

## A Giant Disabling Neurofibroma in a Young Adult Carrier of Neurofibromatosis

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### Abstract

### Case Report

The neurofibromatosis is a genetic disease that affects multiple tissues with different forms of clinical presentation. The neurofibroma is a benign tumor, which can be presented in two forms: solitary in young adults and the multiple forms associated with neurofibromatosis type 1 or Von Recklinghausen disease. The evolutionary profile of this disease is very variable. Anything can go from a few café au lait spots to a disabling state. The treatment of neurofibromas is exclusively surgical. We report the case of a 40-year-old man with Recklinghausen's disease with a giant neurofibroma in the left facial and neck area causing a disability and subsequently traumatized by the patient himself. The craniofacial scan revealed a large left subcutaneous tissue formation measuring approximately 249X22X136 mm and confirming the tomodensitometric appearance of neurofibromatosis type I. Therapeutically a reduction in tumor volume was performed to relieve the patient; which allowed us to make an anatomopathological study. The histology showed a fasciculate tumor proliferation made of fibroblastic and nerve cells. The patient evolved favorably and was satisfied to the point that he no longer wanted additional surgery. Neurofibromatosis is not curable in itself but surgery can restore the patient's functionality by ending the disability caused by certain complications.

**Keywords:** Neurofibromatosis, neurofibroma, Von Recklinghausen's disease, disability.

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## INTRODUCTION

Neurofibromatosis (NF) is a genetic disease that affects multiple tissues with different clinical forms of presentation. Type 1 NF occurs with a frequency in the general population of 1 / 2,000 to 1 / 3,000 inhabitants [1].

Neurofibromas are the cardinal sign of the disease. They are small benign soft tumors, mobile with the skin, sessile or pediculated in elastic and depressive consistency, developing along nerves and nerve sheaths in the dermis and epidermis. The trunk is their main seat, but any other part of the body can be touched. There is orofacial involvement in 1 to 22% of cases [2].

We report a disabling and traumatized giant neurofibroma in a 40-year-old patient with von Recklinghausen's disease. The size of the neurofibroma

and its location are even rarer than we are seen motivated to do this postponement. The treatment consisted of a total excision of the largest and disabling neurofibroma.

## CASE REPORT

This is a 40-year-old patient with a history of neurofibromatosis with multiple tumors on the body. One of them has evolved in a disproportionate way for about two years to cause a real handicap by preventing him from carrying out his daily activities. On physical examination, a very large tumor mass was found in the left cervicofacial area with ulceration in the lower part accompanied by peripheral inflammatory signs. In addition he had an inability of the head to support the tumor requiring him to be assisted by his upper or lower limbs as appropriate (fig.1).



**Fig.1.a**



**Fig-1.b**

Figure 1: Clinical aspect of giant neurofibroma. The tumor is carry up by the patient by superior (Fig.1.a) or inferior (Fig.1.b) limbs according the situation. We can observe too on all the body small neurofibroma attesting the neurofibromatosis.

A CT scan was indicated and indicated the presence of a large subcutaneous tissue formation at the left temporal starting point measuring approximately 249X22X136 mm. This formation was heterogeneous hypodense with a discrete enhancement after injection of PDC seat of a fluid pocket of 189X132 mm at the

bottom containing thick wall air bubble in relation to the puncture trauma. Inside the mass came into contact with the masseter and left sterno-cleido-mastoid muscles with persistence of greasy edginess. It came into contact with the ipsilateral mandibular ramus without detectable bone lysis. At the back it came into contact with the left jugulo-carotian axis with repression of the jugular vein which remains permeable. In conclusion, he evoked the CT appearance of a type I left temporal neurofibromatosis with an inflammatory complication of traumatic origin very probably (fig.2).





**Fig-3: Immediate post operatory stat of the patient. The suture lines are observable.**

The anatomical piece weighed 7 kilograms and was sent to anatomy pathology for study. She objectified a fasciculate tumor proliferation made of fibroblastic cells and fibroblastic cells and nerve cells. Proliferation also has an inflammatory reaction made of lymphoplasmocytes and neutrophils. Absence of cytonuclear atypia. The lymph nodes examined show hyperplasia of lymphoid follicles with clear germinal centers without cytonuclear atypia. The pathological

study concluded a neurofibroma remodeled by inflammation with adenitis reaction. The patient was hospitalized for 3 days. No complications were observed. He was released and followed outpatient. The removal of the threads takes place on the 10th day without complications. Definitively released the 14th day it was reviewed 6 months 10 days later with a fairly acceptable result (fig.4).



**Fig-4: Post operatory stat of the patient at 45 days after. We can observe the overflow of tissue which would be removed in a second time**

## COMMENTS AND DISCUSSION

Pathologically, the neurofibroma is composed of polymorphic cells including Schwann cells, perineural cells and fibroblasts that are contained in a mucopolysaccharide matrix. It infiltrates between fascicles of the nerve, along its path, which could make resection difficult and dangerous [3].

The neurofibroma results from tumor hyperplasia of all nerve components, both schwannian and fibroblastic elements [4].

Neurofibromas are benign tumors. Cutaneous neurofibromas are small soft tumors, mobile with the skin, sessile or pediculated to the type of molluscum

pendulum. In pulpit, rosy or purplish color, their consistency is particular, elastic and depressible. Cutaneous neurofibromas do not appear until puberty and are exceptionally absent in adulthood (95% of adults with NF1 are carriers) [5].

The neurofibroma is a benign tumor, which can be presented in two forms: solitary encountered in young adults and the multiple forms associated with neurofibromatosis type 1 or VON Recklinghausen disease [6] like in our case.

The evolutionary profile of this disease is highly variable. Anything can range from some café au lait spots to a disabling attack. This disability can sometimes be traced to the only cosmetic consequences such as those observed in cases with neuvrome plexiform whose frequency fortunately remains limited to less than 5 per cent of patients [4]. Neurofibromas can reach large dimensions without producing a neurological deficit; the facial or cervical deformity being the most remarkable by the size of the tumor itself [7].

The compromise caused by neurofibromas depends largely on their location. Generally the skin lesions cause deformities while the deeper lesions tend to generate a functional included [8].

Malignant tumors of the nerve sheaths (formerly neurofibrosarcoma) are the main complication of NF1 in adulthood. Their risk of occurrence during the life of a patient with NF1 is of the order of 3 to 4p100. The diagnosis must be early. It develops from isolated nodular neurofibromas or plexiform, cutaneous or visceral neurofibromas. The signs of call are a rapid increase in size of an old or newly emerged neurofibroma, pain, the appearance or modification of pre-existing neurological signs. The biopsy excision of suspicious nodules must then be performed without delay [5].

The treatment of neurofibromas is exclusively surgical. Complete resection is the ideal treatment. However, it is sometimes not possible due to the significant functional deficit that it would cause, in which case partial resections could be accepted [7].

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