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Giant cell tumor of soft tissues: A case report of extra-articular diffuse-type giant cell tumor of the quadriceps



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ABSTRACT

INTRODUCTION: Giant cell tumors of soft tissue (GCTs) are a relatively rare entity. It is a distinct but uncommon group of neoplasms morphologically identical to osseous giant cell tumor. The diffuse type of extra-articular GCT arising within muscle is a rare benign soft tissue tumor with a wide spectrum of clinical presentation.

PRESENTATION OF CASE: This article reports a rare case of a 44-year-old woman with a mass arising from her right thigh. MRI showed only a few areas of low T2 signal in a mass that was hyper intense to muscle. Histopathology of this lesion located within the right quadriceps muscle revealed admixture of multinucleated giant cell with mononuclear cells. This patient was treated by surgical resection and followed up for recurrence.

DISCUSSION: Diffuse-type GCTs are commonly located in the periarticular soft tissues, but on rare occasions these lesions can be purely intramuscular or subcutaneous and can be challenging to diagnose. Characteristic findings include gradient echo secondary to hemosiderin deposition, and the low signal on T2.

CONCLUSION: Because extra-articular diffuse-type GCTs are rare, the differential diagnosis is challenging. The clinical outcomes of diffuse-type GCTs are unclear because of their rarity. Benign clinical course is expected if the lesion is excised adequately.

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1. Introduction

Tenosynovial giant cell tumors (GCTs), also known as pigmented villonodular synovitis, are benign soft tissue tumors that arise from the synovium of the joints, bursae and tendon sheath [1,2]. These tumors are classified into two forms: localized and diffuse (Fig. 1). Diffuse-type giant cell tumor is an extra-articular form of pigmented villonodular synovitis [3].

Localize-type tenosynovial GCTs are frequent, indolent, and arise from the synovium of the joint, bursa, or tendon sheath. In contrast, the less frequent diffuse-type tenosynovial GCTs are locally aggressive and commonly affect peri-articular soft tissue, but on rare occasion these lesions can be intramuscular [2,4]. The yearly incidence of diffuse –type tenosynovial GCTs is two per 1,000,000 cases. Extra-articular diffuse-type GCTs represent only 5–15% of all diffuse-type tenosynovial GCTs. Despite the rarity of intramuscular

diffuse GCTs, it is important to know that they infiltrate surrounding the soft tissue and have a high recurrence rate [2]. Localized versus diffuse forms of tenosynovial GCTs may cause different clinical symptoms.

Here we describe the case of 44-year-old woman who presented with a thigh pain and a mass in the right quadriceps. The final diagnosis was an extra-articular diffuse type intramuscular giant cell tumor treated by a complete surgical resection. It is a rare benign tumor but locally aggressive which must be known as a differential diagnosis of sarcoma.

This case report is in line with the SCARE criteria [5].

2. Case report

The patient was 44-year-old, a sedentary housewife, with three-year history of vague pain in the right thigh without any antecedent of trauma or infection. She visited our institution, on June 2015, with a right mass above the knee first noted 10 months prior to presentation. There was no restriction of movement of the right knee, and the patellar compression test was negative.

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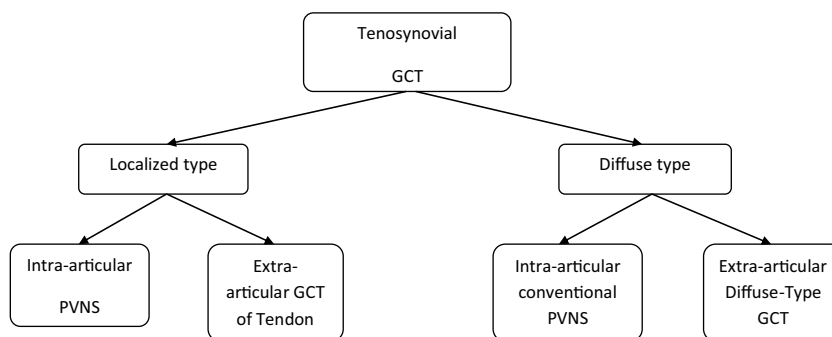


Fig. 1. Classification of tenosynovial GCTs. GCT: giant cell tumor; PVNS: pigmented villonodular synovitis.



Fig. 2. An initial anteroposterior radiograph of the right thigh shows a soft tissue mass.

On examination, it appeared to be a non-tender soft tissue mass measuring about 7.0*4.0cm in size. The mass was fixed to the muscle.

The radiographs of the knee and the femur demonstrated a lobulated soft tissue mass at the anterior and lateral aspect of the right lower thigh. There were no calcification related to the lesion (Fig. 2).

On sonographic examination, the mass was heterogenous echoic mass with a hypoechoic central portion (Fig. 3).

