

Primary Soft Tissue Giant Cell Tumor: A Rare Case Exhibiting Features of an Aneurysmal Bone Cyst

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Özet

Yumuşak dokunun düşük malign potansiyelli dev hücreli tümörü (YD-DHT), nadir bir tümördür. Morfolojik olarak kemiğin dev hücreli tümörüne eşdeğerdir. Tümör yüzeysel ve derin yumuşak dokuyu tutar, genç ve orta yaşlarda daha sıktır ve her iki cinsten eşit olarak karşımıza çıkar. Geniş lokal eksizyondan sonra nadiren nüksler görülebilir. Genellikle benign davranış gösterir. Metastaz nadiren görülür. Biz bu yazıda anevrizmal kemik kisti özellikleri de içeren bir yumuşak doku dev hücreli tümörünü sunmak ve histolojik ayırıcı tanısını tartışmak istedik.

Anahtar kelimeler: Dev hücreli tümör, Yumuşak doku

Abstract

Soft tissue giant cell tumor (ST-GCT) is an uncommon neoplasm with low malignant potential that is morphologically identical to osseous giant cell tumor. This tumor involves both superficial and deep soft tissues and presents more frequently during early adulthood as well as in middle age. It equally affects both sexes. Recurrences occur rarely after wide local excision, and metastases are not frequent. In this manuscript, we present a soft tissue giant cell tumor exhibiting features of aneurysmal bone cyst and discuss the histological differential diagnosis.

Keywords: Giant cell tumor, Soft tissue

Introduction

Soft tissue giant cell tumor (ST-GCT) with low malignant potential is a rare neoplasm. In 1972, Salm and Sisson first termed 10 cases identical to osseous giant cell tumor as primary ST-GCT and noted recurrence in 2 cases¹. At the same time, Guccion and Enzinger reported that these tumors demonstrate a more aggressive clinical course and resemble malignant fibrous histiocytomas². In 1999, Folpe et al. classified these tumors as the soft tissue equivalent of giant cell tumors with low malignant potential due to their variable biological behavior³. In 2002, the World Health Organization accepted these tumors as intermediate malignancies of fibrohistiocytic tumors⁴.

ST-GCT involves both, superficial and deep soft tissues; it presents more frequently during early adulthood as well as middle age; and equally affects both sexes¹⁻¹⁰. It is most frequently noted in the lower limb and trunk followed by the upper limb and the head and neck region. Clinically, it manifests as an asymptomatic, multinodular mass with benign behavior.

The other osseous and soft tissue giant cell tumors and aneurysmal bone cysts should be considered in the differential diagnosis. Recurrence occurs rarely after wide local excision. ST-GCT is usually benign with rare instances of metastases⁶. Histologically, it is similar in appearance to its osseous counterpart. It consists of mononuclear cells and spindle-like cells as well as osteoclast-type giant cells, which are equally scattered between these cells in the stroma. There is limited mitosis and atypia, and necrosis is usually absent. It has been reported that aneurysmal bone cyst-like changes are noted in 30% of the cases⁵⁻⁷.

We report this rare disease presentation because it is the first case in our 35-year database, with areas resembling aneurysmal bone cysts.

Case Report

A 54-year-old male presented to the Orthopedic Department of Erciyes University Medical School with left shoulder pain of 6 month duration. On physical examination, a nodular lesion was detected posterolateral to the left shoulder, with tenderness to palpation. Although the skin tissue overlying the lesion appeared reddish-brown, there was no history of trauma or previous intervention at that site.

On MR imaging, a nodular soft tissue mass (3.5x5.5 cm in size) causing edema in the adjacent muscle group was observed within the deltoid muscle posterolateral to the left shoulder, with cystic necrotic areas within a hypointense capsule on T1- and T2-weighted images, as well as contrast enhancement in the solid component (Figure 1). On the screening CT scan, no abnormal findings were noted in the lungs, lymph nodes, thyroid gland and bones.

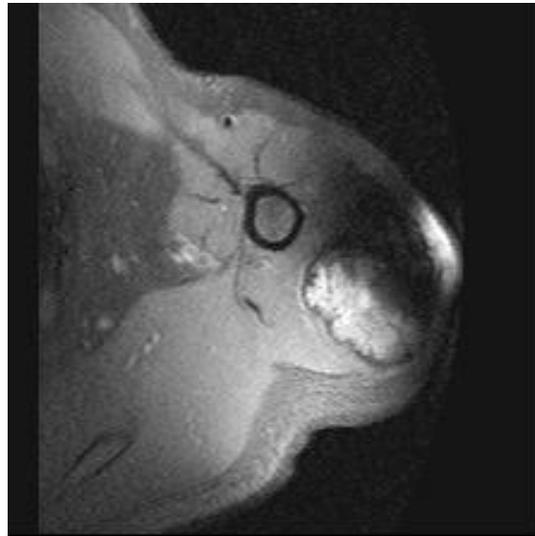


Figure 1

Preoperative MR imaging demonstrating a soft tissue mass (3.5x5.5 cm in size) with a hypointense capsule and cystic and necrotic areas.

