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CASE REPORT

Giant Cell Angiofibroma Scalp: A Rare Neoplasm

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ABSTRACT

Giant cell angiofibroma (GCA) is a rare benign pathologic entity that shows a predilection for the mesenchymal tissue of the orbit and head and neck region. It is a highly vascular, well-circumscribed benign soft-tissue neoplasm that may simulate a malignant or aggressive process. GCA usually presents as a painful mass that grows slowly or remains stable in size over many years. The incidence of extraorbital GCA is very rare, with only two cases of GCA of scalp reported in the literature. The GCA is characterized by a patternless spindle cell proliferation containing pseudovascular spaces, floret-like multinucleated giant cells and a richly vascularized stroma showing hyalinization or myxoid change. CD34 immunoreactivity remains an important immunohistochemical finding of potential diagnostic value. Surgical excision is often curative. We report a case of a 42 years old male with the primary complaint of a painful solitary nodule arising on the occipital region of the scalp. Complete tumor removal through surgical resection was achieved, and diagnosis of Giant cell angiofibroma was rendered on the histopathological and immunohistochemical examination. The postoperative period was uneventful.

Keywords: Giant cell, Angiofibroma, Scalp.

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INTRODUCTION

Giant cell angiofibroma (GCA) was first described in 1995 by Dei Tos et al in a study of seven patients with a distinctive orbital tumor.[1] WHO defines GCA as a 'non-recurring, benign neoplasm containing multinucleated giant stromal cells and angiectoid spaces'.[2] It has been postulated that GCA belongs to the spectrum of solitary fibrous tumors,[2] with histologic features intermediate between those of solitary fibrous tumor (SFT) and giant cell fibroblastoma (GCF) of soft tissue.[1,3] Approximately, 40 cases of GCA involving different body areas, while only two cases of scalp GCA have been reported in the literature so far.[4] Although GCA of orbit shows a male predilection, however, the extraorbital lesions predominate in females, accounting for 66% of all the reported cases.[5] Diagnostic imaging is a helpful tool in determining resectability of the tumor, assessing its surgical risks, and in evaluation of the tumor recurrence. We are describing here a case of GCA arising in the scalp of a 42 years male in order to emphasize the extraorbital involvement of this rare benign neoplasm and to highlight its salient clinical, histopathological and immunohistochemical features that would differentiate this tumor from its close mimickers and will help in reaching to a conclusive diagnosis. To the best of our knowledge, this will be the third case of giant cell angiofibroma arising in the scalp.

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CASE REPORT

A 42 years male presented with a painful swelling arising on the occipital region of the scalp which was gradually increasing in size since 1 year. He had no history of trauma, systemic disease or any neurological deficit. On local examination, a soft, tender swelling was observed in the occipital region of scalp measuring approximately 5 × 4 cm. The overlying skin was loosely attached to the underlying soft tissue and appeared normal without any ulceration, discharging sinus or scarring. Neurological examination was unremarkable. Computed tomography (CT) scan of head revealed a 4 cm, noncalcified, uniformly enhancing well-defined soft-tissue mass in occipital region without any evidence of osseous erosion or invasion (Fig. 1). Fine needle aspiration was painful with

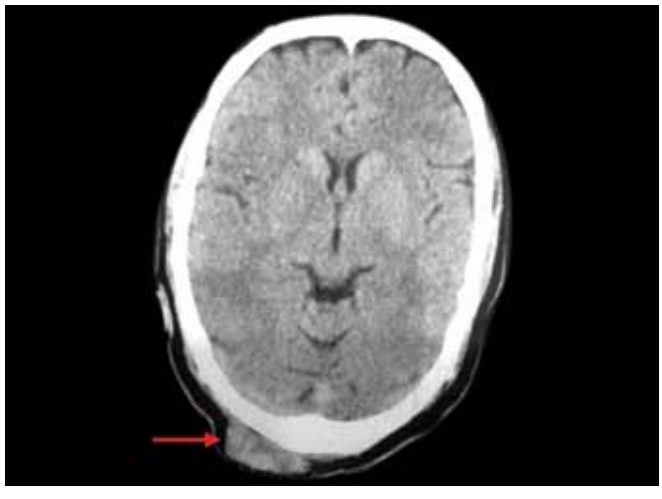

Fig. 1: Computed tomography scan of head showing a 4 cm, noncalcified, uniformly enhancing well-defined soft-tissue mass in occipital region (red arrow), evidence of osseous erosion or invasion not seen

bloody aspirate, cytology (FNAC) of the tumor revealed scant cellularity smear with few dispersed spindle cells with pink collagenous material in a hemorrhagic background, suggestive of some spindle cell neoplasm with high vascularity. The tumor was completely excised by accessing the tumor mass through a horseshoe-shaped scalp incision.

