Unusual Evolution of Plexiform Neurofibroma in the Scalp: A Case Report

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ABSTRACT

The neurofibromatosis is a frequent and polymorphic genetic disorder. The severity is related to the complications. The degeneration of neurofibroma is a very rare complication of neurofibromatosis. In the literature, a few cases of solitary neurofibroma, which turned into a malignant tumor were reported. In our case, we described a very rare clinical case of neurofibrosarcoma in the scalp, and surgical treatment.

KEYWORDS

Neurofibroma; Neurofibromatosis; Scalp; Schwannoma; Surgery

INTRODUCTION

Neurofibromatosis type 1 is a congenital disorder, affecting one individual in 2500 births.1 It is one of the most common genetic diseases, resulting due to a mutation of the NF-1 gene; The mutation is spontaneous in half of the cases and inherited as autosomal dominant in the other half.2 Penetrance is 100% at the age of 5 years, however expressiveness, whether for the severity or location, is extremely variable even within families whose members have inherited the same mutation.3 Neurofibromatosis is characterized by café-au-lait spots, lentigines and neurofibromas; with eye signs like Iris hamartoma; also CNS glioma and bone signs like dysplasia, pseudoarthrosis and scoliosis.4 The severity of disease usually is due to its complications. One of the more morbid complications is malignant degeneration of plexiform neurofibromas.5 The malignant Schwannoma is one of the forms of degeneration of the neurofibroma in neurofibrosarcoma.6 Here, we described a very rare clinical case of neurofibrosarcoma in the scalp, and surgical treatment.

CASE REPORT

We report our experience with a forty-three years old patient, known carrier of Neurofibromatosis type 1, who presented to us with a large occipital tumor. Biopsy revealed presence of malignant Schwannoma (Figure 1). The treatment consisted of wide local...
Fig. 1: A forty-three years old patient, known carrier of Neurofibromatosis type 1 with a large occipital tumor and malignant Schwannoma.
excision and reconstruction. Intra-operatively, the tumor was found to infiltrate the periosteum and abutting the outer table of the occipital bone. A wide local excision of the tumor, the periosteum and the outer table of the bone was done (Figure 1). Histopathology confirmed malignant Schwannoma infiltrating into the outer table. The wound was initially managed by moist occlusive dressings in the interim till the histopathology report was available. One month after the initial surgery, the wound was closed with a large bipedicled bucket handle flap from the parieto-occipital scalp region (Figure 1). There was some graft loss on the donor area and hence the patient was again scheduled for skin grafting of the donor area after two months (Figure 1). The postoperative course was unremarkable and without any complications. The patient was then referred to the Oncology unit for radiation therapy.

**DISCUSSION**

The degeneration of neurofibromas is a rare complication of neurofibromatosis, and the lifetime risk for a patient of NF1 is of the order of 3 to 5 percent. Histologically, the neurofibrosarcoma is characterized by the presence of fusiform cells, probably derived from Schwann cells. Collagen fibers are rare. There is a presence of coarse reticulin fibrils, in parallel rows between fusiform cells which is very characteristic of the lesion. Signs of malignancy are represented by pleomorphic cells, giant cells, mono or polynuclear cells, an excess of mitoses, an invasion of surrounding tissues, and vascular invasion.

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**REFERENCES**