A magnetic resonance imaging (MRI) study was performed which showed a mass confined to the Rectus Femoris muscle. The size of the mass was 7.2*5.3*2.5 cm. The signal intensity was less than of subcutaneous fat on T1-weighted sequences. On T2-weighted images, the tumor was heterogenous, it was hyper intense with focal areas of low signal intensity within the mass. The

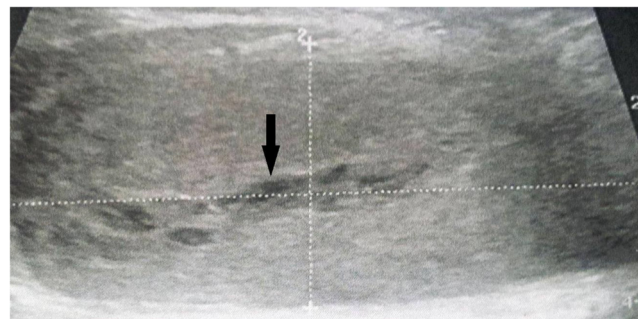


Fig. 3. Sonographic findings of the intramuscular diffuse-type giant cell tumor. Sonography revealed a heterogeneous echoic mass with a hyperechoic central portion.

major portion of the mass showed enhancement after gadolinium enhancement (Fig. 4).

We provided a differential diagnosis of rhabdomyosarcoma, synovial sarcoma and desmoid tumor based on these MRI features.

A biopsy of the tumor was performed. In histological findings, The tumor was densely cellular and was composed of sheets of mononuclear cells admixed with multinucleated giant cells, the mononuclear cells are round or polygonal «histiocytic» and exhibit little pleomorphism nuclei. Some of these cells contain intracytoplasmic hemosiderin. The cells dysplasia are occasional mitotic figures. Multinucleated giant cells are intermingled scattered throughout the lesion and have a variable number of nuclei, rangin from 3 to 20. Inflammatory cells, particularly lymphocytes, siderophages and xanthoma cells are scattered randomly throughout the tumors but very greatly in number. Pseudogranular spaces are focally seen. The tumor infiltrates the tendon sheath. The stroma was collagenous and inconspicuous. The pronounced cellularity coupled with the polymorphic population of cells is likely to lead to a diagnosis of malignancy.

Immunohistochemical study was done. The tumor cells were positive for cluster for differentiation (CD) 68 and CD163, and were negative for CD34, cytokeratin (AE1,AE3), S100 protein, HMB45, melan A, desmin, myogenin and MDM2. The cells don't express the colony stimulating factor CSF-1.

The histopathological diagnosis was a diffuse-type giant cell tumor (Fig. 5).

After preoperative evaluation and preparation, a carefully complete surgical resection of the tumor was done. Infection prevention, venous thrombosis prevention, analgesia and other conventional medical treatments were given according to standard clinical pathway in perioperative period of soft tissue tumors.

Abundant hemosiderin pigment was seen in macroscopic examination. The diagnosis of diffuse-type GCT was confirmed by the histological findings of the final piece of tumor.