Gross examination of the resected specimen showed grayish-brown ill-defined mass measuring 3 × 3 × 1 cm which was soft in consistency. Cut section showed grayish-white vague nodularities with areas of hemorrhage (Fig. 2). Microscopic examination of the paraffin embedded hematoxylin and eosin (H&E) stained section revealed a well-circumscribed tumor, consisting of alternating hyper and hypocellular areas with oval to spindle-shaped cells arranged in a patternless manner surrounding angiectoid spaces in a hyalinized stroma (Fig. 3), intermixed with multinucleated giant cells with nuclei arranged in the center of the cells (Fig. 4), resembling floret giant cells and infrequent mitotic figures. Also, seen were blood vessels of varying caliber with perivascular hyalinization. Correlating the clinical, radiological and histopathological findings a diagnosis of rare giant cell angiofibroma was derived. Immunohistochemistry showed positive CD34 (Figs 5 and 6) and CD99 staining, while Bcl2, S100, desmin, smooth muscle actin (SMA), CD117, CD31, and CD68 were negative, which helped in reaching to a conclusive diagnosis of giant cell angiofibroma of scalp.

Postoperative period was uneventful, and the patient was discharged from the hospital, after a week stay with the follow-up advise.

DISCUSSION

Cutaneous and subcutaneous soft-tissue masses on scalp are commonly encountered in clinical practice and are usually benign, however, possibility of metastases from other sites should also be considered. Most common

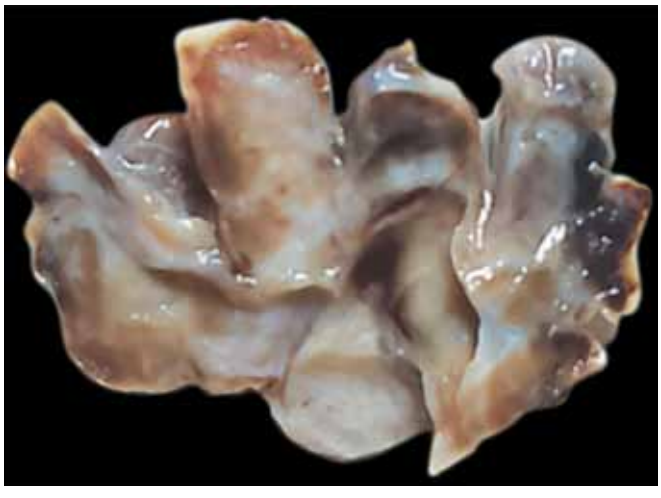

Fig. 2: Cut section showing grayish-white vague nodularities with areas of hemorrhage

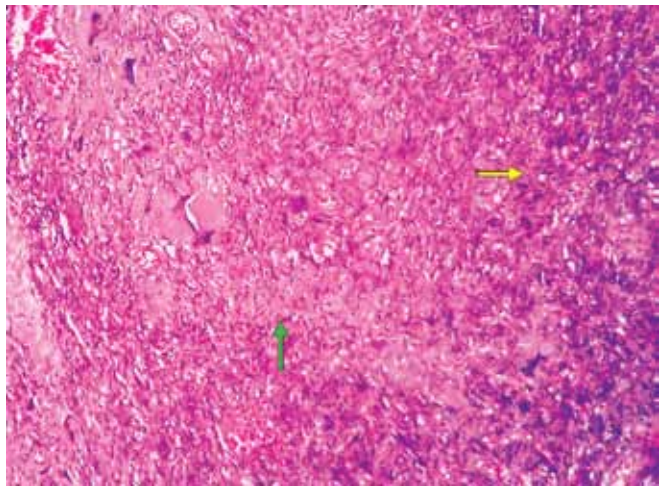

Fig. 3: Alternating hyper (yellow arrow) and hypocellular areas (green arrow) with oval to spindle-shaped cells arranged in a patternless manner, surrounding angiectoid spaces in a hyalinized stroma (H&E, 100×)

