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Case Report

Giant cell angiofibroma of the scalp: A benign rare neoplasm with bone destruction

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Abstract

Background: The incidence of extraorbital giant cell angiofibroma (GCA) is rare, with only one case located in the scalp reported in the literature. The morphological hallmark is histopathological examination showing richly vascularized pattern-less spindle cell proliferation containing pseudovascular spaces and floret-like multinucleate giant cells.

Case Description: We report a case of a 30-year-old female with a primary complaint of a painless solitary nodule arising on the left parietal region of the scalp. Complete tumor removal through surgical intervention was achieved, and the postoperative period was uneventful.

Conclusion: Diagnosing a highly vascularized tumor in the head and neck is challenging. Our case is unique in that it is presented as a GCA of the scalp, which is an extremely rare clinical entity, and also demonstrated bone destruction.

Key Words: Bone destruction, giant cell angiofibroma, scalp

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INTRODUCTION

Giant cell angiofibroma (GCA) is a nonrecurring benign neoplasm that was first described in 1995 in a study of seven patients with a distinctive orbital tumor.[2] Since then, approximately 39 cases have been reported in the literature. Most of these lesions involve the orbital region, and it predominates in males; however, females predominantly present GCA at extraorbital sites, including the nasolacrimal duct, buccal mucosa, submandibular region, trunk, mediastinum, retroperitoneum, vulva, and extremities.[2,8] The incidence of extraorbital GCA is low, and only one case of scalp GCA has been reported so far.[9] GCA may belong to a solitary fibrous tumor (SFT) group, as it contains multinucleated giant stromal cells and angiectoid space.[14] GCA may also grow rapidly and stimulate an aggressive or malignant process. It may also have an indolent course; it sometimes presents as a painful mass that grows slowly or remains stable in size over many years.[5,8]

CASE REPORT

A 30-year-old female presented with a bony swelling over her left parietal region that had increased in size over the past 2 years. She had no history of trauma, bone pain, systemic disease, or neurological symptoms. Physical examination found a swelling that was 6 × 5 cm

in size, soft, and without any tenderness [Figure 1a]. The skin over the swelling area was loosely attached to the underlying soft tissue. There was no venous dilatation, scars, sinuses, or any bruit heard over the swelling. Neurological examination was unremarkable and fundoscopy examination revealed no papilledema.

Radiological examination of a skull x-ray showed a lytic lesion that involving the left temporoparietal bone, and computed tomography (CT) scan confirmed the presence of extensive bone destruction [Figure 1b], with a well-defined soft tissue lesion expanding into the temporal muscle. We were unable to obtain a magnetic resonance imaging (MRI) scan for this patient. On clinical grounds, the decision was made to operate immediately.

The tumor mass was approached through a horseshoe scalp incision [Figure 2a and b]. During the operation, the mass appeared to be eroding through the internal tabule of the bone and infiltrating the muscle, but no mass transversed or invaded the dura mater. Temporal muscle that been infiltrated by the GCA (size 2 × 1 cm) was removed [Figure 1c]. The bone surrounding the tumor mass appeared rarefied and was removed with rongeurs until a normal border was identified [Figure 2c and d]. The postoperative bone defect was 7 × 6 cm in size and was closed using titanium mesh. The patient's neurological status was intact postoperatively.

The histopathological examination showed both cellular areas with especially oval and round- to spindle-shaped cells intermixed with floret-like giant cells in no particular pattern and hypocellular areas. Giant cells often lined the pseudovascular spaces [Figure 3a and b]. The pathologic exam also showed bone erosion without any signs of reformation [Figure 3c] and hypervascularization demonstrated by varying calibers of blood vessels. Immunohistochemistry staining showed positive CD34 staining [Figure 3d]. Mitotic activity was inconspicuous. On the basis of these findings, a diagnosis of GCA of the scalp was made.

She was discharged from the hospital uneventfully, and no recurrence of the mass was detected at her one-year follow-up.

