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Case Report

Rapidly growing diffuse neurofibroma of the scalp with calvarial defect in a young woman: A rare entity

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ABSTRACT

Background: A diffuse neurofibroma, a variant of neurofibroma, most commonly occurs in young adults and involves the head and neck. In the absence of neurofibromatosis, associated calvarial defect with these swellings is rarely seen.

Case Description: An 18-year-old woman presented with a history of rapidly progressive painless large swelling over the bilateral parieto-occipital region of scalp. It was soft and boggy with brownish discoloration of overlying skin. Imaging study showed brilliantly enhancing diffuse lesion involving the bilateral parieto-occipital region of scalp and extending into the extradural region. She underwent excision of lesion. Postoperatively, she developed flap necrosis and it was allowed to heal with the secondary intention. The biopsy findings were consistent with neurofibroma. The patient is on regular follow-up, without any evidence of recurrence at 1 year.

Conclusion: Rapidly growing solitary diffuse neurofibroma is rare in children and adolescents. Preoperative diagnosis may be difficult and surgical treatment needs to be individualized. These patients need regular follow-up for early detection of recurrence.

Keywords: Diffuse neurofibroma of the scalp, Neurofibroma, calvarial defect, Reconstructive surgery, Surgical excision

INTRODUCTION

A diffuse neurofibroma, a variant of neurofibroma, most commonly occurs in young adults and involves the head and neck. Associated calvarial defect with these swellings is also rarely seen in the absence of neurofibromatosis.

We report a young woman with rapidly progressive diffuse neurofibroma of the scalp with a small calvarial defect, without any features of neurofibromatosis.

CASE REPORT

An 18-year-old woman presented with a history of painless rapidly progressive large swelling over bilateral parieto-occipital region of scalp for the past 6 months. She underwent surgery for the swelling at occipital region 5 years back, but reports of the previous treatment were not available with the patient. Local examination showed soft boggy swelling over the scalp with

brownish discoloration of the overlying skin. Healed linear scar was noted near the center of the swelling [Figure 1]. Neurocutaneous markers were not present and. No significant family history suggestive of neurofibromatosis was present.

Computed tomography scan of the head revealed a diffuse soft-tissue mass in bilateral parieto-occipital region with the left occipital bone defect near the asterion [Figures 2a and b]. Magnetic resonance imaging revealed brilliantly enhancing diffuse lesion involving extracranial bilateral parieto-occipital region and small extradural component with infiltration of the scalp [Figures 2c and d]. Fine-needle aspiration cytology of the swelling was inconclusive and showed hemorrhagic fluid only.

The patient was planned for the excision of lesion with reconstructive surgery. Due to the possibility of loss of a large area of the scalp, only surgical excision of the mass was done preserving the overlying scalp. The lesion was soft, fleshy, moderately vascular, infiltrating scalp at places but was free from pericranium. During surgery, hair follicles were also exposed at some places. Small extradural part was also present near the asterion with bony defect. The lesion was excised, with small part of infiltrating scalp was also removed, preserving the pericranium. Calvarial defect is left as such and as calvarial defect was small; no cranioplasty was performed during surgery. Scalp was closed primarily without any difficulty.

Postoperatively, the patient has developed skin flap necrosis and pus discharge from the center of the skin flap. The wound was allowed to heal by the secondary intention under appropriate antibiotics after the debridement of necrotic tissue.

The histopathology report was consistent with neurofibroma [Figure 3]. The patient is on regular follow-up since then, without any evidence of recurrence at 1 year on repeat imaging.

DISCUSSION

Diffuse neurofibroma is a rare, but distinctive variant of neurofibroma. Diffuse neurofibroma is most commonly seen in the head-and-neck regions of adolescents or young adults.[10] Occurrence of giant solitary neurofibroma of the scalp without neurofibromatosis is rare. In the present case, neurocutaneous markers such as café au lait spots and axillary or inguinal freckling were absent with normal orthopedic and ophthalmological findings in the examination. No family history suggestive of neurofibromatosis 1 was present.

Most reported cases in the literature are in adults age group, with a gradually progressive tumor growth over decades.[3] This woman had noticed a rapid increase in the size of the lesion for the past 6 months. A rapid increase in the size of lesions may be seen in cases with malignant transformation of a lesion or intratumoral hemorrhage.[9] However, the histopathological examination of the lesion had not revealed any features suggestive malignant degeneration or intratumoral bleed.

Diffuse neurofibroma is composed of elongated, spindle-shaped cells with round or fusiform nuclei and eosinophilic cytoplasm within a loose matrix of fine fibrillar collagen. The lesion diffusely infiltrates the dermis and subcutaneous tissue. The tumor encircles the surrounding structures rather than destroying them.[10] On immunohistochemistry, tissue shows S-100 protein positivity.

A calvarial defect is usually not associated with neurofibroma in the absence of syndromic association. Skull defect having sclerotic margins is suggestive of an underlying neurogenic tumor.[1] In this patient, small cranial defect was seen on radiology. Intraoperatively, we found that the cranial defect margins were smooth and were not involved. Hence, bone around the defect was not drilled. There are only few reports of calvarial defects in association with neurofibromatosis in children and young adults in the English literature [Table 1].

