

Soft Tissue Giant Cell Tumour of Low Malignant Potential: A Rare Tumour at a Rare Site

AMOOLYA BHAT¹, GEETHAMANI V.², VIJAYA C.³

ABSTRACT

“Soft tissue giant cell tumour of low malignant potential” is considered as the soft tissue counterpart of osteoclastoma of the bone. It is a primary soft tissue tumour which is classified under the category of fibrohistiocytic tumours of intermediate malignancy. Seventy percent of the tumours involve the extremities and only about seven percent of them arise in head and neck region. They are composed of nodules of histiocytes in a vascular stroma, with multinucleated osteoclast-like giant cells positive for vimentin, smooth muscle actin (SMA), CD68 and Tartrate Resistant Acid Phosphatase (TRAP). We are presenting a case of a 75-year-old man who had a nodule on the ala of the nose. Histopathology showed a histiocytic lesion. Benign fibrous histiocytoma, plexiform fibrohistiocytic tumour, solitary reticulohistiocytoma and histioid leprosy were ruled out by using special stains and immunostains. Expression of smooth muscle actin and CD68 confirmed the diagnosis of a soft tissue giant cell tumour with a low malignant potential.

Keywords: Giant cell tumour, Malignant potential, Soft tissue

CASE REPORT

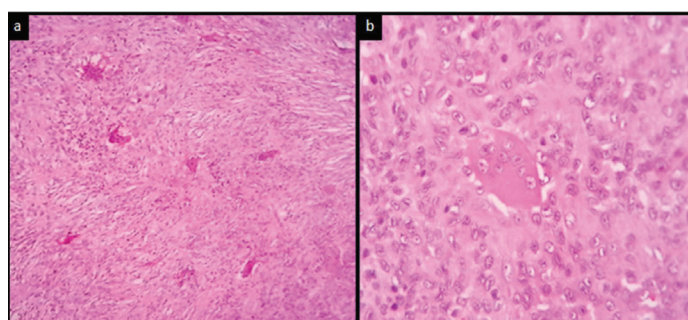
A 75-year-old man presented with a painless, slowly growing swelling over the ala of nose, of six months duration. There was no significant past or present history. On examination, a non-tender, firm, round nodule, raised above the level of surrounding skin, measuring 0.5 × 0.5 cm was seen over the ala of the nose. The skin over the swelling was stretched out, hyperpigmented and ulcerated. The nasolabial fold was unremarkable and it was uninvolved by the nodule. Fine-needle aspiration of the nodule was not attempted, as the size of the nodule was small and it was diagnosed as a sebaceous cyst, based on clinical features. A complete surgical excision of the lesion was performed.

The specimen measured 0.5 × 0.5 cm; cut surface was grey-brown. Microscopically, there was a nodular dermal tumour covered by hyperkeratotic, parakeratotic and ulcerated epidermis. The tumour was composed of sheets of round to oval cells with open nuclear chromatin and convoluted nuclei admixed with osteoclast-like giant cells having similar nuclear features, arranged in a nodular pattern [Table/Figs-1a and 1b]. Mitotic index was 0-1 per high power field. Angiectatic spaces [Table/Fig-2a] and spindle cell areas were noted [Table/Fig-2b]. Scattered haemosiderin laden macrophages and clusters of foamy histiocytes were also present. There was no necrosis. The deep surgical margin was free of tumour invasion. Benign fibrous histiocytoma, soft tissue giant cell tumour with low malignant potential, plexiform fibrohistiocytic tumour, solitary reticulohistiocytoma, nodular fasciitis with giant cells, and histioid leprosy were considered under the differential diagnosis. Fite-Feraco staining showed negativity. Immunohistochemical markers included CD68 and smooth muscle actin.

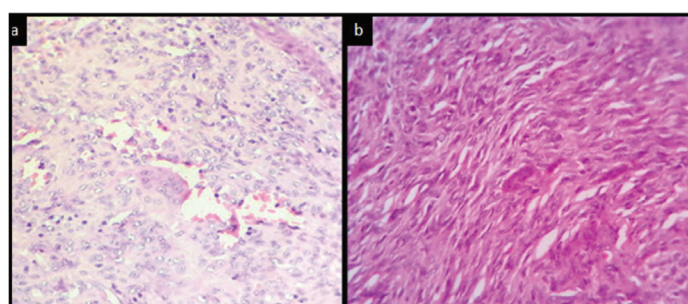
There was strong cytoplasmic CD68 expression in the multinucleated giant cells and in occasional mononuclear cells [Table/Fig-3a]. Smooth muscle actin was detected in mononuclear histiocytes [Table/Fig-3b]. Thus, a final diagnosis of soft tissue giant cell tumour with low malignant potential was established. Orthopaedic examination did not reveal bony lesions.