DISCUSSION

GCA was first described in 1995 by Dei Tos *et al*. as a distinctive orbital tumor occurring exclusively in male adults.[2] GCA is a rare mesenchymal neoplasm. According to some authors, the histologic features are intermediate between SFT and giant cell fibroblastoma of soft tissue.[2,4] GCA was initially described as a potentially recurrent tumor in the orbit of adults. However, it can also present in other locations, and 18 cases have been reported in extraorbital sites, such as submandibular, parascapular, and retroauricular sites and the thigh, mediastinum, back, scalp, retroperitoneum, vulva, hip, forearm, groin, parotid, and neck.[1,6,10,12] The median age of affected patients is 45 years, with a range of 18–81 years. Interestingly, extraorbital lesions predominate in females, who account for 66% of the cases.[1] Patients typically present with a solitary painless soft tissue mass. Other symptoms depend on the site of the tumor and are secondary to the local mass effect, including pain, visual disturbances, and respiratory difficulty.[2,10]

Although GCA is considered a benign neoplasm, it has the potential to recur locally, especially after an incomplete resection. Histologically, the tumor displays a varying combination of a cellular area composed of bland

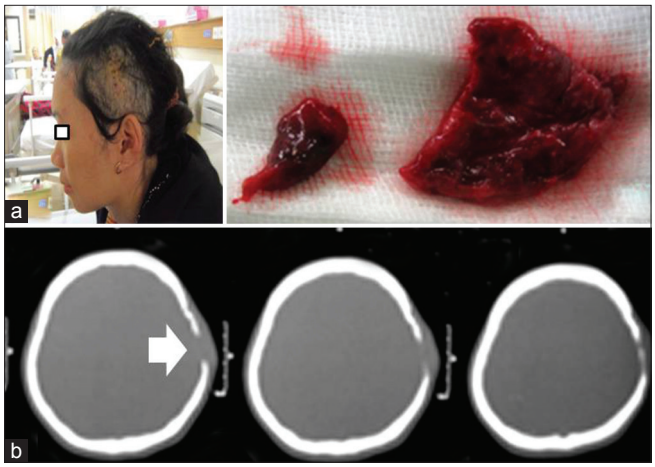

Figure 1: Clinical picture of the patient that had a bony swelling over the left parietal region (a). A CT scan at regio left parietal reveal bone discontinuity that showed bony destruction (b). On macroscopic examination, reddish mass, size 6 × 5 cm and the temporal muscle, size 2 × 1 cm that had been infiltrated with GCA, were removed (c)

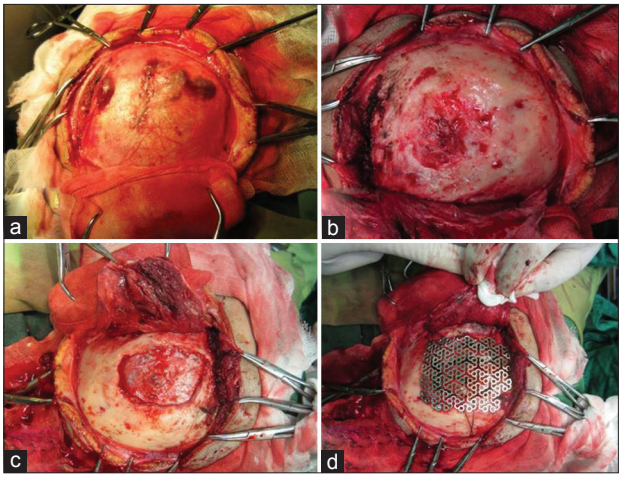

Figure 2: Exposure of the lesion at regio left parietal. (a) Bone discontinuity at regio left parietal, suggested as lytic lesion, as we confirm during operation (b), Postoperative; the bone defect, size 7 × 6 cm was closed with mesh (c-d)