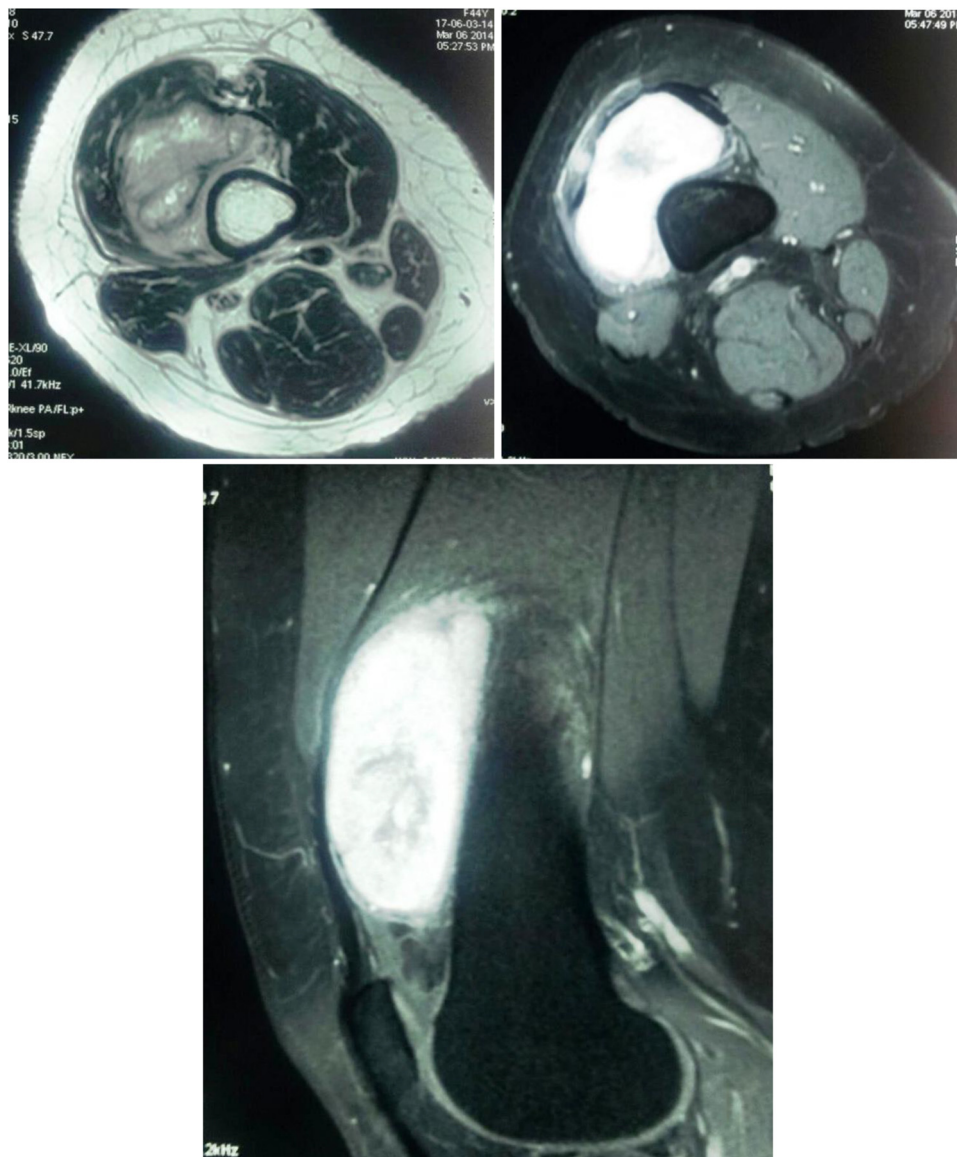


Fig. 4. T1-weighted axial (a), enhanced T1-weighted fat saturated axial (b) and T2-weighted sagittal (c) MRI of the intramuscular diffuse-type giant cell tumor. T1-weighted images of the thigh shows a soft-tissue mass confined to the Rectus Femoris muscle (a). The signal intensity was less than of subcutaneous fat. The mass contains regions of low signal intensity. The major portion of the mass showed enhancement after gadolinium enhancement (b). On T2-weighted images (c), the tumor was heterogenous, it was hyper intense with focal areas of low signal intensity within the mass.

The patient remained symptom-free with no local recurrence 1 year after surgery; MRI should have been made, but the patient was lost in follow up.

3. Discussion

The extra-articular form of diffuse-type tenosynovial GCT was first described by Jaffe et al. [1]. Diffuse-type GCTs are now classified as «fibrohistiocytic tumors» in the World Health Organization system of classification of bone and soft-tissue tumors [7]. Unlike localized types, these tumors are aggressive and recur in 33–50% of the cases, often with multiple recurrences. A long-term following up is needed in our case.

Diffuse-type GCTs have a slight female preponderance [8]. These lesions are seen usually under the age of 40 [7].

Diffuse-type tumors are commonly located in the periarticular soft tissues, but on rare occasions these lesions can be purely

intramuscular or subcutaneous and can be challenging to diagnose [6].

Most intramuscular diffuse-type GCTs are located in the lower extremities such as the thigh, buttock, and lower leg, but all the muscles can be affected [6].

In a review of 50 cases of diffuse-type giant cell tumors by Somerhausen and Fletcher [6] there were only three cases (6%) with involvement of the subcutaneous tissues, of which two were in the knee region.

MRI is highly sensitive and specific for the diagnosis of intra-articular PVNS and its extra-articular localized and diffuse forms. Characteristic findings include gradient echo secondary to hemosiderin deposition, and the low signal on T2 [8]. In comparison, our case showed only a few areas of low T2 signal in a mass that was hyperintense to muscle.

Because extra-articular diffuse-type GCTs are rare, the differential diagnosis is challenging. Our pre-operative differential diagnosis included soft-tissue sarcoma and desmoid tumor [9].