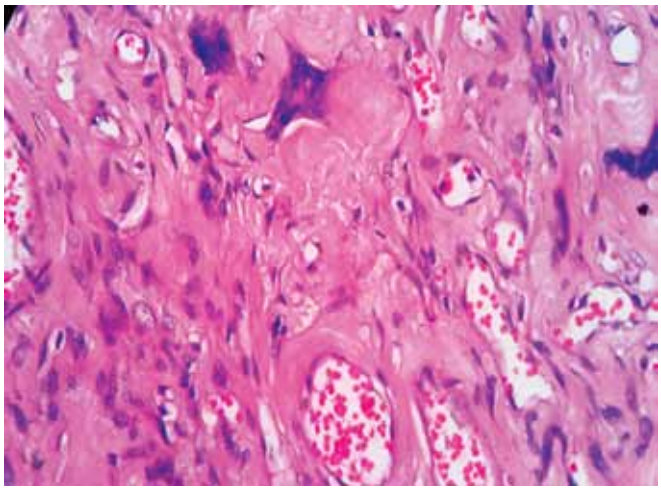

Fig. 4: Multinucleated giant cells with nuclei arranged in the center of the cells (H&E, 400×)

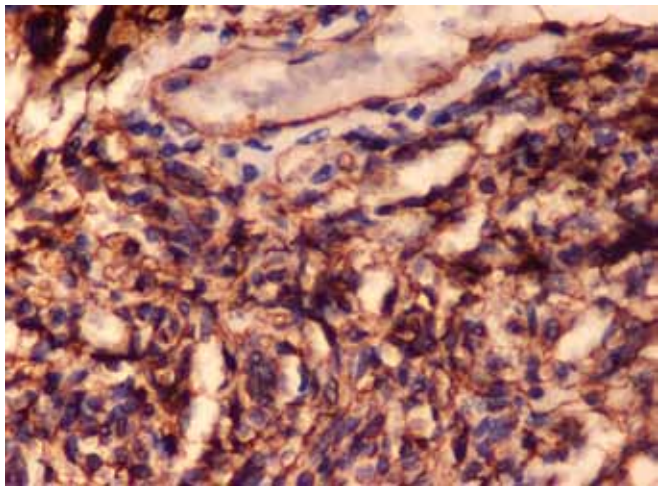

Fig. 5: Tumor cells showing CD34 immunoreactivity (IHC CD34, 400×)

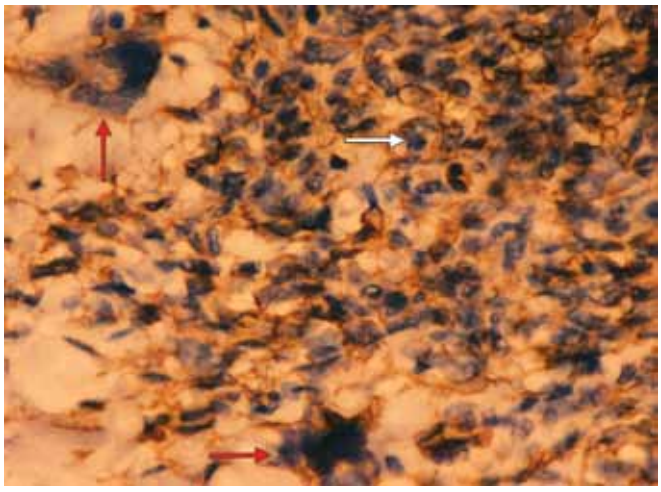

Fig. 6: Mononuclear (white arrow) and multinucleated giant cell (red arrows) showing CD34 positivity (IHC CD34, 400×)

clinical differential diagnosis of benign solitary scalp lesions include keratinous cyst (epidermoid and trichelemmal type), dermoid cyst, benign lipoma, spindle cell lipoma, hibernoma, eosinophilic granuloma, chondroid syringoma,[6] also included in the list are fibrohistiocytic lesions with giant cells which can be either predominantly histiocytic tumors or giant cell tumors. However, malignant lesions includes malignant pilar tumor, malignant nodular hidradenoma and liposarcoma[6] FNAC is a rapid, safe, cost-effective modality for determining the nature of a palpable scalp lesions and precludes invasive costly investigations.[7] The role of FNAC in the diagnosis of malignant scalp nodule was studied by Mondal et al,[8] who found majority of metastases from primaries in head and neck region, out of which 30% from thyroid carcinomas, and to lesser extent from abdominopelvic organs[7] Scalp metastasis usually seen within 3 months of occurrence of primary tumor.[9]

