component. The term

giant cell tumor of soft tissue of low malignant potential, has been suggested for low histological lesions, having low mitotic activity as well as little nuclear atypia.(Ref 7).These neoplasms araise from superficial regions of soft tissues predominantly from lower extremities and upper extremities according to the literature(Ref 6,10,11).Next comes pelvis,neckand trunk regions and very rarely breast, skinand mediastinum can also be involved(Ref 12,13and14). More commener age group being after 5th decade but here in our case report patient is an young adult of 29 yrs which is a rarity by itself.

Histological Features: This tumor is composed of mixture of round to oval cells that are mononuclear and osteoclast-like giant cells that are multinucleated, with both cell types immersed in a richly vascularisedstroma. Metaplastic bone formation is noted in most of the cases which is in the form of woven bone. This is due to TGF 1 and 2 secreted my tumor cells. Histiogenesis of GCTST is not clearly understood few studies suggest mesenchymal origin due to spontaneous origin of stromal cells due to alteration of blood flow due to surgery which cause fusion of monocytic giant cell (Ref 15).

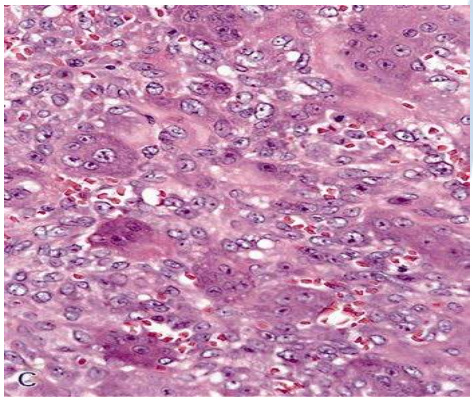

Figure 7: Microscopic appearance of soft tissue GCT

Differential Diagnosis: There are very close differential diagnosis includes conditions which shows predominant giant cell component like: giant cell tumor of tendon sheath, peripheral giant cell granuloma and giant cell malignant fibrous histiocytoma. Radiological similarities are seen in extra osseous aneurysmal bone cyst, paraostealosteo sarcoma of telengiectatic variant and soft tissue sarcoma. (Ref 8).

Immunohistochemical: Study can provide furthur information if required, shows positive strain for CD68, vimentin, tartrate resistant acid phosphatase (TRAP), cytokeratin and smooth muscle actin in both mononuclear cells and multinuclear giant cells (Ref 9).

Treatment: Surgical resection is the mainstay of treatment for this rare neoplasm Microscopically, the GCT includes mononuclear and reactive neoplastic stromal cells and giant cells of the multinucleated osteoclast type. Based on these data, several studies have been conducted to demonstrate the role of bisphosphonates as a potentially attractive therapy, to reduce osteoclast activity and to control tumor growth. In this context, denosumab is the first medical treatment that proved its profit on GCT. It's a fully human monoclonal antibody that specifically inhibits the RANK ligand, which enhance osteoclast's activity. In recent years, several studies on Denosumab in patients with GCTB has shown objective clinical and histological changes in the tumor's behavior. Zoledronic acid is effectively used for conditions with giant cell tumours with rich osteoclastic activity.

CONCUSION

GCTST is a rare slow growing neoplasm which usually araises from superficial soft tissue but here in our case it was araising from deep intra muscular region, This kind of presentation was not reported in the literautre earlier, Through this case report we brief the radiological features of this rare neoplasm which helps in adding GCTST as a differential diagnosis in soft tissetumours. One more rarity in our case report is the age of presentation as GCTST is seen in 5th decade where as the patients age is 29yrs here in our case. Histopathological correation with radiological features helps in diagnosis of this rare neoplasm which implies the role of Radiological imaging with histopathological correlation.

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