An incisional biopsy was performed. Pathology was reported as “A mesenchymal tumor; morphological appearance primarily supports soft tissue giant cell tumor”. A wide excision was performed by the orthopedic department based on the pathology report. Postoperatively, the patient was discharged without complications, and no recurrence has been observed over a follow-up period of 8 months.

The specimen was 9x8x7 cm in size with proximal, distal and posterolateral surgical margins. Sectioning along the long axis revealed a reddish, well-defined nodular lesion (4.5x5.5 cm in size) with a thin capsule, as well as acystic, hemorrhagic appearance at its center within the muscle tissue (Figure 2).

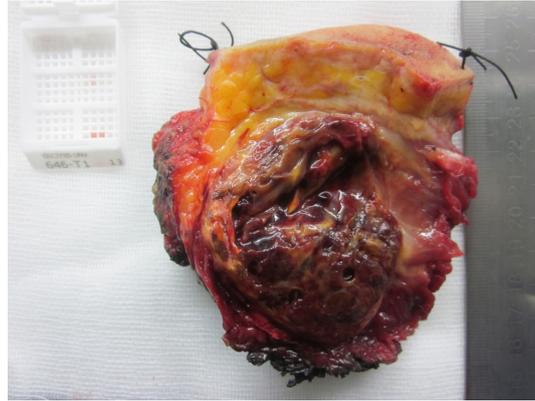


Figure 2

Gross evaluation shows a a reddish-brown, encapsulated nodular lesion within muscle tissue with well-defined margins and central cystic, hemorrhagic areas.

On microscopic evaluation, there were mononuclear cells with round and spindle-like appearance within the tumor tissue with occasional demarcation from the surrounding muscle tissue on H&E-stained sectioning. These cells had round, oval and vesicular nuclei. There was a striking presence of abundant multi-nuclear giant cells, which were uniformly scattered between mononuclear cells (Figure 3).

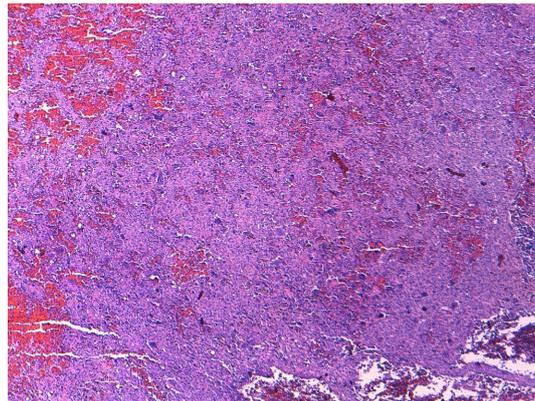


Figure 3

Round-oval to spindle-like stromal cells with abundant multi-nuclear giant cells uniformly scattered over stroma.

The nuclei of giant cells had similar characteristics to those of stromal cells. In addition, blood-filled spaces with cystic dilatation and areas of proliferation involving spindle-like cells forming short fasciculi surrounding these areas of cystic dilatation were noted (Figure 4).

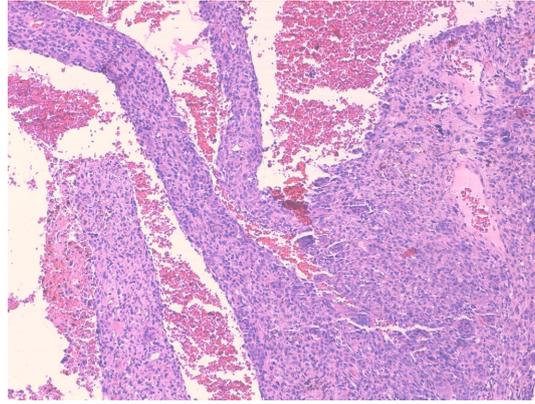


Figure 4

Hemorrhagic, dilated spaces with surrounding spindle-like and giant cells (Hex200)

There were areas with fibrohistiocytic changes. Although 3 mitotic figures were observed within fasciculi on 10x magnification, no atypical mitotic figures, necrosis and marked pleomorphism were noted (Figure 5). In some fields, extravasation of erythrocytes and fibrosis were observed. No osteoblastic activity or bone formation was observed within the tumor or peripherally.

