Learning Point of the Article:

Giant cell tumors of soft tissues should be diagnosed and treated appropriately to avoid reoccurrence.

Abstract

Introduction: Giant cell tumor of soft tissue (GCT-ST) is a low-grade ST tumor that occurs most frequently below the skin and sometimes, extending deeper into the tissues. The tumor is usually hard, painless and considered rare in ST.

Case Report: We report a rare case of a middle-aged woman that presented with a tumor arising from the right tensor fasciae latae muscle. The patient initially opted for traditional Chinese herbal treatment, which we believe, aggravated the growth of the tumor. Radiographic imaging showed clear tumor margins. The histopathological biopsy of the lesion located in the tensor fasciae latae muscle showed a mixture of mononucleated and multinuclear giant cells composed of mesenchyme spindle-shaped cells positive for CD34 and CD88. The patient was treated through surgical resection of the tumor and scheduled for close follow-up.

Conclusion: Since GCT-ST is rare, they are often initially misdiagnosed. These tumors have the tendency to increase in size exponentially within a short period of time, which is why clinical presentation and imaging are necessary for a better pre-operative diagnosis. Radical surgical excision or radiotherapy with close follow-up is recommended for all cases due to the high rate of reoccurrence.

Keywords: Giant cell tumor, tensor fasciae latae, intramuscular tumor, case report, middle-aged female.

Introduction

In surgical practice, soft tissue (ST) tumors are known to be extremely rare with unpredictable behavior. Salm and Sissons first described giant cell tumors-STS(GCT-STSs) in 1972. Such tumors present in different shapes and sizes and can be found in any part of the body with the majority of reported cases appearing in the extremities, with the thigh being the most commonly affected site [1, 9]. Clinically, GCT-STSs manifest as an asymptomatic mass, but some of these tumors are very aggressive and have the potential to increase in size within a short period of time [2]. As a result of this aggressive behavior, they often metastasize to surrounding connective and adipose tissues, skeletal muscles, and blood and lymph vessels [3]. The vast majority of these tumors are successfully treated with clean surgical excision, while others may require additional treatment regimens. Lipoma, hemangioma, and giant cell carcinomas are among the most commonly seen ST tumors, radiographic imaging and tissue biopsies are the best diagnostic techniques in determining the marginal location and the type of tumor, respectively. In this paper, we present the clinical, histologic and radiologic features of a case of a malignant GCT-ST in a 44-year-old female who presented with an extremely painful right hip mass that aggressively progressed over 6 months. The tumor has been completely excised with surgery and follow-up has been advised to monitor reoccurrence.
A 44-year-old Asian female presented with an extremely aggressive intramuscular, tender upper right thigh mass of 350mm × 250mm which had progressed over a 6 month duration without any antecedent of infection or trauma. The patient mentioned the size of the tumor was non-remarkable in the first 2 months when she first discovered it in October 2017 but progressed rapidly despite using traditional Chinese medicine and moxibustion. The lesion at the time it was presented to our department on March 2018 had a fungating skin appearance and ST involvement (Fig. 1 and Fig. 2) with a foul-smelling odor. The patient before presentation had used traditional Chinese herbal medicine and moxibustion to treat the tumor, but there was no remarkable improvement after 4 months of treatment. Apart from a previously treated mild thalassemia, there was no other significant preexisting medical history or any known allergies. On examination, the tumor appeared to be tender and soft to touch. Computed tomography and positron emission tomography scans showed a well-demarcated tumor margin with no bone involvement confined to the tensor fasciae latae muscle (Fig. 3 and Fig. 4). After all standard pre-operative evaluation and preparation, the tumor was successfully resected (Fig. 5 and Fig. 6), followed by additional post-operative medical treatments of pain management, infection prevention, diet modifications, and other conventional rehabilitation procedures.

**Case Report**

Microscopic study of the tissue sample revealed that the tumor was located in the intramuscularly with a relatively clear local boundaries, the tumor cells were diffuse and rich in cytoplasm. The nucleus fat showed spindle, oval, round, and polygonal shapes. There was pleomorphic and heteromorphosis mixture of mononucleated and multinuclear giant cells. There was an obvious hemorrhagic necrosis due to the blockage of venous drainage. However, there was the presence of rich small interstitial blood vessels, part of the region seems to have tumorous osteogenesis. In accordance with clinical history, the tumor conforms to giant cell carcinoma of ST. (Fig. 7a and Fig. 7b) Ck(−), Vimt (+), S100(−), smooth muscle actin (SMA) (+), desmin (−), CD34 (blood vessel+), CD88(increase+), ki-67(about 50%+).