DISCUSSION

The low incidence and rare site of occurrence made us think of other



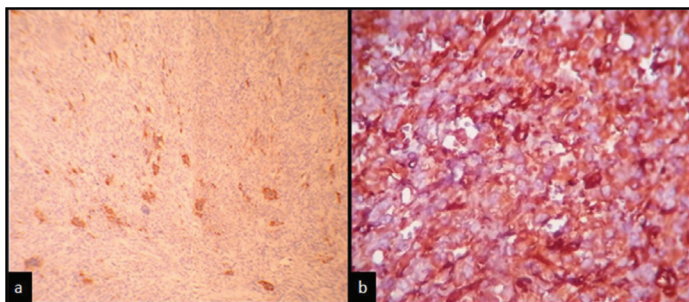
[Table/Fig-1]: (a) Showing mononuclear histiocytes and scattered osteoclast-like giant cells, H&E 100X; (b) Showing mononuclear histiocytes and a multinucleated osteoclast-like giant cell having similar nuclear features, H&E 400X



[Table/Fig-2]: (a) Angiectatic spaces surrounded by histiocytes and giant cells, H&E 400X; (b) Showing spindling of the cells, H&E 400X

common lesions than Soft tissue giant cell tumour of low malignant potential, in this case. Presence of foamy histiocytes and dermal location pointed towards a possibility of Histioid leprosy. However, the lack of other clinical lesions of Hansen's disease and negative Fite-Feraco staining ruled out this diagnosis.

The spindle cells, histiocytes and haemosiderin laden macrophages are characteristic of benign fibrous histiocytomas which are frequently seen in head and neck area and commonly involve dermis [1,2]. Osteoclast-like giant cells can occur in benign fibrous histiocytomas [3,4]. The lack of whorling, predominance of histiocytes having indented nuclei and expression of smooth muscle actin in the histiocytes, made this diagnosis unlikely.



[Table/Fig-3]: (a) Showing CD 68 expression in the osteoclast like giant cells and occasional mononuclear cells; b) Showing expression of smooth muscle actin in the mononuclear histiocytes

Angiomatoid fibrous histiocytomas show histiocyte-like cells and vascular spaces lined by histiocytes, but they also have a dense lymphoplasmacytic cuff at the periphery, which was lacking in our case. They are rare in adults and smooth muscle actin is expressed in spindle cells only [1].

Plexiform fibrohistiocytic tumours occur exclusively in children and they are composed of histiocytes and giant cells arranged in nodules, circumscribed by short fascicles of fibroblastic cells in deeper dermis and subcutis. Although these histiocytes and multinucleated giant cells express CD68, they lack smooth muscle actin. Only spindle cells express smooth muscle actin [1].

Nodular fasciitis with giant cells can occur at any age and site. It is composed of plump myofibroblasts arranged in short irregular bundles in a myxoid matrix admixed with scattered lymphocytes, macrophages and multinucleated osteoclast-like giant cells. However, smooth muscle actin is expressed only in the spindle cells; the mononuclear and multinucleated histiocytes are negative [1].

Solitary reticulohistiocytomas develop in adults at any site. They show oncocyctic epithelioid histiocytes positive for CD68 and lysozyme and multinucleated giant cells. Smooth muscle actin is usually not expressed [5]. The histiocytes in our case lacked the abundant eosinophilic cytoplasm and they expressed smooth muscle actin.

Giant cell tumour of the tendon sheath has similar histological features, but they arise within the tendon sheath. Giant cell forms of malignant fibrous histiocytomas and epithelioid sarcomas display significant nuclear atypia, necrosis and increased mitosis, which

were lacking in our case [1]. Amelanotic melanomas show marked pleomorphism and are negative for smooth muscle actin [6].

Complete excision with uninvolved surgical margins is the treatment of choice for soft tissue giant cell tumours of low malignant potential [7]. Local recurrences are known to occur following incomplete excisions [2]. A recurrence rate of twelve percent has been observed, but metastasis and death are unusual [1,2].

CONCLUSIONS

We have described a rare case of "Soft tissue giant cell tumour of low malignant potential", involving the head and neck area. Combination of histopathology and immunostains for smooth muscle actin and CD68 confirmed the diagnosis and ruled out the various neoplastic and non neoplastic differential diagnoses which vary in therapeutic aspects. The knowledge on existence of such an entity and its natural course help in providing better care for these patients.

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