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CASE REPORT

Solitary giant diffuse neurofibroma of the scalp with calvarial defect

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ABSTRACT

Neurofibromas of the scalp can have protean presentations. Most of the swellings are small, solitary and are easily diagnosed clinically. Diffuse swellings on the other hand are rare and are commonly seen in adults. The skull defects with these swellings are also rarely reported in the absence of neurofibromatosis. There is only one report of child having diffuse neurofibroma with skull defect. We report a second case in literature in a child with progressive, painful, diffuse neurofibroma along with calvarial defect.

Key words: Calvarial defect, neurofibroma, neurofibromatosis, scalp neurofibroma

Introduction

Neurofibromas are tumors of nerve sheath origin and may arise from Schwann cells, perineural cells,[1] or fibroblasts.[2] They are usually solitary, but upto 10% of patients have multiple lesions and a proportion of them have von Recklinghausen neurofibromatosis.[3] Diffuse neurofibroma is an uncommon but distinctive form of neurofibroma, which canpresent invariablesizes, often as large areas of marked dermal and subcutaneous thickening. Most of them are located in the trunk or head and neck region of adults. Theselesions have also been termed 'paraneurofibroma' to indicate the extension of the tumor beyond the confines of the perineurium.[4] Intracranial extension of extracranial variety of this tumor has also been reported in adultsbut most have been in patients with neurofibromatosis or pre-existing plexiform neurofibromas.[5] Scalp neurofibroma with calvarial defect in the absence of neurofibromatosis or phakomatosis is very rare. We report a case of a 15-year-old girl with anuncommon solitary giant diffuse neurofibroma of the scalp with defect in the skull bone.

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Case Report

Anadolescent girl aged 15 years presented with painless and progressive swelling over the left temporal region since the age of eight years. Five years ago, that is, in 2006, she had developed headache and vomiting and was diagnosed with post-tubercular meningitis hydrocephalus for which ventriculoperitoneal (VP) shunt was done. On examination, there was a soft-to-firm, boggy swelling in the left fronto-temporo-occipital region, which was nontender, noncompressible, nonpulsatile, nonreducible, and nontransilluminant with normal skin overlying the swelling. It had variable consistency, measuring about 15 × 12 cms, andthe underlying bony margins could not be appreciated. There was no evidence of neurofibromatosis or phakomatosis. Clinically, tubercular osteitis with osteomyelitis, soft tissue tumor, and vascular malformations were considered in differential diagnosis. Angiography was done and it revealed no vascular abnormality. Fine needle aspiration cytology (FNAC) of the swelling was inconclusive. X-ray of the skull revealed soft tissue density in left temporo-occipital region, defect in the left occipital bone, and burr hole of the VP shunt in right parietal bone [Figure 1]. Computed tomography (CT) scan of the head revealed a calvarial soft tissue mass in left temporo-occipital region with left occipital bone defect near the asterion. It was enhancing on contrast scan. There were calcified focal densities

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in the right basal temporal lobe, right cerebellum, right tectal plate, and near left frontal horn [Figure 2]. Magnetic resonance imaging (MRI) of the brain revealed soft tissue mass in the same region, which was iso-hypointense on T1, iso-hyperintense on T2 with multiple flowvoids, and heterogeneously enhancing with contrast. The VP shunt was noted coursing through the right parietal lobe with its tip in the frontal horn of the left lateral ventricle [Figure 3]. The patient underwent surgical excision of the mass. The tumor tissue was soft, fleshy, moderately vascular, and mostly encapsulated with certain areas of ill-defined margins. It was eroding the left occipital bone near the asterion but was lying extradurally. The mass was excised along with pericranium [Figure 4]. No cranioplasty was performed at this stage. The histopathology report was consistent with benign neurofibroma [Figures 5a and b]. The postoperative period was uneventful. The patient was followed up with regular radiological studies without any evidence of recurrence in one year.

