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Case Report

Giant Cell Tumour of Soft Tissue – A Rare Neoplasm

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Abstract

Giant cell tumour of soft tissue is a very rare tumour that is clinically and histologically similar to giant cell tumour of the bone. The WHO currently classifies the tumours in the category of Intermediate (rarely metastasizing tumours). GCT-ST is required to be distinguished from giant cell rich tumours of the bone and other soft tissue neoplasms with giant cells. This is a case of a 63-year-old male with a giant cell tumour of soft tissue of the thigh. This tumour was not suspected clinically and the diagnosis was made only after histopathological examination. The case is presented for its extreme rarity and the differential diagnosis with other giant cell rich tumours is discussed.

Key words: Soft tissue neoplasms, Giant cell tumour of Soft Tissue, Thigh.

Introduction

Primary Giant Cell Tumour of Soft Tissue (GCT-ST) is a recently described rare neoplasm of low malignant potential. GCT-ST is a very rare primary soft tissue neoplasm that clinically and histologically resembles its counterpart, the giant cell tumour of bone. It is also known as osteoclastoma of soft tissue, and giant cell tumour of low malignant potential (GCT-LMP) and giant cell tumour of soft parts.[1,2,3] This tumour shows characteristic histological features and clinical behaviour that render it a specific entity.[3]

Case History

A sixty three year old male presented with a painless but gradually progressive swelling in the medial aspect of the left thigh since six months. There was no history of fever or previous trauma in that region. On examination, there was a soft cystic swelling measuring 6x6 cms. The swelling was fixed to the underlying soft tissues. Ultrasonography was done and the findings were suggestive of an organised abscess. An X-ray of the thigh was taken which did not show bone involvement [Fig 1]. A wide excision of the swelling was done and the specimen was sent for histopathological examination.

The specimen consisted of a well encapsulated globular mass measuring 7x5x1.5 cms. Outer surface was unremarkable. The cut surface appeared cystic and filled with hemorrhagic material [Fig 2]. Microscopy showed a highly cellular neoplasm composed of sheets of oval to elongated mononuclear cells admixed with evenly distributed multinucleated osteoclast-like giant cells [Fig 3]. The mononuclear cells had an open chromatin with one to two small nucleoli and ill-defined cytoplasm. Both cells types were neither pleomorphic nor atypical. The mitotic count was less than one mitosis per 10 high power fields. Bony trabeculae are seen at the periphery of the tumour. A fibrous capsule is seen around the tumour which appears focally breached. Areas of haemorrhage are seen. Angiolymphatic invasion was not identified. The histopathological findings were consistent with those of Giant Cell Tumour of Soft Tissue.

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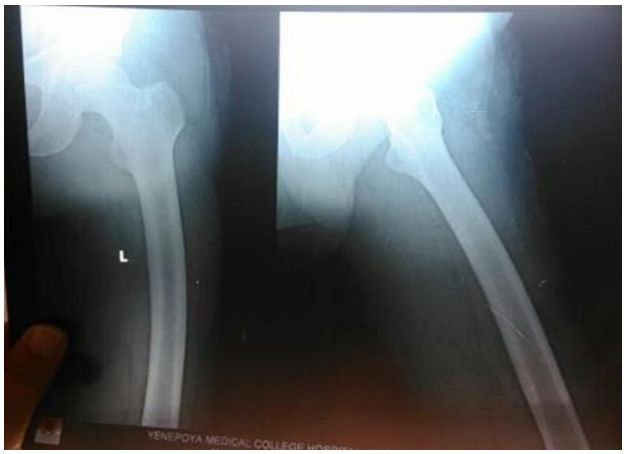

Figure 1: X-ray of the thigh showing uninvolved bone.

Figure 2 - The specimen consisted of an encapsulated globular mass measuring 7x5x1.5 cms. The cut surface was cystic and filled with hemorrhagic material.

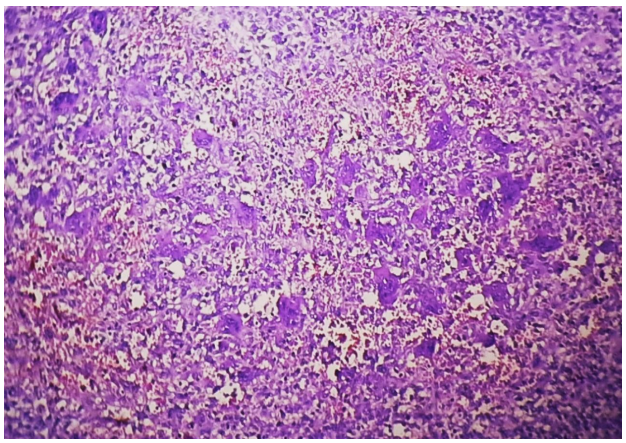

Figure 3 – Microscopy – a cellular neoplasm composed of sheets of oval to elongated mononuclear cells admixed with multinucleated osteoclast-like giant cells evenly distributed (H&E x100).

