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Beret Shaped Diffuse Neurofibroma: A Case Report

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Abstract
Diffuse neurofibromas are rare benign tumors, mostly located in the cephalic extremity, and are associated with neurofibromatosis type 1 in 10% of cases. They are known in the literature for their clinical appearance, haemorrhagic surgery and prognosis.
In this article, we report the case of a patient with beret-shaped diffuse neurofibroma of the scalp occurring in the context of Von Recklinghausen's neurofibromatosis.
Keywords: *Diffuse Neurofibroma, neurofibromatosis type 1, scalp*

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I. Introduction

Neurofibromas are benign lesions of the peripheral nerves, they may occur in syndromic or sporadic forms.

We identify multiple types in the litterature: classic, myxoid, cellular, pigmented, pacinian, epithelioid, hyalinised,, plexiform, diffuse, dendritic cell neurofibroma, granular cell.(1)

Diffuse neurofibroma is a rare tumor that occurs primarily in children or young adults, and typically appears in the cephalic and neck region. (2,3). It is a benign tumor of the peripheral nerve that develops from proliferative schwann cells, perineural cells and endoneurial fibroblasts. Diffuse neurofibroma has a neuroectodermal origin(4)

We present a case of a 35-year-old man who had a giant diffuse neurofibroma on the scalp since the age of 15 years.

II. Case Report :

A 35-year-old patient with no family history, presented with a giant pedunculated scalp mass with a 20-years evolution.

On physical examination,he had a left fronto-parietal mass,beret shaped, measuring 21x15cm, firm in consistency, slightly fixed in relation to the deep plane, non-pulsatile and veiling the left orbit. The patient used to always wear a hat to hide it (Figure 1).

In addition there were diffuse cafe-au-lait skin spots all over his body, as well as exophytic tumors(Figure 2). The neurological examination was normal. Blood tests showed increased hemoglobin to 17g.

The CT-scan (Figure 3) reveals a large extracranial mass containing calcifications and fatty islands, with intra-tumoral veins draining into the superficial venous network of the face and the scalp. In addition, a discrete irregularity of the left parietal external table without erosion of the Diploe and no intracranial extension.

Under general anesthesia, in supine position, after infiltration with epinephrine-lidocaine, transfixing haemostatic stitches were made at the base of the tumor using 2/0 silk. Cutaneous incision, followed by monobloc excision using an electrocautery scalpel, and haemostasis. Then a direct suture with 2/0 silk at scalp and 4/0 silk in the frontal region. (Figure 4) The patient was transfused with two units of blood.

The aesthetic and functional results were very satisfactory (Fig 5) with a 3-year of follow-up, there was no recurrence.

The histological study reveals a diffuse neurofibroma in the context of type 1 neurofibromatosis (Von Recklinghausen's Disease)

Genetic analysis was not performed due to lack of resources.

