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Case Report

The rare giant cell tumor of soft tissue – An Indian perspective: A case report with review of the available Indian literature

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Abstract

Primary giant cell tumors of soft tissue (GCT-ST) bearing a resemblance to giant cell tumors of bone are rare but distinct entities, first described by Salm and Sissons in 1972, closely followed by Guccion and Enzinger. The low malignant potential but high propensity of recurrence is well recognized. The tumor is very rare in India, with approximately ten cases having been reported. Here we report a case of a giant cell tumor of soft tissue in a 63 year old male that was managed by performing a wide excision.

Keywords: giant cell tumor, histiocytoma, low malignant potential, mononuclear cells, multinucleated giant cells.

1. Introduction

Primary giant cell tumors of soft tissue (GCT-ST) bearing histological resemblance to their bony counterparts, are rare, but distinct entities.[1-3] Though classified initially along with more malignant forms of giant cell tumors, the low malignant form with an unpredictable clinical behavior has gradually gained prominence.[2] Altogether 56 cases had been reported till 2002,[2] and a search over the internet reveals altogether eleven cases being reported in India till date. Here we report the case of a 63 year old male who presented with a GCT-ST in the thigh.

2. Case report

A 63 year old gentleman came with the complaints of a painless swelling on the medial aspect of the upper one-third of the left thigh, for a period of six months, gradually progressing in size. Local examination revealed a single hemispherical swelling, approximately six cm in diameter. Fluctuation was negative, consistency, firm with restricted mobility. USG of the swelling revealed an organized hypoechoic collection measuring 6.4 x 4 cm in the intermuscular plane along the medial aspect of the thigh with diffuse internal echoes with central hypoechoic areas. Attempted aspiration yielded minimal blood. A computed tomogram revealed a 6 x 4 cm heterogeneous soft tissue mass with enhancing margins adjacent to the sartorius muscle. He underwent a wide excision of the swelling under spinal anesthesia (Figure 1).

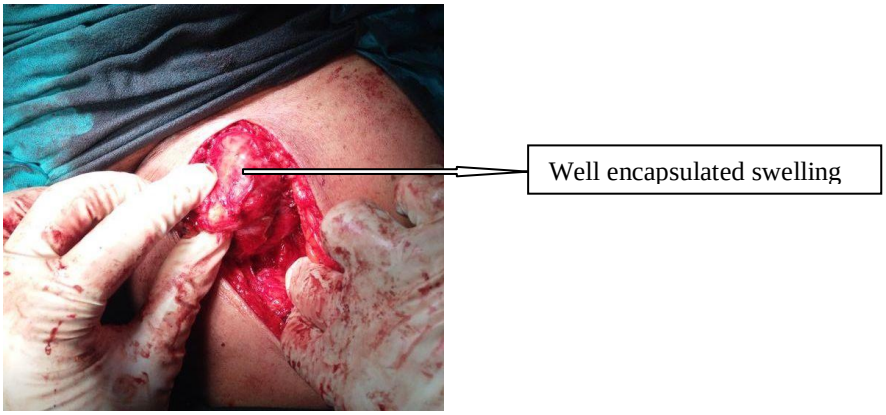

Figure 1: Intra operative photograph

Histopathological report showed a neoplasm composed of sheets of oval to elongated mononuclear cells with open chromatin, with one to two small nucleoli and ill-defined cytoplasm, admixed with well spaced multinucleated osteoclast-like giant cells (Figure 2).

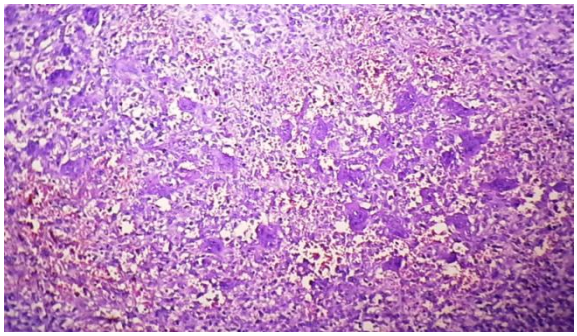

Figure 2: Microscopic appearance

Mitotic figures were occasional. Bony trabeculae (reactive bone) were seen at the periphery of the tumor suggestive of giant cell tumor of soft tissue.

3. Discussion and review of literature from India

Giant cell tumors of soft tissue of low malignant potential, also known as the primary giant cell tumor of soft tissue, are referred to as the soft tissue counterparts of giant cell tumours of bone.[1-4] These entities were first described in 1972 by Salm and Sissons as "Giant-cell tumors of soft tissues".[1] In the same year Guccion and Enzinger reported 32 cases, having a more aggressive histologic appearance and clinical behavior, and coined the term "Malignant giant cell tumor of soft parts",[1] postulating them to be a related form of malignant fibrous histiocytoma.[1-3,7,8] Later, Folpe *et al* reclassified it as "giant cell tumor of low malignant potential".[3] At present the general consensus is that two forms exist, one being of low malignant potential and the other one referred to as malignant giant cell tumor of soft parts, the latter having a higher propensity to recur. The first Indian case was reported by Mazhari *et al*, about a 22 year old male with a GCT-ST in the paravertebral region, presenting with bilateral lower limb weakness and bowel and bladder incontinence.[6] This was followed by a retrospective study of 7 cases located in the fingers (3 cases), thigh, palm, foot and the anterior abdominal wall (1 each), by Chand *et al* in 2006.[8] These preceded 3 individual case reports, by Mardi K in 2007 about a GCT-ST in a finger,[7] by Asotra S in 2009 about a GCT-ST in the thigh[4] and by Bhat A in 2013 about one over the ala of the nose.[5] Ours is the third case in India with a GCT-ST in the thigh. Studies done outside India showed maximum involvement in the extremities[1,3] and age range between 14 to 89 years.[1,3] Indian cases reported so far have ages ranging from 18 to 75 years.[4-8] All the tumors were subjected to wide excision and except one, none recurred.[4-8] Histologically, the common picture is the same as that in our case except that, ours is the only case which has shown metastatic reactive bone formation.[4-8] Transformation to a high malignant form is shown histologically in one case reported by Chand *et al*, that recurred.[8] No deaths have been recorded in India so far, though a few have been overseas.[1,2]

4. Conclusion

Our aim is to:

i) Assimilate the available Indian literature and compare histopathological trends of this rare entity with world literature, to effectively differentiate microscopically between low and high malignant forms that would aid in treatment
ii) Explore new treatment options including chemoradiotherapy
iii) Follow up treated patients to evaluate recurrence rates

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