Abstract

Tenosynovial giant cell tumors (TGCT) are a group of generally benign intra-articular and soft tissue tumors with common histological features. TGCT is also known as pigmented villonodular synovitis. There are localized and diffuse forms. Localized types include giant cell tumors of tendon sheath and localized pigmented villonodular synovitis, whereas diffuse types encompass conventional pigmented villonodular synovitis and diffuse type giant cell tumor. Localized tumors are generally indolent, whereas diffuse tumors are locally aggressive.

In this article, we report the case of a diffuse-type extra-articular TGCT found in the left thigh of a 73-year-old woman who presented with a painless but gradually progressive swelling in the left thigh since eighteen months. On examination, there was a soft cystic swelling measuring 22 cm. The swelling was fixed to the underlying soft tissues. She had a Computed Tomography scan of the left thigh showing a mass of fluid density, well encapsulated between the muscles of the thigh, measuring 20x10 cm of major axes, compressing the femoral vessels without invading them. The patient had a complete marginal resection of the tumor. The immune-histopathological findings were consistent with those of a diffuse type of Giant Cell Tenosynovial Tumor. At 18 months follow-up, the patient is asymptomatic with no evidence of disease recurrence.

The extra-articular diffuse type TGCT is more aggressive than the localized type. Although these tumors are benign in the majority of cases, malignant transformation has been reported. Therefore, close follow-up is recommended after tumor excision.

Keywords
Giant Cell Tumor; Giant Cell Tumor of Soft Tissue; Thigh; Diffuse Variant.
Introduction
Giant cell tumors are classified according to their site of origin into bone, soft tissue and tenosynovium. They can be divided into a localized or nodular and a diffuse type or pigmented villonodular synovitis. The nodular type is confined to a distinct area of synovium, it affects the smaller joints of the fingers, and it is generally extra-articular. In contrast, the diffuse type shows extensive involvement of the synovial membrane and capsule, it usually affects larger joints and it is often intra-articular and infiltrative. It is generally arising from the synovial membrane that composes the lining of joints, tendons and bursa. Rarely, the diffuse type can be extra-articular. Herein, we present the case of a diffuse-type extra-articular tenosynovial giant cell tumor found in the left thigh of a 73-year-old woman diagnosed and treated in January 2015 in the department of oncologic surgery in Salah Azaiez Institute. The case is presented for its extreme rarity and the difficulty of differential diagnosis.

Case report
A seventy three year old woman presented with a painless but gradually progressive swelling in the medial aspect of the left thigh since eighteen months. There was no history of previous trauma in that region. On examination, there was a soft cystic swelling measuring 22 cm. The swelling was fixed to the underlying soft tissues. She had a Computed Tomography scan of the left thigh showing a mass of fluid density, well encapsulated between the muscles of the thigh, measuring 20x10 cm of major axes, compressing the femoral vessels without invading them (Figure 1). The magnetic resonance image was not done because it was not available in our institution, and the patient could not afford this examination in a private centre. A preoperative biopsy was performed but it was not decisive. The patient underwent a surgery consisting of a complete marginal resection of the tumor (Figure 2). The intraoperative findings showed that the tumor was adhesive to the adjacent muscles of the anterior and the medial compartment of the thigh, without invading them. The specimen was sent for histopathological examination. Macroscopically, it was a well circumscribed mass measuring 22x12cm with focal

Figure 1: CT Scan of the left thigh showing a mass of fluid density (A+B).
cystic change. Microscopic examination showed a tumor composed of a mixture of mononuclear round to oval cells and osteoclast-like giant cells that were multinucleated. The immunohistochemical results revealed that CD68 immunoreactivity was diffuse in multinucleated giant cells, whereas it was focal in mononucleated cells. These immuno-histopathological findings were consistent with those of a diffuse type of Tenosynovial Giant Cell Tumor. At 18 months follow-up, the patient is asymptomatic with no evidence of disease recurrence.

Discussion

Tenosynovial giant cell tumors is a group of tumors that originate in tendon sheaths, joints, bursa, or adjacent soft tissue. TGCT was first described in 1852 by Chassaignac as synovial membrane proliferation involving the flexor tendons of the fingers and was later redefined by Jaffe et al. in 1941 [1]. TGCT can be subtyped into diffuse and localized types, and intra-articular and extra-articular types, according to the growth pattern and location [2]. Giant Cell Tumor of Soft Tissue was first described in 1972 in two simultaneous publications, one series reporting benign cases and another series with tumors showing aggressive behavior [3, 4]. This entity was originally called “malignant fibrous histiocytoma, giant cell type” based on the fact that these tumors were linked histogenetically to malignant fibrous histiocytoma [3]. Giant cell tumors of soft parts have now been reclassified as Giant Cell Tumor of Soft Tissue (GCT-ST) and undifferentiated pleomorphic sarcoma with giant cells (giant cell malignant fibrous histiocytoma or malignant giant cell tumor of soft parts in the current WHO Classification of Tumors of Soft Tissue and Bone) [5]. Diffuse-type TGCT is extra-articular type in about 5% to 15% of cases [6]. The tumor can be locally aggressive, even if benign in nature, and 33% to 50% can recur. Metastasis to other organs is rare after multiple recurrences [6-7]. These tumors
present as painless growing masses [8, 9] with an average duration of symptoms of 6 months [8, 9]. No etiological factors have been identified [5]. The treatment remains controversial because of the small number of reported cases. Current treatment of choice for extra-articular diffuse type TGCT is surgical excision [10, 11]. Complete removal of the tumor with negative surgical margins assures no recurrence. But there is a significant risk of multiple recurrences with aggressive diffuse disease. A local adjuvant treatment is usually not necessary [10]. However, radiation therapy has been reported as primary treatment for unresectable disease or as local adjuvant treatment after incomplete excision or locally recurrent tumors [10].

Conclusion
The extra-articular diffuse type TGCT is more aggressive than the localized type.

It presents a diagnostic challenge. These tumors are slow growing with atypical presentation, difficult differential diagnosis, and higher local recurrence rate.

Although these tumors are benign in the vast majority of patients, malignant transformation has been reported [6]. Therefore, close follow-up is recommended after tumor excision.

References
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