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Surgical Management of Diffuse Plexiform Neurofibroma in Von's Disease Recklinghausen: About a Case at Mopti Hospital

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Abstract **Case Report**

***Introduction*:** Facial plexiform neurofibromas are considered a rare but disfiguring and devastating complication of facial neurofibromatosis. The objective of this work is to show the difficulties of the surgical management of the facial manifestations of neurofibromatosis. ***Observation*:** A 39-year-old woman consulted for a monstrous-looking swelling with an irregular texture on the left hemiface, evolving for 13 years. She underwent external carotid artery ligation, total tumor excision and thin skin grafting. Histology concluded to a facial plexiform neurofibroma. ***Discussion*:** Neurofibroma is a benign tumor arising from the connective elements of the Schwann sheath by proliferation of the endoneurial matrix. Complete surgical excision is the treatment of choice for neurofibroma. ***Conclusion*:** Facial neurofibromatosis surgery remains a challenge for the surgeon. We must have a critical look at our gestures and indications. In particular, provide for surgery after the age of puberty, ligation of the external carotid artery and reconstructive cosmetic surgery.

Keywords: Neurofibroma, plexiform, facial.

INTRODUCTION

Plexiform neurofibromas are considered a rare but disfiguring and devastating complication of neurofibromatosis type 1. Diffuse plexiform neurofibroma is a characteristic lesion of Von Recklinghausen disease [1]. The treatment of plexiform neurofibromas is a real challenge. Medical treatment of NFPs has been frustrating with little evidence of effectiveness. Standard chemotherapy has not been shown to be beneficial and is associated with the risk of treatment-induced secondary malignant neoplasms [2]. Currently surgery is the only effective treatment. This is difficult because of its hemorrhagic nature and the infiltrating aspect of the lesions. In these situations, although temporary cessation of bleeding is usually possible with packing and pressure, permanent control of bleeding has always been achieved either by ligation of the external carotid artery (ECA) or by selective embolization [1, 6].

We report a case of diffuse plexiform neurofibroma of the face successfully operated without hemorrhagic accident after ligation of the external carotid artery at the Sominé Dolo hospital in Mopti (Mali).

CLINICAL OBSERVATION

A 39-year-old woman with no particular pathological history consulted for a monstrous swelling with an irregular texture on the left hemiface (figures 1, 2, 3); evolving for 13 years.

Figure 1, 2, 3: Bulky photo tumor of the left hemi face from the front and profiles

On clinical examination, there were 2 mobile swellings in relation to the skin, they were painless, soft, ulcero - necrotic, the largest of which is in the left orbito -malar seat and the smallest is located at the level of the left preauricular region.

The rest of the examination found cafe-au-lait spots, axillary lentigines.

The diagnosis of Von Recklinghausen's disease was made in the presence of diffuse neurofibroma, café-au-lait stain and axillary lentigines.

She underwent ligation of the external carotid artery and total tumor excision initially (figures 4, 5, 6). The postoperative course was simple. Secondly, she benefited from a thin skin graft (figures 7, 8). With complete healing after 2 months. The anatomo-pathological examination of the surgical specimen concluded to a neurofibroma.

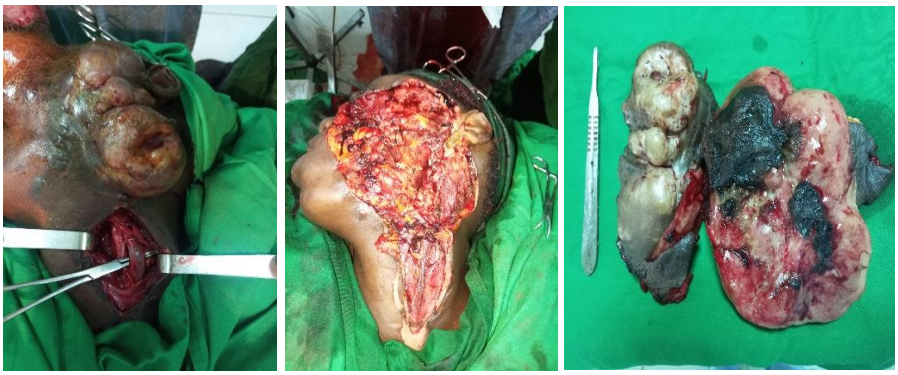

Figure 4, 5, 6: Photo Hospital Ligation of the external carotid artery + resection and surgical specimen

