An Extremely Uncommon Case of Giant Cell Tumor of Skin: A Case Report in a 16-year-old Female

Lalit Kumar, Pooja Agarwal, Afreen Parvez, Yatendra Chahar[1]

Department of Pathology and [1]Department of Skin, S.N. Medical College Agra, Agra, Uttar Pradesh, India

Abstract

Giant cell tumors of the skin are known to be extremely rare tumors, grossly, and histologically similar to that of giant cell tumors of bone. A 16-year-old girl presented with an asymptomatic swelling over the right cheek, which had progressed over 5 months duration without any antecedent history of local trauma and infection. Grossly, the specimen was brown-colored without any pigmentation, fleshy, and consisted of a skin-covered globular mass measuring 1.5 × 1.0 × 0.5 cm. On histopathologic examination, sections examined show a well-circumscribed lesion involving the dermis and revealed biphasic population of round to spindle-shaped mononuclear cells with intimately admixed osteoclast-like giant cells. On immunohistochemistry, osteoclast-like giant cells and mononuclear cells showed strong cytoplasmic granular positivity for CD68 and final diagnosis of giant cell tumor was given.

Keywords: CD68, giant cell tumor, osteoclast-like giant cells

Introduction

Giant cell tumors of the skin are known to be extremely rare tumors, involving commonly the extremities, head, and neck regions, which are grossly and histologically similar to that of giant cell tumors of bone.[1] Histologically, these tumors show round- to spindle-shaped cells admixed with uniformly scattered osteoclast-like multinucleated giant cells.[2] To the best of our knowledge, less than 10 cases of this entity have been reported in the literature so far. We report the clinical and histologic features of giant cell tumor of the skin in a 16-year-old girl, which is believed to be the ninth reported case of giant cell tumor of skin as a primary lesion at this site [Table 1].

Case Report

A 16-year-old girl presented with an asymptomatic swelling over the right cheek on the facial region, which had progressed over 5 months duration without any antecedent history of local trauma and infection. On local examination, the swelling was well-circumscribed, no skin color changes (similar to adjacent skin), no ulceration, mobile, nontender, adherent to skin, and measuring about 2.0 × 1.5 × 1.0 cm in size. No regional lymph nodes were involved. X-ray showed no bony involvement. The clinical diagnosis of granulomatous lesion of skin was given. The lesion was excised and sent for histopathologic examination. Grossly, the specimen was brown-colored without any pigmentation, fleshy, and consisted of a skin-covered globular mass measuring 1.5 × 1.0 × 0.5 cm. On histopathologic examination, sections examined show a well-circumscribed lesion involving the dermis and revealed biphasic population of round- to spindle-shaped mononuclear cells with intimately admixed osteoclast-like giant cells [Figures 1–3]. The cells had a moderate amount of granular eosinophilic cytoplasm and oval- to spindle-shaped nuclei with vesicular chromatin and prominent nucleoli. The tumor giant cells had multiple nuclei similar to those of mononuclear cells and eosinophilic granular cytoplasm. The mononuclear cells showed mild pleomorphism and occasional mitotic activity.

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Address for correspondence: Dr. Pooja Agarwal, Department of Pathology, S.N. Medical College Agra, 79, Gandhi Nagar, By Pass Road, Agra, Uttar Pradesh 282003, India.
E-mail: drpooja.agarwal@gmail.com

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A provisional diagnosis of giant cell tumor of the skin was made. On immunohistochemistry, osteoclast-like giant cells and mononuclear cells showed strong cytoplasmic granular positivity for CD68 [Figure 4] and final diagnosis of giant cell tumor was given.

Discussion

Giant cell tumors of the skin, which are extremely rare tumors, resembles their osseous variants both grossly and histologically. Age of presentation of these tumors is 68–78 years (median age 73 years) with an M:F ratio of 3:2.[1,6] In 1972, Salm and Sissons[7] first described the giant cell tumor of soft tissue, which formerly comes under the term "malignant giant cell tumor of soft parts." Guccion and Enzinger reported the tumor of soft tissue with the same characteristic features but with aggressive malignant transformation as atypia, abundant mitotic activity, and pleomorphism.[8] Folpe *et al.* reclassified them as "giant cell tumor of low malignant potential" because on further pathological analysis they found lack of cytological atypia even with increased mitotic activity and vascular invasion.[9] The extremities, head, and neck are commonly involved sites by this tumor. These tumors are well-circumscribed, unencapsulated, and multinodular with a mixture of round- to spindle-shaped mononuclear neoplastic cells and osteoclast-like giant cells scattered uniformly. Osteoclast-like giant cells have voluminous eosinophilic cytoplasm with 50–100 small nuclei, which arise due to fusion or by amitotic nuclear division of precursor mononuclear cells. The histogenesis is not clear. However, previously it was considered as one of the histologic types of malignant fibrous histiocytoma but not favored so long.[3] The osteoclast-like giant cells and mononuclear cells show strong positivity for CD68, alpha-1 antitrypsin, and alpha-1 antichymotrypsin, whereas these cells are negative for cytokeratin (AE1/AE3) and S100 protein.[2] Differential diagnosis of this tumor includes benign fibrous histiocytoma, atypical fibroxanthoma, and giant cell tumor of bone with soft tissue extension. Benign fibrous histiocytoma with many osteoclast-like giant cells can be differentiated by the presence of hyperplastic epidermis, hyperpigmentation of