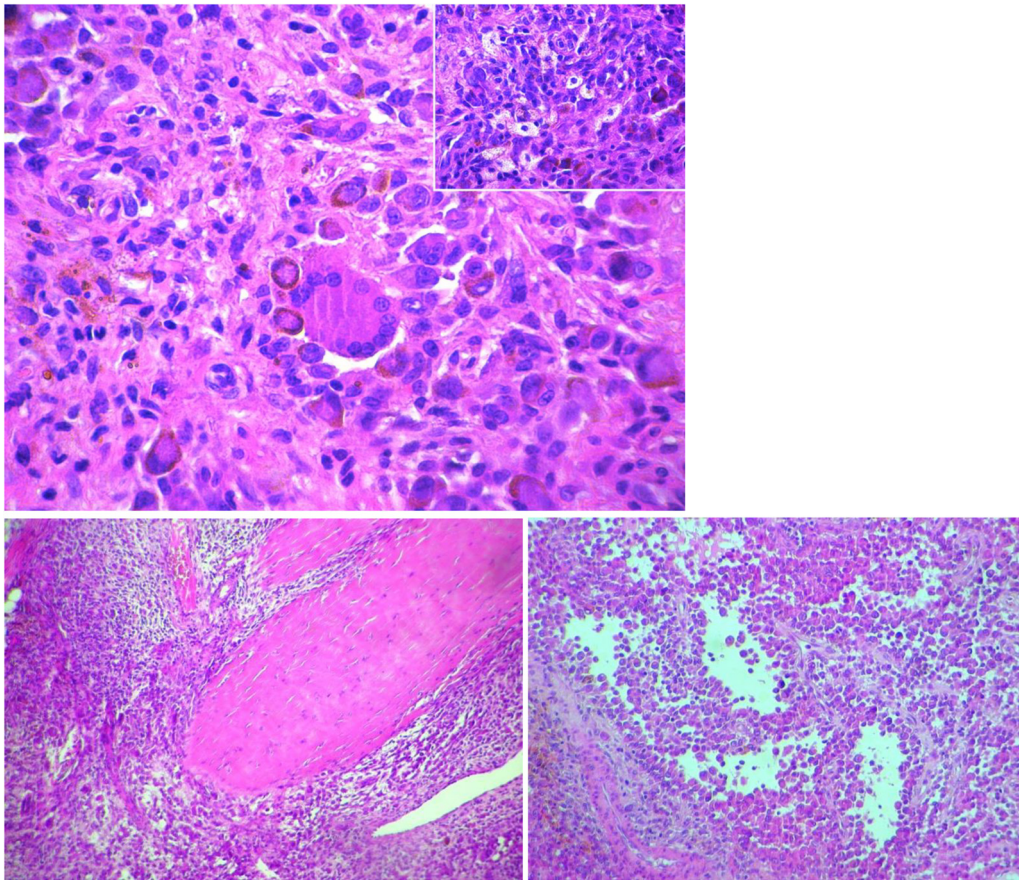


Fig. 5. Histologic findings of the intramuscular diffuse-type giant cell tumor.

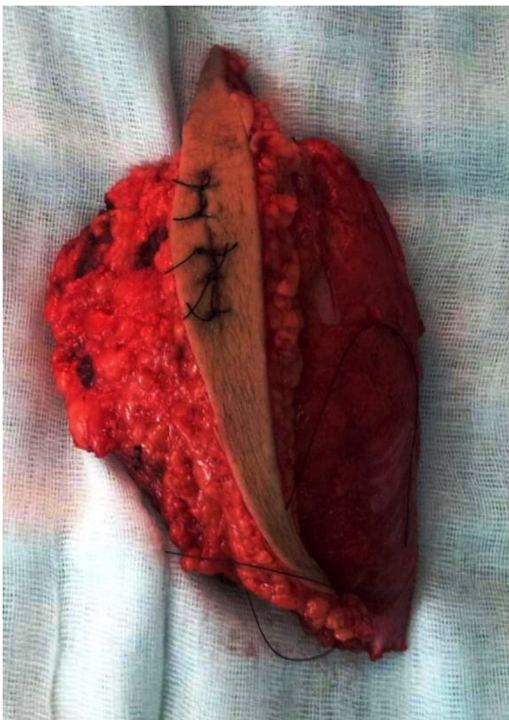


Fig. 6. Gross specimen of the resected tumor.

Giant cell tumors of soft tissue are slowly grown, painless lesions, and are treated with surgical excision. Although benign,

these tumors have a high recurrence rate ranging from 9% to 44%. There are a few case reports of malignant PVNS [10,11]. The clinical outcomes of diffuse-type GCTs are unclear because of their rarity.

In our case, the tumor may have possibly arisen from the tendon or bursal surface (Fig. 6).

4. Conclusion

In summary, we reported a case of intramuscular diffuse-type Giant Cell Tumor in a female of 44-year-old. It is a benign but locally aggressive tumor; its local recurrence cannot be predicted. Proper surgical excision and a long period of follow up are essential in these cases. Because of the unusual type and location and the lack of typical T2 shortening seen in extra-articular GCTs, the present case may be instructive for differential diagnoses of primary intramuscular tumors.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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Ethical approval

Approval has been given by the patient.

Consent

The patient's consent has been obtained.

Author contributions

Kochbati Rateb and Abid Leila contributed in writing the paper. Ben Ghozlen Hassen and Farah Faten contributed in the data collection. Daghfous Med Samir contributed in study concept.

Registration of Research Studies

None.

Guarantor

Kochbati Rateb.

Disclosure

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