Figure 7, 8: Hospital photo of thin skin graft at 3 months post-operative Face /left profile

DISCUSSION

Neurofibroma is a benign tumor arising from the connective elements of the Schwann sheath by proliferation of the endoneurial matrix. The diagnosis is confirmed by histology, which highlights a fibromatous proliferation made up of narrow and luxurious fibers, grouped in loose spans dotted with regular and fusiform nuclei, these fibers are made up of collagen [2]. Localization in the cervico-facial region is rare. Complete surgical excision is the treatment of choice for neurofibroma because recurrence is possible even if its frequency remains low [4, 5]; it is most often intralesional.

The tumor is often very hemorrhagic, sometimes responsible for operative mortality.

Since peroperative hemorrhages of plexiform neurofibroma are often fatal, our patient first benefited from ligation of the external carotid artery before performing total excision of the mass. Plexiform neurofibromas of the face pose complex repair problems [2]. To repair the loss of substance at the surgical site, which was slow to heal, a thin skin graft was performed on the patient. In the study of Dogra B *et al.*, in India [6], subtotal excision was performed due to impossibility of total excision. Their tumors bled profusely during surgery due to the friable nature of the new vessels. Partial regrowth was observed 6 months later. They therefore recommended, that a sufficient amount of blood be arranged before undertaking the surgical excision of the facial PNF; the tumescent technique should be used in all cases and, after excising the overhanging folds, the skin flap should be re- -draped and a few anchoring sutures between the internal surface of the flap and the underlying periosteum should be placed to avoid downward traction of soft tissue and vital structures after surgery.

Three factors influence the results of neurofibroma surgery: extent of resection, tumor location, and patient age. Subtotal resection has a recurrence rate of less than 40% while total resection reduces the recurrence rate to less than 20%. In addition, patients with cervico-facial location, compared to locations in the trunk and extremities, and subjects under 10 years of age have a higher risk of recurrence [6, 7]. Needle *et al.,* [8] demonstrated that the greatest risk of recurrence of operated plexiform neurofibromatosis was observed in lesions involving the head and neck region, recurrence up to 54% over a period of 10 years.

It is now certain that the destruction of neurofibromas does not lead to any risk of accelerated growth of the remaining neurofibromas or of cancerization [9]. The postoperative course was simple in our patient. However, it appears in a study by Kerrary S. *et al.,* [10] The poor prognosis in Recklinghausen disease is related to the location of the tumors (more often trunk or proximal), the larger size and grade, and the fact that some patients develop multiple sarcomas simultaneously. Local recurrences are common, and metastases (lungs, liver, skin, and bone) usually appear within two years of diagnosis. Despite this risk of malignant transformation, benign schwannomas retain a good prognosis if surgical excision is complete, recurrences are exceptional. On the other hand, malignant schwannoma is a tumor with a poor prognosis with an overall survival rate of 20 to 25% in the case of Recklinghausen's disease and 50% in the case of an isolated tumor [11].

CONCLUSION

Despite advances in surgical and imaging techniques, it is clear that surgery for facial neurofibromatosis remains a challenge for the surgeon. We must constantly have a critical look at our surgical gestures and indications.

In particular, provide for surgery after the age of puberty, ligation of the external carotid artery and reconstructive cosmetic surgery.

DECLARATION OF INTERESTS

The authors declare that they have no conflict of interest.

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