Table 1: Brief review of giant cell tumors of skin

Study	Patient age in years	Sex
Hoang *et al*. (2002)[2]	6–78 (Five cases)	3 Male and 2 Female
Kumar *et al*. (2006)[3]	55	Male
Lentini *et al*. (2010)[4]	79	Female
Murphy *et al*. (2011)[5]	92	Female
Present case	16	Female

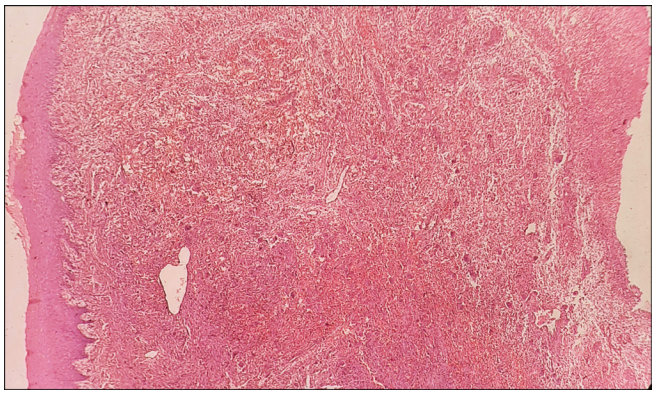

Figure 1: Well-circumscribed lesion involving dermis (H&E, 4X).

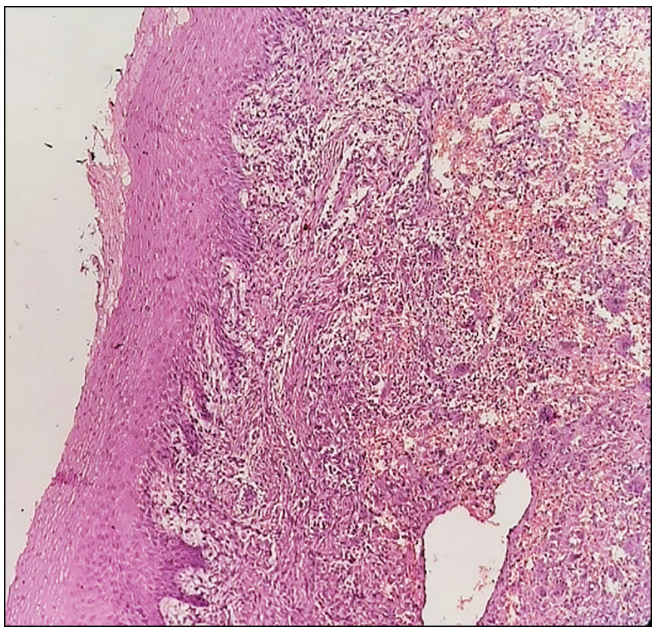

Figure 2: Epidermis along with mononuclear cells and admixed osteoclast-like giant cells with hemorrhage in dermis (H&E, 10X).

