Available online at www.elixirpublishers.com (Elixir International Journal)

Maxillofacial Surgery

Elixir Maxillofacial Surgery 147 (2020) 54885-54886

ISSN: 2229-712X

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

Othmane Bouanani, Olaya El Hamidi, Zineb Zouiti, Salma.Benazzou and Malik Boulaadas

Department of Maxillofacial Surgery Hospital of Specialities-Rabat Faculty of Medicine and Pharmacy-Rabat Mohamed V University

ARTICLE INFO

Article history:

Received: 4 May 2020;

Received in revised form:

6 October 2020;

Accepted: 16 October 2020;

Keywords

Neurofibromatosis,

Van Recklinghausen,

Phacomatoses.

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.

Introduction

NF1 or van recklinghausen's disease is the most common of phacomatoses and neurofibromatoses, it is an autososmic disease that predominates most often by facial damage.

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.(Figure1)

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 cafe au lait spots at the trunk and lower limbs with diameters between 2 and 4cm, 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.

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)

Figure 2. Post operative result after tumoral reduction.

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 craniomaxillo facial sphere can take several forms.The cafe 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 cafe 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

Tele:

E-mail address: bouananiothmane@gmail.com

some of them disappear. They are one of the best signs to dignose a neurofibromatosis type1.(7)

Traditionally bone lesions are present from childhood, plexiform neurofibromas appear before the age of 5 years, cranio-orbital, cervical and parotido-jugal neurofibromas appear before the adult age and simple neurofibroids, cutanes or molluscum can develop throughout life [8]. During puberty, a constant and variable development of the tumors is observed [9].

1. ≥6 cafe au lait spots(>5mm prepuberal; >15 mm post puberal)
2. ≥ 2 neurofibromas/≥ 1 plexiform neurofibroma
3. Axillary /inguinal freckling
4. Optic glioma
5. ≥2 lish 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-angiogenetic agents(10), farsyl transferase inhibitors and cytokine modulators, such as Pirfenidone . Encouraging results from the use of Pirfenidone as an antifibrotic agent have been reported in pulmonary fibrosis, peritoneal sclerosis and desmoid tumours. This drug was therefore proposed in an attempt to combat the growth of plexiform neurofibroids.(11)

All neurofibroma's surgery is associated with a major haemorrhagic 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 and importance of the surgical action to be performed, especially for children, the majority of authors recommend intratumoral resections, incomplete, the maximum conservatism of function and aesthetic appearance, especially since the tumour limits are difficult to detect. Complete removals are neither justified nor feasible . The removals are, therefore, most often partial. It is necessary to be satisfied with a partial tumor reduction, the resections are intratumoral and have for sole purpose a remodelling and a reduction of the tumor mass.(13)

Conclusion

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

References

[1] Pinson S, Cre´ange A, Barbarot S, Stalder JF, Chaix Y. Neurofibromatose 1 : recommandations de prise en charge. Arch Pediatr 2002;9:49–60.

[2] NIH consensus development conference statement. Neurofibromatosis. Arch Neurol 1988;45:575–8, http://www.ncbi.nlm.
nih.gov/pubmed/?term=NIH+Consensus+Developement+

[3]Latham K, Buchanan E, Suver D, Gruss J. Neurofibromatosis of the head and neck: classification and surgical management. Plast Reconstr Surg 2015;135:845–55.

[4]Poole MD. Experiences in the surgical treatment of cranioorbital neurofibromatosis. Br J Plast Surg 1989;42:155–62.

[5]Welkenstein P, Zeller J, Ismaili N. Welkenstein P, Zeller J, Ismaili N. Neurofibromatoses. Encycl Méd Chir, Dermatologie 2002;10:1-10.

[6] Ortone JP, Brocard E, Floret D, Perrot H, Thivole Ortone JP, Brocard E, Floret D, Perrot H, Thivolet J. Valeur diagnostique des taches café-au-lait (T.C.L.). Ann Dermatol Venereol 1980; 107 : 313-27.

[7] Kissel P, Schmitt J, André JM. Kissel P, Schmitt J, André JM. Phacomatoses. Encycl Méd Chir, Neurologie 1975;10: 1-6.

[8]Pinson S, Cre´ange A, Barbarot S, Stalder JF. Recommendations for the treatment of neurofibromatosis type 1. J Fr Ophtalmol 2002;25:423–33.

[9] Littlewood AH, Stilwell JH. The vascular features of plexiform neurofibroma with some observations on the importance of pre-operative angiography and the value of pre-operative intra-arterial embolization. Br J Plastic Surg 1983;36:501–6.

[10]Packer RJ, Rosser T. Packer RJ, Rosser T. Therapy for plexiform neurofibromas in children with neurofibromatosis 1: an overview. J Child Neurol 2002; 17 : 638–41.

[11] Babovic D, Ballman K, Michels V, Mcgrann P, Lindor N, King B, et al. N, King B, et al. Phase II trial of pirfenidone in adults with neurofibromatosis type 1. Neurology 2006; 67: 1860–2.

[12]Heuze Y, Piot B, Mercier J. Heuze Y, Piot B, Mercier J. Difficultés de la prise en charge chirurgicale des manifestations faciales de la neurofibromatose de type 1 ou maladie de von Recklinghausen chez l'enfant. Rev Stomatol Chir Maxillofac 2002; 103 (2): 105-13.

[13]Banzet P, Bodin B. Banzet P, Bodin B. Chirurgie plastique dans la maladie de recklinghausen. Sem Hop Paris 1985; 61: 2705-8.