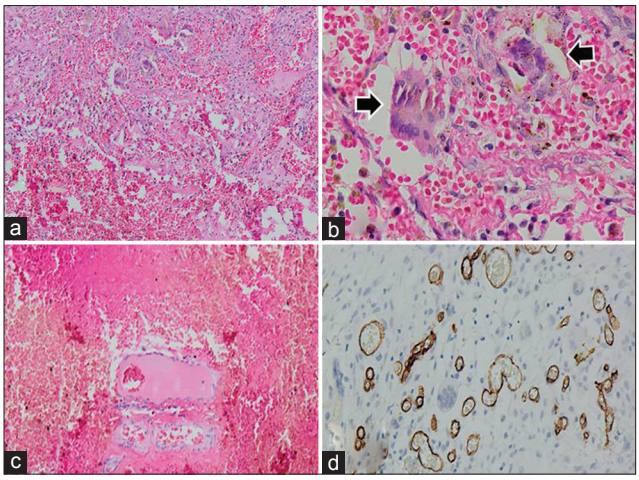

Figure 3: A giant cell angiofibroma of the scalp presenting as a varying combination of cellular areas composed of bland round to spindle cells, collagenous or myxoid stroma with focal sclerotic areas, medium-sized to small thick-walled vessels, and multinucleated giant cells, often lining angiectoid spaces; arrow giant cell (H and E, stain ×100 and ×200, respectively) (a-b). The lesion was observed eroded through the bone, as shown that the lysis of the bone without reformation (c). The lesion was highly vascularized with varying caliber of blood vessels and immunohistochemistry showed CD34 positive staining (d)

round- to spindle-shaped cells, collagenous or myxoid stroma with focal sclerotic areas, medium-sized to small thick-walled vessels, and multinucleated giant stromal cells often lining the angiectoid spaces.[2,14] The number of giant cells may vary from one tumor to another, and pseudovascular spaces may occasionally be absent. GCAs are invariably positive for immunohistochemical staining with vimentin and CD34.[2,8,10,12,13] Cytogenetic testing in one study revealed an associated chromosomal abnormality on 6q.[11]

Diagnostic imaging can play a role in determining tumor resectability, assessing surgical risks, and evaluating tumor recurrence.[1,13] CT scan and MRI may help to detect exact lesion localization and extension, but a diagnosis based on radiological evidence cannot usually be made. In our case, we identified a bone discontinuity at the left parietal region on a bone window CT scan, suggesting a lytic lesion, which was proven intraoperatively. Although the internal tabule of the skull was deformed, there was no invasion into the dura mater. This is unique, as these GCA entities are usually considered benign tumors. The lysis of the bone without reformation is typical of giant cell tumors and assumed to be secondary to osteoclast activity.[7]

Several entities should be considered in the differential diagnosis of GCA. On the basis of both histology and immunohistology, the following diagnoses can usually be easily excluded: hemangiopericytoma (a much rarer entity when strict diagnostic criteria are applied, which is usually more cellular and had a predominant staghorn vascular pattern and weaker CD34 positivity); fibrous histiocytoma (storiform pattern, more collagenous tissue and usually CD34 negative); schwannoma (Antoni A and B areas, Verocay bodies, strong S100 positivity); orbital meningioma (syncytial sheets or whorls of cells, EMA positive); and multinucleate cell angiohistiocytoma (usually dermal-based, CD34-negative, and CD68-positive interstitial cells but CD68-negative giant cells).[3,6] In our case, microscopic examination revealed cellular spindle cell proliferation between hyalinized blood vessels and scattered multinucleated floret-like giant cells. Mononuclear and multinucleated stromal cells were characteristically positive for CD34.

The best treatment appears to be surgical resection with complete tumor removal, as local recurrence may occur after incomplete resection.[12] Although GCA is a rare scalp tumor, it should be considered when the histopathologic examination reveals a well-defined fibroblastic pattern-less, CD34-positive subcutaneous tumor mass.

CONCLUSION

GCA is a benign tumor with a predilection for the orbit and head and neck regions in adults. Diagnostic imaging can play a role in determining tumor resectability, assessing surgical risks, and evaluating tumor recurrence. In our case, its imaging appearance was not specific and suggested a highly vascular soft tissue tumor. We performed surgical resection, and the clinical course was uneventful. GCA is uncommon but should be included in the differential diagnosis of a subcutaneous mass of the scalp.

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