A Great result after Surgical Treatment of a Nasal Neurofibrous Tumor  
(A Case Report and Literature Review)

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**ABSTRACT**

Type I neurofibromatosis (NF) is the most common autosomal dominant disease. It concerns one in 3000 births, the penetrance is close to 100%. Cranio-maxillofacial region is concerned in 10% of the cases. We are reporting the case of a patient who has been surgically cared for in the department of stomatology and maxillofacial surgery at the Rabat specialty hospital, for a nasal plexiform neurofibromatosis with a good post-operative evolution and a very satisfactory esthetic result.

We carried out a tumor reduction surgery with aesthetic objectives, the result of which was satisfactory allowing a good socio-professional insertion of the patient. (Figure 2)

**Observation**

We report the case of a 34-year-old patient who has a history of first-degree inbreeding and has had a progressively increasing nasal swelling since the age of 5 years with no other associated signs.

The clinical examination objectified a soft, painless mass, poorly limited with no inflammatory signs, not increasing in volume in the pro-live position. The mass deformed the nose tip and left nostril wing with a reduction in the nostril opening. (Figure 1)

**Discussion**

Type 1 neurofibromatosis (NF1) is the most common autosomal disease, described by von Recklinghausen in 1883. It belongs to the family of phacomatoses. Its incidence is about 1/3000 births [1]. NF1 [2] is asserted in an individual when two or more diagnostic criteria are present (Figure 3). Ten percent of cases have facial locations [3] and 1% have orbital involvement [4]. Neurofibroma of the face is often associated with other orbito-facial manifestations [5] and extrafacial [5, 6] which must be sought to establish the diagnosis of NF1. Clinically, the lesions can be classified into 2 categories: cardinal signs and the complications.

The clinical expression of the disease has the craniofacial sphere can take several forms. The café au lait spots are the first manifestation of neuro fibromatosis type 1. They are often congenital and rarely appear after the age of 2 years. Their distribution is random, their contours are markedly traced and they are more or less dark brown sometimes at the edge of visibility. The café au lait spots diameters range from 0.5 to 50 cm, but the majority are less than 10 cm. In adolescence, they are present in 90% of cases. In adulthood, they become more pale, barely visible, and

**Figure 1. Nasal mass deforming the nose tip and left nostril wing.**

The rhinoscope examination objectified a reduction in the light of the left nasal cavity without nasal obstruction. The general examination revealed 13 café au lait spots at the trunk and lower limbs with diameters between 2 and 4 cm, with lentigines at the axillary hollow. The ophthalmological examination was normal and the radiographic examination did not show any abnormality of the long bones. The diagnosis of neurofibromatosis type 1 was chosen on 2 criteria and the patient received surgical treatment under general anesthesia.

**Figure 2. Post operative result after tumoral reduction.**
some of them disappear. They are one of the best signs to
diagnose a neurofibromatosis type 1.(7)

Traditionally bone lesions are present from childhood,plexiform neurofibromas appear before the age of 5 years,cranio-orbital, cervical and parotido-jugal neurofibromasappear before the adult age and simple neurofibroids, cutanea nor molluscum can develop throughout life [8]. During puberty, a constant and variable development of the tumors isobserved [9].

1. >5 cafe au lait spots (>5mm prepuberal; >15 mm post puberal)
2. >1 neurofibromas
3. Axillary/inguinal freckling
4. Optic glioma
5. >10 iris nodules
6. Osseous lesion/sphenoid dysplasia (long bone cortex thinning)
7. NF1 in first degree relative

Figure 3. Diagnosis criteria.

The effect of drug treatments on plexiform neurofibroids has so far been disappointing. Several drugs have been tested in Phase I and II studies, such as antihistamines, retinoic acid,anti-angiogenic agents(10), farsyl transferase inhibitors andcytokine modulators, such as Pirfenidone . Encouragingresults from the use of Pirfenidone as an antifibrotic agenthave been reported in pulmonary fibrosis, peritoneal sclerosisis desmoid tumours. This drug was therefore proposed in anattempt to combat the growth of plexiform neurofibroids.(11)

All neurofibroma’s surgery is associated with a majorhaemorrhagic risk and recurrence regardless of its location. Surgery occupies a place of choice, since there is no other therapy, neither preventive nor curative of the disease. This surgery is only palliative because it does not alter the general course of the disease(12). There is no standardisation of the course of action to be taken with regard to the timing andimportance of the surgical action to be performed, especiallyfor children, the majority of authors recommend intratumoralresections, incomplete, the maximum conservatism offunction and aesthetic appearance, especially since thetumour limits are difficult to detect. Complete removals areneither justified nor feasible . The removals are, therefore,most often partial. It is necessary to be satisfied with a partialtumor reduction, the resections are intratumoral and have forsolute purpose a remodelling and a reduction of the tumormass.(13)

Conclusion

This so-called benign condition presents a real localmalignancy. If the diagnosis criteria are clearly defined,there is no specific protocol concerning the age of care and thetherapeutic modalities to be implemented. Its treatment, onlysurgical, still poses many therapeutic problems today.

References