Study of 7 Cases of Giant Cell Tumor of Soft Tissue

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Abstract

Background : Primary giant cell tumour of soft tissues is a distinct but uncommon group of neoplasms morphologically identical to osseous giant cell tumor.

Methods : 7 patients with painless growing soft tissue mass, having no attachment to underlying bone, were identified in a four years retrospective study from two zonal hospitals of armed forces. Histopathology of these lesions revealed admixture of multinucleated giant cell with mononuclear cells. All patients were treated by surgical resection and followed up for recurrence.

Results : There were 5 male and 2 female patients in the age group of 18 to 56 years. All lesions were superficial, circumscribed and involved extremities except one. Histologic transition between benign and malignant lesion was present in only one of the 7 patients that recurred after three months of surgery for which she had to be operated again. 2 of our 7 cases were lost in follow up.

Conclusion : Primary giant cell tumour of soft tissues usually present as a painless mass and needs to be differentiated from other giant cell rich soft tissue tumors. 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.

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Key Words : Giant cell tumour of soft tissue; Giant cell tumour of bone; Malignant fibrous histiocytoma

Introduction

Giant cell tumor of soft tissue (GCT-ST) is a relatively rare entity, and is clinically and histomorphologically indistinguishable from its counterpart in bone [1,2]. It affects adults of both sexes. The lesions are superficial and involve lower extremities and trunk. Although GCT-ST is predominantly a benign condition, its capacity to occasionally recur or evolve into a malignant lesion is well recognized. If surgically treated with wide excision, it is expected to have a benign clinical course [1,2].

Salm and Sisson [1] were the first to coin the term primary GCT-ST in 1972 in their study of 10 soft tissue tumors which were identical to osseous GCT. Recurrence occurred after surgical removal in two of their cases; but there was no instance of metastasis. In contrast Guccion and Enzinger, in the same year, found much aggressive behaviour of these tumors [2]. They noted the resemblance of these tumors to malignant fibrous histiocytoma and other workers gradually accepted this entity to be synonymous with giant cell rich malignant fibrous histiocytoma [3,4]. Later on, other investigators accepted this entity to have two distinct variants, a largely benign and a few having low malignant potential [5,6]. Flope et al preferred to classify these tumors as soft tissue analogue of GCT of low malignant potential due to varied biological heterogeneity [7].

GCT-ST is required to be distinguished from giant cell rich tumors of the bone and other soft tissue neoplasms. Scanty data regarding benign GCT-ST has prompted us to publish a series of 7 cases of this rare entity along with a short review of histopathological findings.

Material and Methods

This is a retrospective study of 7 cases of GCT-ST, identified between the years 2000-2003 from the histopathology files of Military Hospitals Dehradun and Jalandhar. None of these cases were clinically suspected as GCT-ST. For selection of the cases criteria chalked out by Connell et al [5] were followed i.e. primary soft tissue tumor with no attachment to the underlying skeleton, no previous history of GCT and histologic appearance of osteoclast like GC and mononuclear cells (MNC) demonstrating spindle and/or histiocytic/ epitheloid cells comprising at least 75% of the tumor. MNC should lack any specific lineage differentiation such as evidence of osteoblasts or smooth muscle differentiation. Microsections were stained with haematoxylin-eosin (H&E) stain. The tumors were classified as benign and malignant based on the degree of nuclear atypia and mitotic figures.

Results

Of the 7 patients, 5 were male and 2 female in the age group of 18 to 56 years. All lesions were superficial in location.

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The chief complaint was painlessly growing soft tissue mass, present on the upper and lower extremities except in one case where it was on the abdominal wall. All the lesions were circumscribed. There was no prior history of GCT. The provisional diagnosis in 4 out of 7 was implantation dermoid on palms/fingers. History of trauma was present in three cases where the lesions were present on fingers. In the other three, the clinical diagnosis was neurofibroma over the thigh, foot and abdominal wall respectively. Radiological evaluation wherever indicated, was done to exclude involvement of the bone. Routine haemogram and biochemical parameters were within normal limits. In 4 cases FNAC was asked for, which revealed cellular smears having pleomorphic spindled stromal cells having vesicular nuclei, predominant nucleoli and an occasional giant cell. All lesions were removed surgically with wide excision. Post-surgical recovery was uneventful.

Grossly, the tumors were fleshy ranging from 0.5 to 5.4cm in size, gray white or red in colour with tiny brownish areas in three cases. In another two cases cystic changes were seen. Gritty feeling on cut section was felt in two tumors, but no feeling of reactive bone was perceived around the tumor. There was no area of calcification or necrosis.