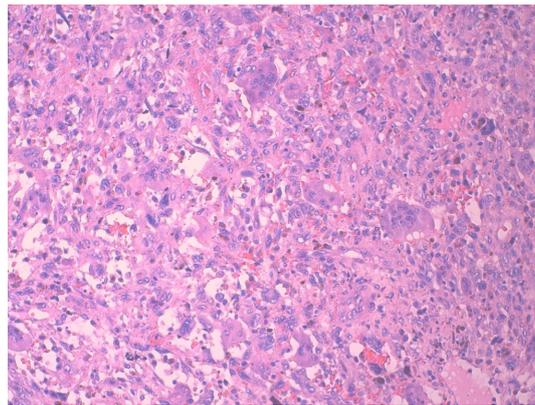


Figure 5

Stromal and giant cells on greater magnification. No marked atypia and mitosis (Hex400)

On immunohistochemical staining, the following results were found: positive staining with CD68 and TRAP in giant cells; diffuse staining with Vimentin and negative staining with Actin, Desmin, S-100, CD117 and CD34 at the spindle-like proliferation areas. The Ki-67 proliferation index was estimated as 15%.

Based on these findings, the lesion was characterized as a “soft tissue giant cell tumor with low malignant potential”

Discussion

Primary soft tissue giant cell tumor is a rare entity which was first defined by Salm and Sisson in 1972¹. It shows similar gross features, microscopic characteristics and clinical behavior to its' osseous counterpart. It may involve the superficial and deep soft tissues. It is usually located in the upper and lower limbs. The trunk and head and neck

regions are less frequently involved. In this case, tumor was located at the shoulder. It is usually a brisk, painless and mobile mass with well-defined borders which does not involve the tendon, muscle or bony structures. It has no particular pattern in terms of age and sex. Similar to its' osseous counterpart, ST-GCT shows positivity of vimentin, CD68 and tartrate-resistant acid phosphatase (TRAP) in both mononuclear stromal cells and multinuclear giant cells on immunohistochemical staining^{3,6,7}. Our case also showed positive staining with the above-mentioned immunohistochemical stains.

In some series, the incidence was found to be lower than local recurrent osseous giant cell tumor (37.5% vs. 6.2%); however, metastasis and mortality rates were found to be higher (0.4-3%vs. 6.2%)⁶.

The differential diagnosis should include other osseous and soft tissue giant cell tumors as well as aneurysmal bone cysts. These include pleomorphic sarcoma (malignant fibrous histiocytoma with abundant giant cells), non-skeletal osteosarcomas as well as giant cell tumors of the tendon sheath. Moreover, the spread of osseous giant cell tumors to soft tissue should be considered as well. Histogenesis of the tumor has not been fully elucidated. However, it has been proposed that the tumor develops by fusion of monocytes of giant cells⁷. As with its osseous counterpart, there are theories proposing that malignant transformation can occur either spontaneously or by impaired blood flow due to radiation or repeated surgical intervention, resulting in sarcomatous transformation in benign stromal cells⁷.

Cells comprising the tumor stroma are mononuclear cells with an appearance ranging from round to spindle-shaped. These cells have nuclei which are similar in appearance to multi-nuclear giant cells. Giant cells include about 20 nuclei, which are uniformly scattered within the lesion. In some series, the number of mitosis has been reported between 1-30 on 10x magnification, and no atypical mitosis has been reported in such cases⁷. In our case, the rate of mitosis was 3 on 10x magnification, and no atypical mitosis was observed.

In ST-GCT, degenerative changes, stromal bleeding and hemosiderin accumulation are seen in 50% of the cases, while foam macrophages and changes of aneurysmal bone cysts are present in 68% and 27% of cases, respectively^{6,7}. In our case, rather diffuse changes of aneurysmal bone cysts were noted. Aneurysmal bone cysts in soft tissue are distinct entities which had emerged in the 1990s^{13,14,15}. It is identical to its osseous counterpart on histology. Non-skeletal osteosarcomas are the primary sample of this entity. In our case, it was challenging to exclude aneurysmal bone cysts of soft tissue as these changes were markedly diffuse in our case. Accompanied with the fact that these changes may occur in ST-GCT, and the knowledge that these were not as prominent as in aneurysmal bone cysts, we were able to differentiate between these two entities. In addition, the uniform distribution of multi-nuclear giant cells rather than an accumulation around blood-filled cystic areas placed ST-GCT at the top of our differential diagnosis. Although reactive and matted osseous areas with a peripheral rim can be frequently seen in aneurysmal bone cysts, such cases are rare in ST-GCT¹⁵. No such changes were observed in our case.

In conclusion, ST-GCT is a rare tumor with low malignant potential, with similar gross features, microscopic characteristics, and clinical behavior to its osseous analog. Soft tissue tumors with abundant giant cells should be considered in the differential diagnosis. A benign clinical course is expected in most cases when complete excision is performed with negative margins. However, the patient should be closely monitored for local recurrence and distant metastasis.

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