**Immunohistochemical study**

The GCT-ST, which is an extremely rare tumor, resembles its osseous variants both histologically and clinically [4,5]. It was first reported as early as 1972 in a case series of 10 benign primary ST tumors [6]. These tumors can exhibit aggressive malignant transformation, abundant mitotic activity, and pleomorphism [6]. However, GCT-ST is predominantly benign in nature, with a high possibility recurrence possibility if inadequately excised. The majority of the cases of GCT-ST occur in the extremities. However, cases occurring in rare locations such as the head, neck, and ears have been reported [7, 7].

**Discussion**

The GCT-ST, which is an extremely rare tumor, resembles its osseous variants both histologically and clinically [4,5]. It was first reported as early as 1972 in a case series of 10 benign primary ST tumors [6]. These tumors can exhibit aggressive malignant transformation, abundant mitotic activity, and pleomorphism [6]. However, GCT-ST is predominantly benign in nature, with a high possibility recurrence possibility if inadequately excised. The majority of the cases of GCT-ST occur in the extremities. However, cases occurring in rare locations such as the head, neck, and ears have been reported [7,
Until now, etiological factors for GCT-ST have not been identified, and some reports suggest that chronic inflammation may have a role in its pathogenesis, which remains unproven to date [7]. GCT-ST mostly presents as a painless mass with an average symptom duration of 6 months–1 year [7,12]. The mass, in our case, was tender, huge, and with loss of the skin. In our case, microscopic examination showed diffuse tumor cells arranged in a crisscross pattern. Furthermore, there were unclear cell boundaries and multinucleated giant cells. Atypia was obvious. Our immunohistochemistry results indicated vimentin(+), CD68 (high), and SMA negative [8], CD34 (positive for blood vessels), CK(−), S-100(−), desmin (−), and Ki-67. The Ki-67 labeling index was approximately (M ref). The osteoclast-like giant cells usually exhibit positive CD68 and negative CD34 immunostaining [9,10]. GCT-ST is often initially misdiagnosed [4, 11], which was the case of our patient. It was initially diagnosed as a lipoma. These tumors can grow exponentially in a short period of time. Radical surgical excision or radiotherapy with close follow-up is recommended for all cases due to the high rate of reoccurrence. In this case, we opted for radical excision because the tumor was extremely large and adversely affected her daily life activities. Second, due to its size, it had invaded the muscular layer. Conservative surgical resection with tumor-free margin is the typical treatment for GCT-ST that assures a good prognosis with very low recurrence rate[11]. Magnetic resonance imaging (MRI) scans are the most reliable radiographic diagnostic choice for identifying the margins and invaded ST because they are highly sensitive.

**Conclusion**

We reported a case of intramuscular diffuse-type GCT-ST in 44 years old. The tumor was exceptionally aggressive and local reoccurrence after surgery cannot be predicted. GCT-STs are clinically and morphologically identical to GCT of the bone, which is why they can progress rapidly and aggressively if not properly and promptly treated. Radical and complete surgical excision is the most reliable treatment approach. Reoccurrence is very common; therefore, close monitoring during follow-up is recommended. In the case of this patient, the growth of this tumor was likely due to the delay in surgical treatment in addition to the use of herbal moxibustion, which likely escalated the growth. Due to how rare GCT-STs are, we hope this case serves as an instructive for prompt diagnosis and treatment of any primary intramuscular tumors.

**Consent**

The patient was asked if the data concerning the case can be used for publication, and they consented.

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**Clinical Message**

Diagnosing GCT-ST can be difficult both radiographically and clinically because they are extremely rare with unpredictable behavior. However, MRI is the most reliable radiographic diagnostic choice for identifying the tumor margins and invaded ST. Radical surgical resection of the tumor is the treatment for GCT-ST that assures a good prognosis. Reoccurrence is very common; therefore, close monitoring of the patient post-operative is recommended.

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Conflict of Interest: Nil

Source of Support: Nil

Consent: The authors confirm that Informed consent of the patient is taken for publication of this case report

How to Cite this Article

Mensah D, Chhantyal K, Lu Y X, Li Z Y. Malignant Giant Cell Tumor of Soft Tissue: A Case Report of Intramuscular Malignant Giant Cell Carcinoma of the Tensor Fasciae Latae. Journal of Orthopaedic Case Reports 2019 Jan-Feb; 9(1): 70-73.