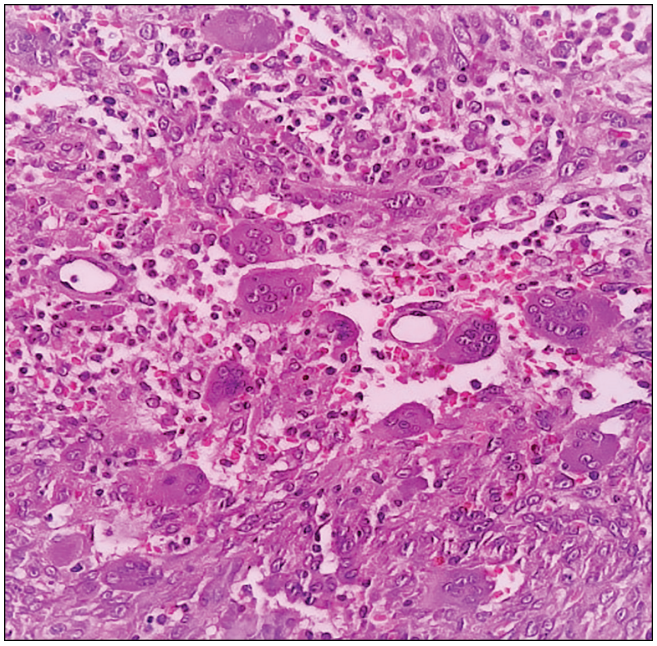

Figure 3: Biphasic population of round to spindle-shaped mononuclear cells with intimately admixed osteoclast-like giant cells (H&E, 40X).

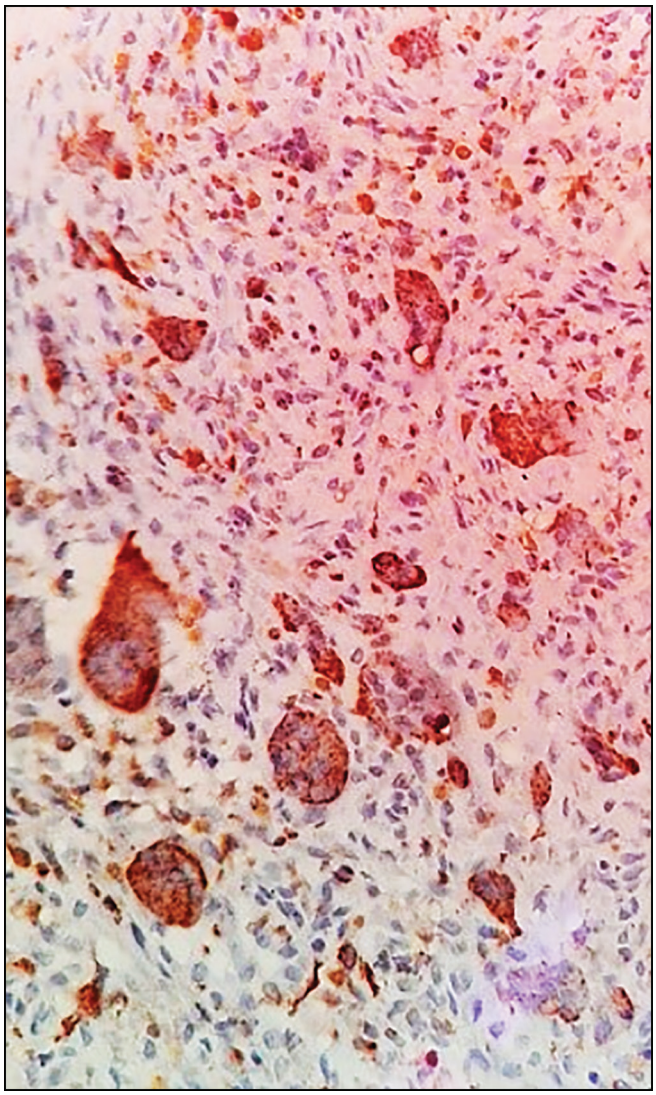

Figure 4: Osteoclast-like giant cells and mononuclear cells showing cytoplasmic granular positivity for CD68 (IHC, 40X).

the basal layer, and elongation of rete ridges.[10] Atypical fibroxanthoma shows pleomorphic histiocytes-like cells and atypical giant cells, often with bizarre nuclei and numerous mitotic figures.[2,10] Giant cell tumor of bone with soft tissue extension shows radiologically, an osteolytic lesion in epiphysis and presence of a rim of ossification at the edge of the tumor.[11] Extraskeletal osteosarcoma can be differentiated by the presence of neoplastic bone or osteoid.[12] Both benign fibrous histiocytoma and atypical fibroxanthoma show resemblance with this tumor and can only be differentiated by histopathologic studies. Giant cell tumor of bone also shows a lot of similarities with tumor and radiologic studies are needed to differentiate between both of them. Cutaneous giant cell tumors are low-grade sarcomas that can recur locally and infrequently metastasize. One case with lung metastasis has been reported in the literature. Superficial tumors have a better prognosis than deeper ones; 75% of superficial tumors recur and 25% metastasize, whereas about 50% of deep tumors recur and about 50% metastasize.[2]

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Conflicts of interest

There are no conflicts of interest.

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