Discussion

Giant solitary neurofibroma of the scalp without neurofibromatosis is rare. Only a few cases have been reported so far. Most reported cases are adults, with a slow rate of tumor growth over decades.[6,7] Only one case of a five-year-old child has been reported.[8] Most reported cases of giant scalp neurofibroma are in Japanese literature.[6,9,10] Because of lack of distinctive clinical characteristics, especially in isolated lesions, it is difficult to diagnose the lesion preoperatively, and such lesions remain largely, a histopathological surprise. Even proper history taking, physical examination, ophthalmological, and radiodiagnostic investigations may fail in clinching the diagnosis in the absence of neurofibromatosis. The presence of skull defect with sclerotic margins is suggestive of an underlying neurogenic tumor. Bone defects are more common in neurofibromas associated with neurofibromatosis.[6,9,10] In our case however, the bone

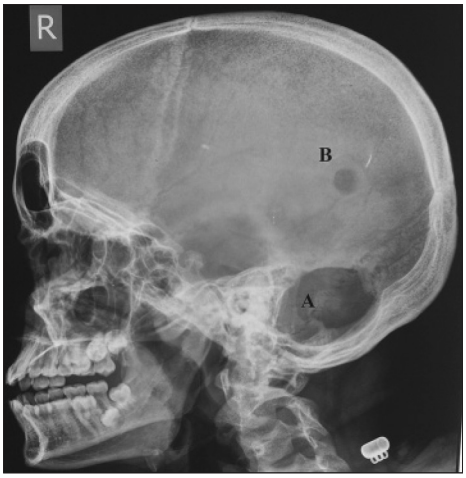

Figure 1: (a) Plain X-ray showing lytic defect in left occipital bone (b) with burrhole at parietal bone

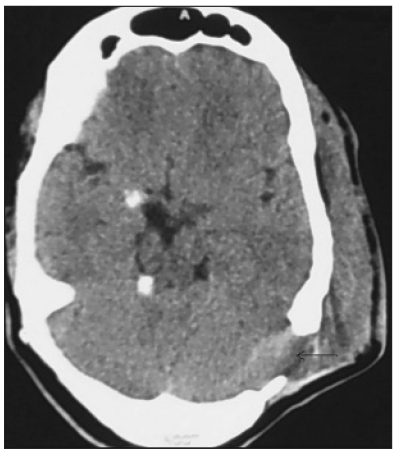

Figure 2: Non contrast CT head showing defect in the occipital bone (thin arrow) with soft tissue swelling. There is evidence of calcification in right cerebellum and medial perisylvian area

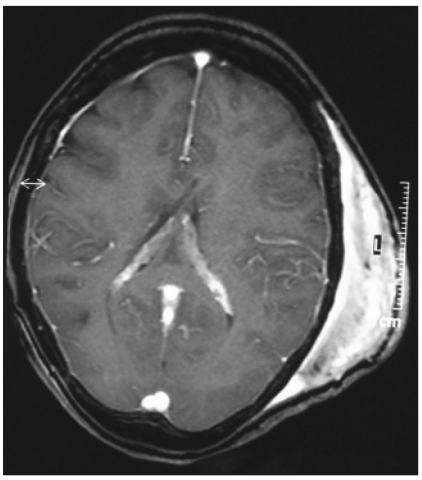

Figure 3: MRI contrast showing enhancing soft tissue swelling with shunt tube in place

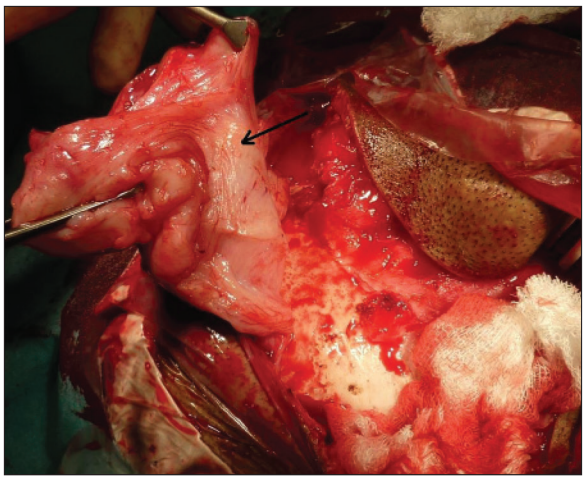

Figure 4: Intraop photograph shows diffuse mass lifted off the skull (arrow)