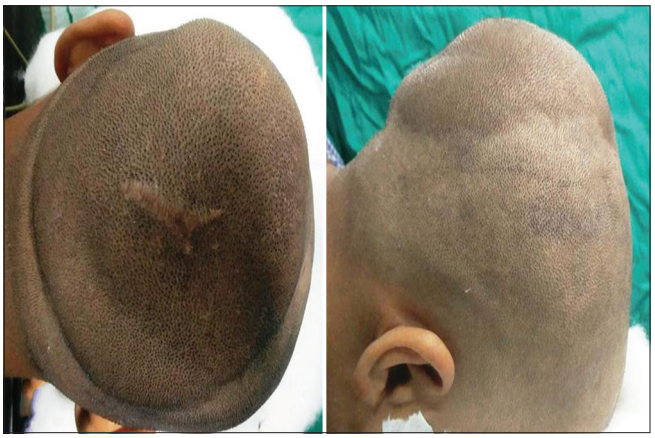

Figure 1: Clinical photograph displaying scalp swelling with overlying brownish skin discoloration.

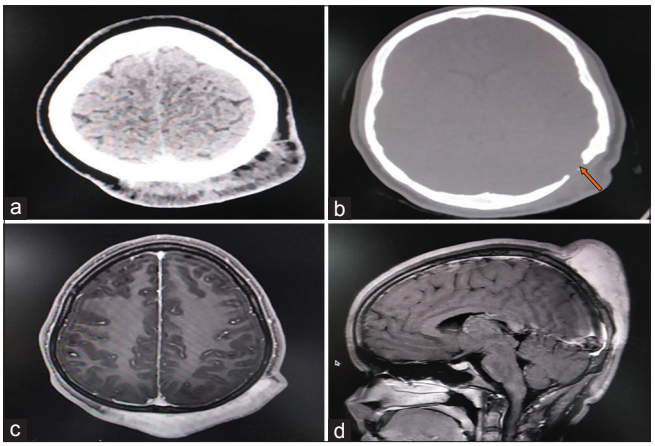

Figure 2: Computed tomography brain axial showing (a) diffuse soft-tissue mass in scalp (b) associated calvarial defect (marked with orange arrow head). (c and d) Magnetic resonance imaging showing diffusely enhancing soft-tissue scalp mass.

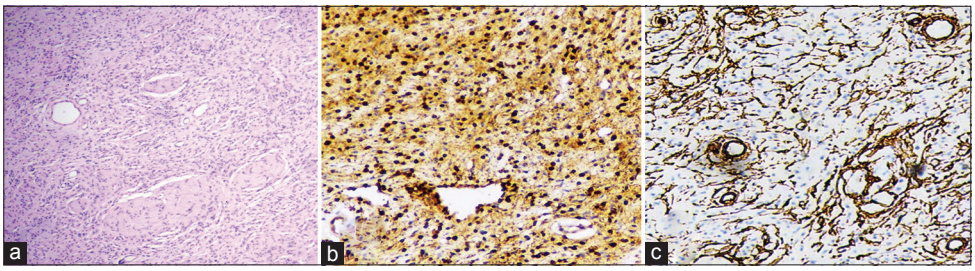

Figure 3: Photomicrograph illustrating histopathology of the lesion (a) shows irregular shaped groups of bland spindle cells with wavy nuclei and pale eosinophilic cytoplasm. There is no nuclear atypia. Mitotic activity is not appreciated (H&E, ×20), (b) diffuse positivity for S-100; (c) CD34 positivity in blood vessels only (IHC, ×200).

Table 1: Diffuse neurofibroma patients with calvarial defect in children and young adult.

S. No	Title	Clinical features	Treatment	Follow-up
1.	Solitary giant neurofibroma of the scalp with calvarial defect in a child[1]	5 years, boy 7 weeks' h/o a progressive painless swelling on the right side of the forehead and temporal region	Tumor excision and cranioplasty	8 weeks no residual lesion
2.	Solitary giant diffuse neurofibroma of the scalp with calvarial defect[3]	15, girl painless and progressive swelling over the left temporal region since the age of 8 years	Surgical excision	1 year, no recurrence
3	Rapid growing diffuse neurofibroma of scalp with calvarial defect: a rare entity and review of literature	18 years, 6 months history of rapidly growing swelling	Surgical excision	1 year no recurrence

The treatment of large neurofibroma consists of surgical excision with or without reconstructive surgery. As these lesions are highly vascular, obliteration of feeding vessels in early stages of surgery helps to minimize intraoperative bleeding. We were able to control bleeding from the lesion by the bipolar coagulation. Lin and Chen highlight the importance of hypotensive anesthesia and preliminary sutures around the lesion to control bleeding.[6] Some surgeons prefers preoperative angiogram[7] and intra-arterial embolization to reduce the risk of hemorrhage. However, in spite of these measures to reduce vascularity, sometimes massive bleeding can occur and in such cases serial excision is one option.[4]

Reconstruction of the defect can be done with expanded myocutaneous flaps and tissue expander[8] or split-thickness skin graft.[2] The anterolateral thigh flap, the latissimus dorsi muscle flap, the myocutaneous rectus abdominis flap, and the radial forearm flap are most preferred free pedicled flaps for scalp and forehead reconstruction.[5] However, it is highly challenging procedure and need expertise. Tissue expansion is a relatively simple procedure and gives essentially matched skin in terms of color, texture, hair-bearing qualities, and sensation.

Rapidly growing solitary diffuse neurofibroma is rare in children and adolescents**.** Preoperative diagnosis may be difficult and surgical treatment needs to be individualized. These patients need regular follow-up as lesion may recur even after complete excision due to the infiltrative nature of the tumor.

CONCLUSION

Rapidly growing solitary diffuse neurofibroma is rare in children and adolescents and has less propensity to involve calvarial bones. These cases need long term follow up after surgical excision for early detection of recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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