The H&E stained sections showed oval or spindle cells admixed with variable number of multinucleated osteoclast like giant cells distributed evenly throughout the tumor (Fig 1). The number of nuclei in a giant cell ranged from 3 to 18 (Fig 2). MNC varied from oval histiocytoid cells to plump spindle cells. Two of the tumors exhibited small areas of necrosis. Mitotic activity limited to MNC was low (mean, 4 mitotic cells per 10 high power fields (HPF)). Areas of haemosiderin deposits were seen in some of the cases. No metaplastic bone formation was identified in our series. Cellular atypia, nuclear pleomorphism, hyperchromasia and raised mitotic figures suggestive of malignant changes were not seen. Angiolymphatic invasion was not identified. A few tumors showed relative hypocellular bands of collagenous tissue. Histologic transition between benign and malignant lesion was present in one arising from the abdominal wall in a female. It recurred after three months of surgery for which

she had to be operated again. Review of the slides did not reveal any feature suggestive of sarcomatous stroma. It was perhaps inadequate surgical removal rather than aggressive nature of the tumor, as the lesion did not recur on follow up for two years. There was no other case of recurrence or metastasis. Two of our 7 cases were lost to follow up.

Discussion

Primary GCT-ST is predominantly a benign tumor. Its capacity to recur or to evolve into a malignant lesion is recognised, but with exceedingly low recurrence rate, provided the lesion is removed adequately [1,2,4,5]. Some workers feel that malignant GCT-ST is a variant of malignant fibrous histiocytoma or fibrosarcoma with osteoclastic differentiation [3,4]. In malignant GCT-ST, there is histological evidence of sarcomatous change as indicated by cellular and nuclear pleomorphism, areas of haemorrhage, necrosis with high mitotic activity of MNC histiocytes and fibroblasts. The MNC in benign lesions are devoid of atypia, pleomorphism and atypical mitosis but no histopathologic feature is pathognomonic of aggressive behaviour of the tumor. An average number of mitotic figures in benign GCT-ST varied from 2-3/10 HPF in one series [7] to 9.5/10 HPF in another series where 6 out of 22 tumors revealed vascular invasion but these tumors did not metastasize [4]. Connell et al noted two striking differences in their study of 18 benign and malignant tumors. These were frequent mitotic figures (mean, 25 mitoses per 10 HPF) and conspicuous nuclear pleomorphism and hyperchromasia in MNC with common occurrence of hyperlobated nuclei in giant MNC [5].

The actual histogenesis of the tumor is not clear but it is postulated that giant cells are the result of fusion of circulating monocytes recruited into the lesion. Fibroblasts are plump or elongated and some resemble histiocytes. In its osseous counterpart, McGarth claimed

Fig. 1 : Multinucleated osteoclast like giant cells evenly distributed in a background of mononuclear cells. A few collagenous bands are also seen (H & E x 100)

Fig. 2 : Spindled mononuclear cells with vesicular nuclei, admixed with giant cells having 3 to 15 nuclei in the same tumour (H & E x 400)

that malignant transformation may occur spontaneously, after irradiation or repeated surgical intervention with curettage suggesting the possibility of mechanical disruption with access to blood stream, which may cause metaplasia of the benign stromal cells to sarcoma [8]. He subdivided malignant transformation of GCT of bone into primary, in which the tumor is malignant from the onset (de-novo), evolutionary, in which the tumor progressed to malignancy in a short period and lastly secondary, where sarcomatous change appeared after a relatively long symptom free period usually after radiation or repeated surgical intervention. This etiopathogenesis should also stand true for GCT-ST, since the GCT of bone and soft tissue are identical.

Majority of the tumors reported in the literature are located in the lower extremity, thigh being the most common site followed by trunk and upper extremities [1-5]. Rarely it may involve superficial and deep body tissue. They present less frequently, as a skin tumor [11]. A case has been reported in the paravertebral region opposite D3-D5 [9]. The tumor involves both sexes at all ages and presents as a painlessly, well-circumscribed, superficial mass. Duration of the complaint is less than one year but cases with duration up to 15 years are also on record [1].

In this small series, the commonest site was the upper limb, finger being the most common location. Most of the tumors in our study were solid, fleshy, reddish brown in colour; a few had tiny areas of haemorrhage. Some workers could shell out the tumor along a pseudocapsule [5]. We did not find foamy histiocytes, epithelioid type of MNC or storiform pattern of arrangement in any of our cases. Fibrohistiocytic areas and metaplastic bone formation, as identified by some workers, were also absent.

Flope et al, in their study of 31 soft tissue tumors having mild to moderate nuclear atypia, proposed the entity as GCT of low malignant potential and regarded it as soft tissue analogue of malignant GCT of bone [7]. They followed-up 19 cases after surgery. Recurrence occurred in 4 but none developed metastasis. This behaviour contrasts significantly from the high-grade behaviour traditionally associated with malignant GCT

of soft parts and malignant fibrohistiocytic tumors [10].

We feel that GCT-ST is a distinct entity histologically identical to its osseous counter part as a primary neoplasm within the soft tissue. Prognosis of GCT-ST varies and the biological aggressive course for its local recurrence cannot be predicted. Proper surgical excision and a long period of follow up are essential in these cases. These neoplasms should be distinguished from other giant cell rich soft tissue tumors with which these may be confused.

Conflicts of Interest

None identified

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