Discussion

Giant Cell Tumour of Soft Tissue was first described in 1972 in two simultaneous publications, one series reporting benign cases and another series with tumours showing aggressive behaviour.[4,5] This pathologic entity was originally called "malignant fibrous histiocytoma, giant cell type" based on the fact that these tumours were linked histogenetically to malignant fibrous histiocytoma.[4] Later, with a report of thirty-one additional cases with the same histopathological features but yet lacking marked cytological atypia it was reclassified as "giant cell tumour of low malignant potential".[6] Giant cell tumours of soft parts have now been reclassified as GCT-ST (or GCT-LMP) and undifferentiated pleomorphic sarcoma with giant cells (giant cell malignant fibrous histiocytoma or malignant giant cell tumour of soft parts in the current WHO Classification of Tumours of Soft Tissue and Bone.[1,3]

GCT-ST usually occurs in superficial soft tissues of the lower[1,5,7,8,9,10,11] and upper extremities.[1,5,7,8,11,12,13,14] Less frequently affected sites are the trunk,[1,5,7,11,15] pelvis [8,16] and neck regions.[1,3,17,18] Recently, tumours with similar histopathological features have been described in numerous different anatomic sites including breast,[19] mediastinum [20] and skin.[21] GCT-ST occurs predominantly in the fifth decade of life and affects both sexes in equal numbers.[1] The tumours present as painless growing masses [4,5,7,8,11] with an average duration of symptoms of 6 months.[4,5,7,8,11] No aetiological factors have been identified.[1]

The tumours are composed of a 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 vascularised stroma. The nuclei in the multinucleate cells are similar to the nuclei in the mononuclear cells. Mitotic activity may be present in every GCT-ST; typical mitoses range from 1 to 30 figures per 10 high-power fields. Atypia, pleomorphism, and tumoural giant cells are absent, and necrosis is found extremely rarely. Absence of bone involvement is a must for the diagnosis.[1,2,22,23] Metaplastic bone formation is present in approximately 50% of the tumours; frequently it is in the form of a peripheral shell of woven bone.[1,2,17,22,23] Metaplastic bone may be perhaps induced by secretion of Transforming Growth Factors beta 1 & 2 by the tumour cells.[22] The main differential diagnosis includes giant cell tumor tendon sheath (GCTTS), diffuse-type giant cell tumour, plexiform fibrohhistiocytic tumour, intravascular fasciitis and giant cell rich malignant tumours.

GCTTS, also called tenosynovial giant cell tumour, is an extra articular counterpart of pigmented villonodular synovitis, that is composed largely of mononuclear histiocytoid cells with scattered osteoclast and Touton type of giant cells, with xanthoma cells, siderophages, lymphocytes. The hand (fingers) is the common location. It is different from GCT-ST because of its usual localization near joint spaces or bursae, a generally uninodular rather than multinodular growth pattern which may be seen in GCT-ST, and paucity of the giant cells. Metaplastic bone production is not commonly seen in GCTTS. Although both tumours are composed of large mononuclear histiocytoid cells and osteoclast type giant cells, Touton type giant cells are also present in GCTTS. The ratio of giant cells to mononuclear cells is reversed in these two lesions and there is a heterogenous population of cells in GCTTS. Unlike GCTTS, large amounts of dense fibrous tissue with or without hyalinisation are absent or scarce in GCTSP. [1,2,3,24]

Diffuse–type giant cell tumour (pigmented villondular synovitis/tenosynovitis) is typically seen in younger patients. It may be intra articular (knee, hip, ankle, elbow, shoulder) or extra articular (knee, thigh, foot). It is a destructive proliferation of synovial-like mononuclear cells admixed with multinucleated giant cells, foam cells, siderophages and inflammatory cells.[1,2,3,24] The differential diagnosis also includes plexiform fibrohistiocytic tumor (PFT). This tumour has a dermal/subcutaneous location, and identification of bimodal population of cells in these tumours often allows for its distinction. Unlike GCTSP, which shows coarser multinodular pattern, PFT reveals plexiform growth pattern with minute nodules or short fascicles of fibroblasts at the dermal subcutaneous interface. In addition, PFT tends to arise in younger patients in a truncal location.[1,2,3,24]

Intravascular fasciitis (IF), are variant of nodular fasciitis may resemble GCTSP because of considerable number of multinucleated giant cells. Typical areas of nodular fasciitis characterized by somewhat randomly arrayed bland myofibroblastic cells that are commonly associated with with microcystic change should be searched for the diagnosis of IF. IF involves small or medium-sized veins or arteries in contrast to GCTSP wherein the bulk of the tumour is situated in the dermis. [1,2,3,24] GCTST should be considered different from giant cell rich malignant fibrous histiocytoma (MFH) (undifferentiated pleomorphic sarcoma with giant cells) because the latter is a more aggressive, high grade malignant tumour. Giant cell rich MFH is a multinodular, storiform-pleomorphic MFH with marked cytologic atypia, atypical mitoses, and benign osteoclast-like giant cells. However, GCTST lack the numerous atypical mitoses and coagulative tumour cell necrosis typical of giant cell rich MFH. The current choice of treatment is to combine aggressive surgical resection and postoperative radiotherapy for these malignant tumours. Therefore, GCTSP should be separated from giant cell rich MFH, because of their quite different biologic behaviour, and far better prognosis.[1,2,3,24] Similarly the absence of nuclear atypia, atypical mitoses and necrosis distinguishes it from other malignant tumours with giant cells.[1,2,3,24]

The treatment of GCTSP remains controversial because of the small number of reported cases.[3] A benign clinical course is expected if the lesion is excised adequately. Its biological behaviour to have low malignant potential is recognized; but this cannot be predicted and metastasis does occur rarely.[11] Complete removal of the GCT-ST with negative surgical margins assures no recurrence, but lung metastases have been reported in cases with positive surgical margins. The incidence of local recurrence is high. The recurrence rate is 28 % in skin and 45 % in deep soft tissue. All recurrent cases were positive in the surgical margins. Four cases of distant metastasis, all in the lungs, have been reported.[3,9,16] Radical surgery has been accepted as a mode of treatment for GCTST by some surgeons. However, because of the possibility of local recurrence, clinical follow up with or without postoperative radiotherapy is advised after excision. Although some authors advocate adjuvant radiotherapy as a treatment of GCTSP, many aspects of this mode of treatment have not been clarified.[3]

In conclusion, GCTSP should be kept in mind in the differential diagnosis of other osteoclast-like giant cell -rich tumours as the biological behaviour and treatment vary.

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