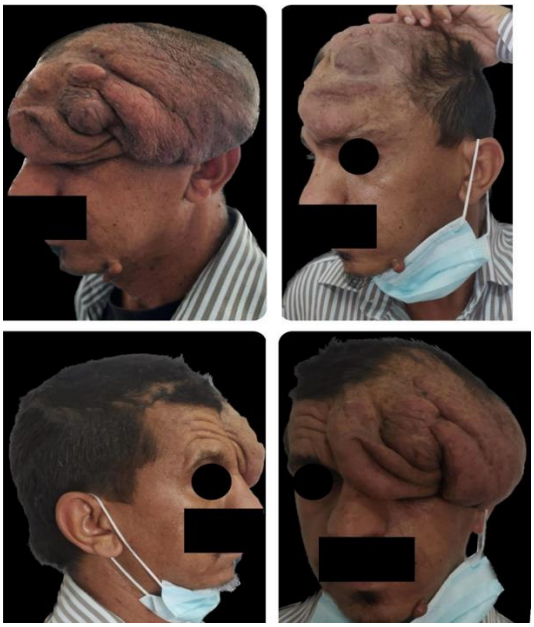

Fig 1 : Preoperative Aspect Of The Neurofibroma And

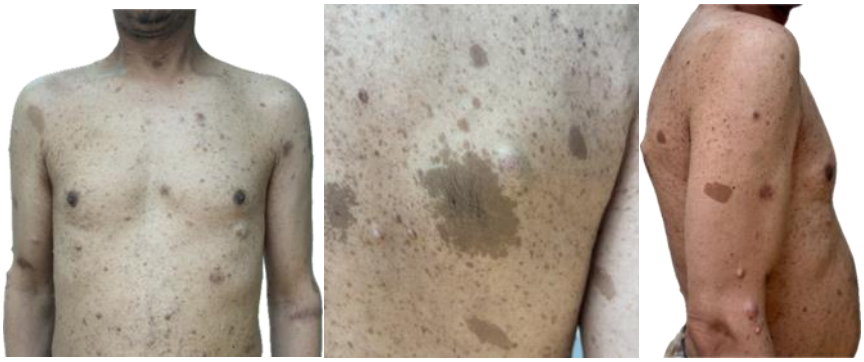

Fig 2 : Café Au Lait Skin Lesions

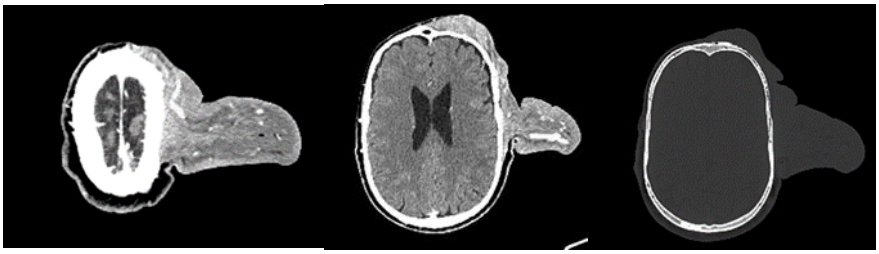

Fig 3: CT Scan Showing A Large Extracranial Mass With Intratumoral Veins

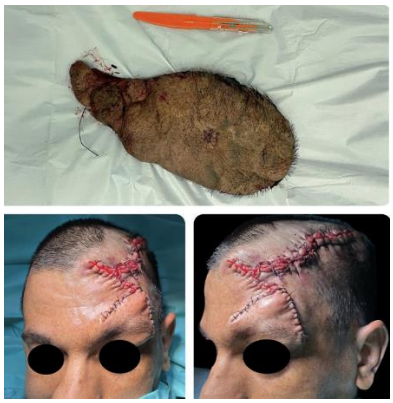

Figure 4: Immediate Postoperative Results

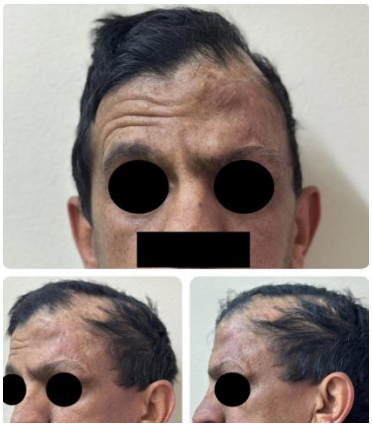

Fig 5: Late Postoperative Outcomes

III. Discussion

We described a case of beret-shaped diffuse neurofibroma of the scalp occurring in Von Recklinghausen's Disease background. A Similar case was described in the literature with a beret shaped tumor (3) It is associated with neurofibromatosis type 1 (Von Recklinghausen's Disease) in 10% of cases, and occurring in 90% of cases in the absence of neurofibromatosis. (2,4,5)

Intracranial extension of extracranial neurofibroma is possible especially in von Recklinghausen Neurofibromatosis patients(5)

Diffuse neurofibroma should be evoked in the case of scalp swelling (2)

The fatty component on imaging is highly suggestive of the diagnosis, but it may suggest other differential diagnoses of fatty tumors, which is found in the literature .(4)

This is a highly vascularized tumor whose surgery is highly hemorrhagic. In fact, the literature recommends operating under hypotensive conditions, using transfixing sutures around the tumor, as we did in this reported case, and ligating the feeding pedicles. (2,6), Kareem et al described a haemostatic technique utilizing multiple overlapping interrupted sutures before resection.(7) Even though some authors in the literature do not transfuse their patients, we recommend intraoperative transfusion to compensate for the massive hemorrhage that can occur during resection.

Treatment is based on total or partial excision, as the infiltrative features of this lesion make it at risk of recurrence, especially in cases involving patients with neurofibromatosis.

In this case we performed direct suture with local plasty for the reconstruction, in the literature, forehead and scalp reconstruction can be done using tissue expander, split-thickness skin graft, flaps such as latissimus dorsi, anterolateral thigh flap, rectus abdominis flap, and radial forearm flap.(8)

Tissue expanders are a good option for scalp reconstruction, with their advantages of providing sensitive, aesthetic, and hairy skin. (8)

Considering the risk of cancerization, we recommend performing complete exeresis if possible.

IV. Conclusion

To conclude, we report the rare case of a patient presenting beret-shaped diffuse neurofibroma of the scalp in the background of neurofibromatosis type 1. This is a rare tumor that is associated with Von Recklinghausen's Disease in 10% of cases. Surgery is performed not only for aesthetic purposes, but also to prevent malignant transformation.

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