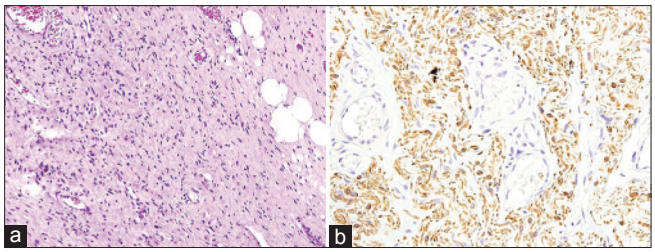

Figure 5: (a) Hemotoxiline and Eosine slide showing tumor composed of Schwann cells displaying thin wavy nuclei. (b) Tumor showing focal but strong positive areas for S100

defect was not associated with either sclerotic margins or any stigmata of neurofibromatosis.

There is no consensus about the role of cranioplasty in such cases. There is a potential risk of recurrence; therefore, some authors suggest deferring the cranioplasty fearing recurrence.[8,11] The literature is rather silent about the timing of cranioplasty in such cases. As the child was in a growing stage, and there was a risk of recurrence, the cranioplasty was deferred in our case for the time being.

Solitary giant neurofibroma of the scalp with calvarial defect is rare. Clinical diagnosis may be difficult, and surgical treatment must be individualized. Regular radiological studies during follow-up visits are necessary to detect early recurrence.

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Conflicts of interest

There are no conflicts of interest.

References

1. Claus EB, Persing JA. Scalp tumors. Youman's neurological surgery. Philadelphia: Saunders; 2004. p. 1409-18.
2. Kenneth WL, Bone I. Intracranial tumors-pathological classification. Neurology and neurosurgery illustrated. Edinburgh: Churchill Livingston; 2004. p. 301.
3. Bodhey NK, Gupta AK. Neurofibromatosis type I with occipital encephalocele. Neurol India 2006;54:103-4.
4. Chowdhary RP, Little BW. Large vascular plexiform neurofibroma of scalp: Excision and coverage with free tissure transfer. Ann Plast Surg 1990;24:75-9.
5. Ergün SS, Emel E, Karabekir S, Buyukbabani N. Extracranial diffuse neurofibroma with intracranial extension. Plast Reconstr Surg 2000;105:801-3.
6. Aya T, Mitsuo H, Tadashi O, Hideyuki T, Masamitsu K, Aiko S. Giant neurofibroma of the scalp: Case report. Jpn J Plast Reconstr Surg 1999;42:413-7.
7. Yoo KH, Kim BJ, Rho YK, Lee JW, Kim YJ, Kim MN, *et al.* A case of diffuse neurofibroma of the scalp. Ann Dermatol 2009;21:46-8.
8. Ismail NJ, Shehu BB, Lasseini A, Hassan I, Shilong DJ, Obande JO, *et al.* Solitary giant neurofibroma of scalp with calvarial defect. J Surg Tech Case Rep 2010;2:24-6.
9. Tsutsumi M, Kazekawa K, Tanaka A, Ueno Y, Nomata Y, Nii K, *et al.* Rapid expansion of benign scalp neurofibroma caused by massive intratumoral haemorrhage--Case report. Neruol Med Chir (Tokyo) 2002;42:338-40.
10. Nakasu S, Nakasu Y, Natsuda I, Handa J. Giant neurofibroma of the occipital scalp associated with lambda defect. No To Shinkei 1981;33:181-5.
11. Mislow JM, Proctor MR, McNeely PD, Greene AK, Rogers GF. Calvarial defects associated with neurofibromatosis type I: Report of two cases. J Neurosurg 2007;106:484-9.