A giant cell rich form of hemangiopericytomas-solitary fibrous tumors (HPC-SFT) was described in 1995 by Dei Tos et al in a study of seven patient with a distinctive orbital neoplasm as 'giant cell angiofibroma' (GCA).[1] Giant cell angiofibroma displays all features of a classic solitary fibrous tumor (SFT) but it is identified by pseudovascular spaces lined by multinucleated stromal giant cells. The age of patients with giant cell angiofibroma ranges from 20 to 80 with mean age of 45 years.[10] Giant cell angiofibroma has a much wider anatomic distribution, it usually occurs in orbital region, including the eyelids, the nasolacrimal duct and the lacrimal sac region but to a lesser extent it is also seen involving the head and neck region outside the orbit as in scalp, retroauricular region, parotid gland, cheek, submandibular region, buccal mucosa, as well as in the posterior mediastinum, back, axillary and inguinal regions, retroperitoneum and vulva.[5,11,12] Orbital giant cell angiofibroma predominates in males,[4] while extraorbital lesions predominate in female patients.[5] GCA usually presents clinically as a slowly growing mass which sometimes can be painful.[13] CT scan and MRI help to detect the exact localization of the lesion, assessment of its extent and resectability. Grossly, giant cell angiofibromas are well circumscribed, they may be variably encapsulated, they are often a small lesion approximately 3 cm average size, however, soft-tissue lesions tend to be larger than those in orbital-region, sometimes reaching up to 10 cm.[13] Upon cut section of these tumor, hemorrhagic and/or cystic changes may be observed.[14] Microscopically, these tumors are well circumscribed, patternless lesions composed of bland looking round to spindle-shaped cells with pale, indistinct cytoplasm set in a variably collagenous or myxoid matrix containing numerous small blood vessels (often with hyalinized vessel walls), irregularly-shaped pseudovascular spaces lined by mononuclear and multinucleate tumor cells; multinucleate giant cells are often floret type, present even in the solidly cellular areas. Tumor cells show consistent positive immunoreactivity for CD34 and CD99 and less frequently, for BCL2.[15]

Giant cell angiofibroma must be differentiated from other fibroblastic soft-tissue tumors having overlapping spectrum of morphologic and immunophenotypic findings, such as hemangiopericytoma (HPC) (a much rarer entity when strict diagnostic criteria are applied, which is usually more cellular and shows a predominant staghorn vascular pattern and a weaker CD34 positivity), solitary fibrous tumor (SFT) (shows a storiform pattern of cellular arrangement, tumor cells are vimentin, CD34, and bcl-2 positive[16] complicating its distinction from GCA, however, pseudovascular spaces and lining by multinucleated giant cells are not seen in SFT) and giant cell fibroblastoma (GCF) (most often presents in early childhood as a slow growing infiltrative soft tissue mass, at a wide variety of sites and is characterized by a high rate of local recurrence, while no case of GCA has been reported in children till now and also it is a well circumscribed with exceptional instances of local recurrences).

Surgical excision is the treatment of choice for lesions amenable to resection. Nearly, all giant cell angiofibromas shows a benign behavior; local recurrences after complete excision is exceptionally reported at only two instances.[1,14,15]

CONCLUSION

1. Giant cell angiofibroma (GCA) is a highly vascular, benign, non-recurring soft-tissue neoplasm having predilection for the orbit and head and neck region.
2. Although GCA is an uncommon tumor, however, it should be considered in the differential diagnosis of subcutaneous mass of the scalp.

3. Diagnosis of GCA is rendered when the histopathologic examination reveals a well-defined, patternless, fibroblastic subcutaneous tumor mass showing CD34 positivity.

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