Case Report

Giant Neurofibroma of the Scalp

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Abstract

We are reporting a rare case of big scalp neurofibroma in a patient not known to have Neurofibromatosis type 1 (NF1) or Neurofibromatosis type2 (NF2) aged 24yr. The mass was present since birth with progressive enlargement ,total surgical resection done with Z-plasty to close the scalp defect.

Key words: neurofibroma, scalp mass, progressive enlargement.

Introduction

Neurofibroma is a well Known benign tumor of the peripheral nerve sheath[5], two histologically and perhaps biologically, distinct lesions have been termed neurofibromas, the most common form occurs in the skin (cutaneous neurofibroma) or in peripheral nerve (solitary neurofibroma), these arise sporadically or in association with neurofibromatosis type 1. The second type is the plexiform neurofibroma wich is considered by some to occur only in patient neurofibromatosis type 1 [4].

We reported a big scalp not plexiform neurfibroma in patient not known to have neurofibromatosis type 1 and the mass was present since birth and it got progressive enlargement over years.

Case Report

Twenty four year old married lady not known as Neurofibromatosis type 1 or Neurofibromatosis type 2, presented with progressively enlarging big scalp mass at left parieto teporal area of scalp, the mass was present since birth but it was small then progressively enlarging mainly during pregnancy period then 2 months after delivery she advised medical care.

Physical examination showed a large scalp mass measuring length 14 cm, width = 11cm, height = 10 cm, on left parietoteporal area the mass was soft, non pulsating, well defined margin, non tender, at the tip of the mass there is hair fall with just like paede orange appearance of the skin and the skin was hyperpigmented (figure 1).

All investigation were within normal. Brain CT scan showed hypo and hyper dense lesion, no skull defect, no hematoma (figure 2)

Total surgical resection was done. The mass was very vascular with tortous scalp vessels, the periosteum was edematous but not involved, and no intratumoral hematoma. The scalp was involved Z- plasty used to close the defect. Histopathological study showed closely packed spindles cells proliferation with serpentine orientation and scattered chronic inflammatory cells with mast cells (figure 3).

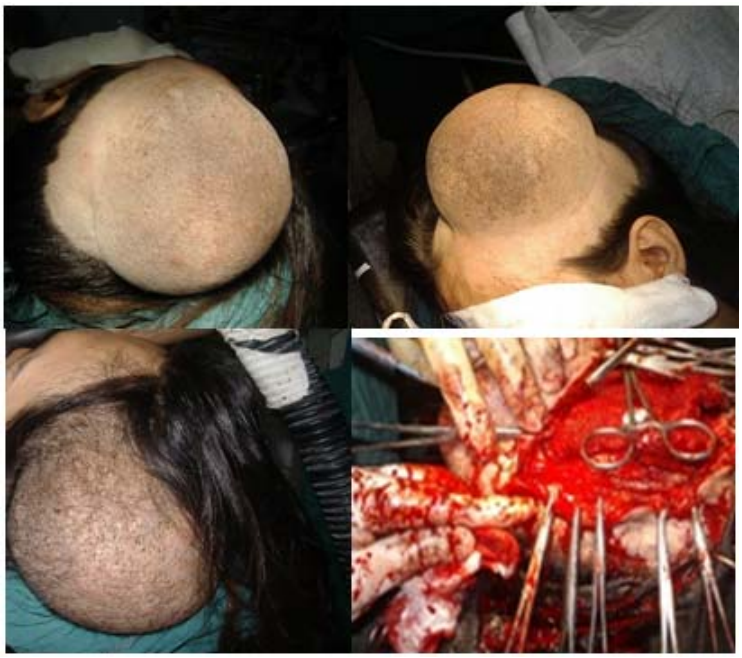

Fig 1. scalp mass.

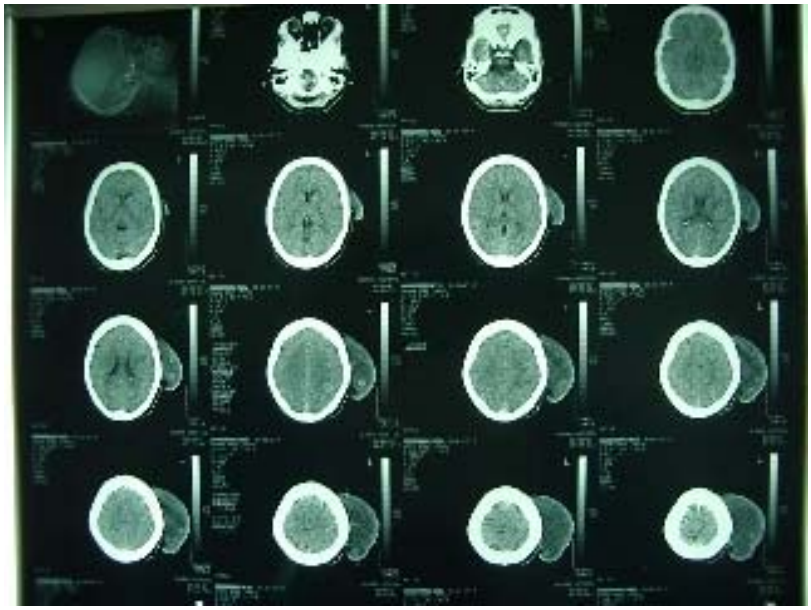

Fig 2. CT scan of the brain & skull

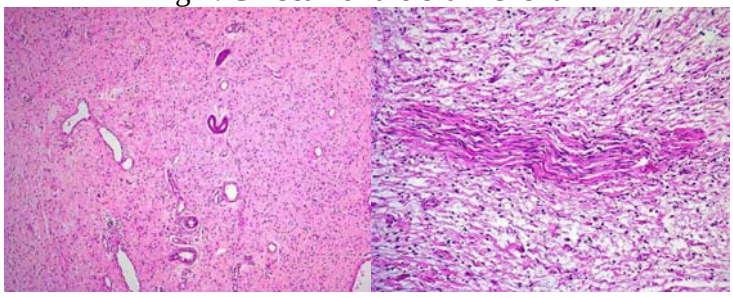

Fig 3. Histopathological study

Discussion

Scalp Neurofibroma is relatively rare. Although, association with evidence of NF1 is strongly indicative of diagnosis[2], but in our case, the patient was not known to have NF1.

Scalp nearofibroma is characterized by very slow progression, occipital coction, associated with skull defect or thinning and salient hyper vascularity[7,8], while our patient had mainly parietotemperal mass with no skull defect or even thinning.

Four reported cases of scalp neurofibroma presented with spontaneous intratumoral hemorrhage[3,7] while we did not find such hemorrhage but we reported significant enlargement of the mass during pregnancy period which is mostly due to increasing blood volume which occurs as physiological changes during pregnancy.

To our knowledge there are only four reported cases of neurofibroma of the scalp without other manifestations of Neurofibromatosis[6] and this case to be the fifth one, noting that most of the reported cases are in eastern countries.

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