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Plexiform neurofibromas

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Case Report

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Abstract: The plexiform neurofibroma is a rare and benign tumor often associated with neurofibromatosis type 1. The aim of this work is to show, through three clinical cases, the clinical characteristics and the therapeutic means of plexiform neurofibromas in our department. The constant elements found in all patients are benign peripheral tumors developed at the expense of nerve sheaths. An extensive tumor or malignant transformation is the surgical indication. The majority of authors admit the difficulty of this surgery in view of the difficulty of the dissection, the importance of the haemorrhagic risk, the frequency of recurrences. Neurofibromatosis is characterized by a very great variability of its clinical expression. The plexiform neurofibromas of the face are especially difficult to control. The results remain modest and recidivism is frequent.
Keywords: Neurofibroma, plexiform, tumor

INTRODUCTION

The plexiform neurofibroma is a rare and benign tumor often associated with neurofibromatosis type 1 (NF1) or Von Recklinghausen disease which is an autosomal dominant disease. While it is associated with a variety of phenotypic presentations, it is the plexiform variant that is of particular concern because it can become extremely disfiguring and has a predisposition to malignant degeneration. The aim of this work is to show, through three clinical cases, the clinical characteristics and the therapeutic means of plexiform neurofibromas in our service through three clinical cases.

MATERIAL AND METHODS

Three clinical cases have been followed up to scarring that present a plexiform neurofibroma

RESULTS

Case 1: Patient aged 53, followed for neurofibromatosis type 1 with excision of a neurofibroma of the face at the age of 7 years who had a recurrence of neurofibroma with progressive increase in volume. The examination found a patient in good general condition; plexiform, pedicled, multilobed 20 cm by 14 cm at the right fronto-temporo-parietal. CT showed a locally aggressive fronto-temporo-parieto-occipital tumor process with lysis of the zygomatic arch and infiltration of the right temporal muscle and the outer cantus and a discreet scalopping on the parietal bone. Total excision of the mass with excision of the cutaneous excess opposite and direct suture with satisfactory aesthetic result. The operative specimen weighed 1,2 Kg.

Case 2: A 16-year-old patient followed for neurofibromatosis who had a progressive increase in left leg volume. Conscious examination in good general condition, presence of coffee-milk stains; the left leg was enlarged with a soft plexiform consistency with no inflammatory sign or pain on palpation or dorsiflexion. MRI of the leg showed diffuse muscle and subcutaneous infiltration of the left leg with bone involvement of the left ankle. She benefited from a partial excision of the tumor and excision of the skin opposite and direct suture.

Case 3: Patient aged 17; followed for neurofibromatosis type 1; operated at the age of 12 years for a plexiform tumor of the right hemiface; presents itself for right hemifacial ptosis of progressive aggravation. The examination finds a patient in good general condition, complete right hemifacial ptosis interesting the eyelids; cheek and lips with milk coffee spots next to it. CT showed right hemifacial involvement infiltrating the deep spaces with dysplasia of the great wing of the sphenoid, right microphthalmia and right hemicranial atrophy. The patient underwent exeresis of the tumor with suspension of the lips and the right external cantus.

DISCUSSION

The NF1 or Recklinghausen disease has been known for over a century thanks to the works of Friedrich Daniel von Recklinghausen (1833-1910) [1]. It is characterized by a great clinical variability. Despite this variable clinical expression, the constant elements found in all patients are benign peripheral tumors

developed at the expense of nerve sheaths: neurofibromas. Histologically, these tumors consist mainly of Schwann cells and fibroblasts. Neurofibromas can be classified into four types: cutaneous tumors, subcutaneous tumors, deep nodular tumors and finally plexiform tumors [2]. The first three types tend to be localized and small while plexiform neurofibromas can be enormous, infiltrate adjacent tissues and lead to disfigurement. Plexiform neurofibromas progressively infiltrate adjacent tissues, deform bone structures, and interfere with sensory functions (vision, hearing), and with facial growth leading to significant psychological repercussions. There are also compressive syndromes, which give breathing difficulties, dysphagia, and speech problems. Given that these tumors are benign and most often quiescent, a simple follow-up of the patients may suffice. However, a functional deficit, an extensive tumor or a malignant transformation pose the surgical indication [3].

Indeed, up to 10% of neurofibromas undergo a malignant transformation in neurofibrosarcoma of formidable prognosis. The primary goal of the surgery is not the complete excision of the tumor, which would require mutilating excisions, but its reduction with the least possible of sensitivomotor functional sequelae. The majority of authors admit the difficulty of this surgery in view of the difficulty of the dissection, the importance of the haemorrhagic risk, the frequency of recurrence as is the case with our patients. Preoperative arteriography of neurofibromas may be required with selective embolization to decrease intraoperative bleeding risk. The recurrence varies indeed between 20 and 40% for the subtotal resections, and goes to 60% when the resection is less than 90% of the tumor [4]. Craniofacial localizations have a poorer prognosis and surgery before the age of ten leads to recurrence during puberty. The life expectancy of these patients ranges from 54 to 59 years due to malignant changes and vascular complications of the disease. Hence the need for rigorous monitoring of these patients by a multidisciplinary team without forgetting the psychological care and genetic counseling [5].

CONCLUSION

Neurofibromatosis is characterized by a very great variability of its clinical expression. Plexiform neurofibromas, particularly on the face, are difficult to control because of soft tissue infiltration and associated bone dysplasia. The results are modest and recidivism is frequent. The risks of tumors and the totally unpredictable evolution of the disease require regular and multidisciplinary monitoring of patients.

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