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Giant Cell Tumor of Soft Tissues with Neurofibromatosis: A Case Report

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Abstract

Giant cell tumor of soft tissues (GCT-ST) is a rare entity with unpredictable clinical behavior. However, metastasis and tumor-related death are rarities if these are adequately treated. GCT-ST is required to be distinguished from giant cell-rich tumors of the bone and other soft tissue neoplasms. Coexistence of multiple neurofibromas with GCT-ST is cited in very few literature reports, hence this case report.

Key words: *Giant cell tumor, soft tissue, neurofibroma, osteoclastoma*

Introduction

A Giant cell tumor of soft tissue (GCT-ST) is a relatively rare entity affecting adults of both sexes. These are superficial lesions described in wide anatomic sites, including extremities, trunk, head and neck, fascia and skeletal muscle [1-4]. GCT-ST is required to be distinguished from giant cell-rich tumors of the bone and other soft tissue neoplasms [5]. Although GCT-ST is a benign tumor, recurrence and malignant transformation are well recognized [2,4,5]. Lesions are likely to have a benign clinical course if wide surgical excision is done.

Case report

We report a case of a 26-year-old male with multiple painless nodules over the face, neck, trunk and back, along with a solitary painless swelling over the scalp for 6 months. Routine haemogram and biochemical parameters were within normal limits. Computed tomography of the swelling scalp showed a well-defined soft tissue ovoid lesion in the frontal region, more to the right side, measuring 38x26 mm. The adjoining frontal bone showed diploic widening. No focal lytic lesion or intracranial extension was seen. Fine-needle aspiration

from multiple nodules showed clusters of spindle-shaped cells with twisted nuclei in places, confirming them to be neurofibromas. FNAC from the swelling scalp revealed a giant cell-rich lesion with osteoclastic giant cells favoring GCT-ST (Figure 1). Excision biopsy was done a month later. The H&E-stained sections showed a tumor with relatively hypocellular bands of collagen dividing the tumor into partial nodules. The tumor was composed of oval to spindle cells with irregularly interspersed numerous multinucleated osteoclast-like giant cells. The number of nuclei in these giant cells ranged from 3 to 32. The mitotic count of 3 per 10 high-power fields was noted in stromal cells. Vascularity was prominent (Figure 2). Focal areas with hemosiderin deposits were seen, which was confirmed on Perl's Prussian blue reaction. Cellular atypia, nuclear pleomorphism, hyperchromasia and a raised mitotic count suggestive of malignant changes were not seen, nor was lymphovascular invasion seen. Areas of necrosis, cystic change or metaplastic bone formation were absent.

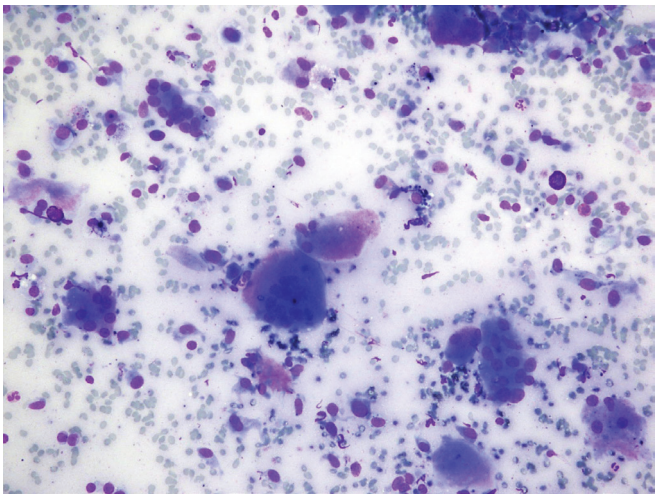

Figure 1. FNA smear showing multinucleated giant cells with dispersed stromal cells (MGG X 200).

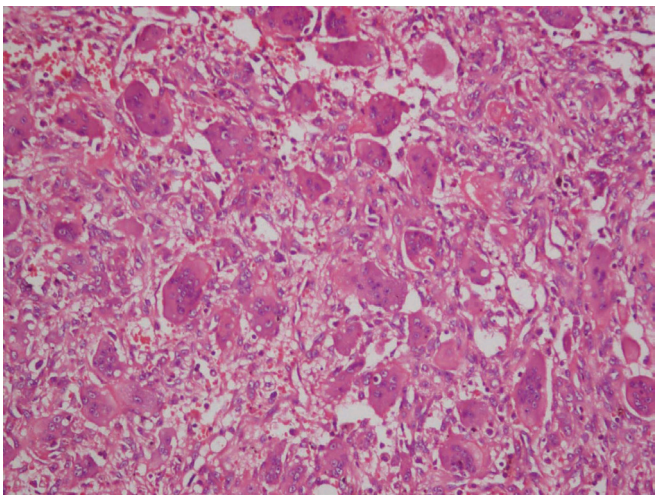

Figure 2. Photomicrograph showing uniformly distributed osteoclast-like giant cells with stromal cells in the background and pronounced vascularity (H&E X 200).

Discussion

GCT-ST is a rare tumor first described by Salm and Sissons [5] in 1972. More recently, Flope et al. [6] proposed the term 'giant cell tumor of low malignant potential'. Not many cases of GCT-ST have been described in the literature. Furthermore, the concomitant presence of multiple neurofibromas and GCT-ST is exceptional. The actual histogenesis of the tumor is unclear, with a postulation that giant cells result from the fusion of circulating monocytes recruited into the lesion [7]. It has been shown that human osteoclast precursors circulate in the peripheral blood in the monocytic fraction [8].

Most cases have been described in the thigh, trunk and upper extremities, and rarely in the head and neck, as in the present case [1-4]. Duration of the complaint is less than one year in most cases, like ours, but cases with longer duration are on record [5].

Histomorphologically, GCT-ST is similar to its bony counterpart [9]. An admixture of mononuclear cells with round to oval nuclei and osteoclast-like multinucleated giant cells is seen. The average number of mitotic figures in benign GCT-ST varied from 2-3/10 high-power fields (HPF) to 9.5/10 HPFs in different studies [4,6]. We observed a count of 3/10 HPFs. Other changes reported include metaplastic bone formation at the periphery in 40-50% and aneurysmal bone cystic changes in approximately 30% of the tumors[4,10]. Our case lacked both of these changes. Foci of necrosis are rare. Although not pathognomonic, cellular and nuclear atypia, pleomorphism, high mitotic activity, necrosis and hemorrhage are some of the pointers towards malignant transformation [5,7,10]. O'Connell et al.[10] observed frequent mitotic figures (mean 25/10 HPFs), and conspicuous nuclear pleomorphism and hyperchromasia along with hyperlobated nuclei in giant cells in malignant tumors feature, which were not seen in the present case.

Histopathologically, GCT-ST should be separated from other giant cell-rich lesions as it has a prognostic and therapeutic connotation. These include giant cell

tumors of tendon sheath, extraskeletal osteosarcoma, giant cell-rich tumors, pleomorphic sarcoma, epithelioid sarcoma, and leiomyosarcoma, or other benign reactive processes containing abundant osteoclast-like giant cells [11].

GCT-ST is a distinct entity with a histological resemblance to its osseous counterpart. The prognosis of GCT-ST is variable, with rare recurrence and metastasis on record [12]. The need for distinction from other giant cell-rich lesions cannot be overemphasized. The coexistence of this distinct entity with multiple neurofibromas is incidental, and extremely rare [13].

Conflict of interest statement

The authors have no conflicts of interest to declare.

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