

Giant Malignant Peripheral Nerve Sheath Tumor of the Scalp

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Abstract-

Purpose: Malignant peripheral nerve sheath tumors (MPNST) constitute a group of soft tissue neoplasms with neurogenic origin. Association of MPNST with neurofibromatosis type 1 (NF1) has been frequently reported as a result of malignant transformation of a pre-existing plexiform neurofibroma. However, most of the cases are small and few have been described with giant dimensions. We describe a case of a giant MPNST in an unusual location (scalp), in a NF1 patient.

Case Report: A fifty-six years old woman with a history of NF1 since childhood was admitted due to a rapid growing of a scalp tumor in occipital region. The mass had 15 cm in the largest dimension and a central ulceration with purulent secretion. Computed tomography revealed no bone invasion. The tumor was surgically removed and the histopathological examination revealed a neurofibrosarcoma. The patient had a good post operative evolution and notwithstanding the tumor size, no metastasis was found and she was discharged to oncologic follow-up.

Conclusion: Given the poor prognosis of MPNST, patients with NF1 should be aware that neurofibromas can undergo malignant transformation and report signs such as rapid growth of a pre-existing lesion.

Key Words: nerve sheath tumors, neurofibromatosis type 1, giant, malignant transformation, scalp, neurofibromas.

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INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNST) constitute a group of soft tissue neoplasms with neurogenic origin. Association of MPNST with neurofibromatosis type 1 (NF1) has been frequently reported as a result of malignant transformation of a pre-existing plexiform neurofibroma¹. It is important to note that MPNST typically occurs in the extremities, with few reports in other locations. In the present report, we will

describe a case of MPNST in an unusual location (scalp). The tumor reached a giant dimension in few weeks and required surgical intervention.

CASE REPORT

A 56-year-old white woman with NF1 was admitted to the hospital due to a rapid growing (in a three-month span) of one of the tumors in the occipital region of the scalp. She also complained of neck pain, because of a

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Figure 1. Preoperative appearance of occipital region demonstrating a huge mass with central ulceration and purulent discharge.

continuous tendency to head extension caused by the tumor weight. The diagnosis of NF1 had been made in her childhood, based on clinical assessment. Café-au-lait spots and a skin biopsy compatible with neurofibroma had confirmed the diagnosis. Regarding her family history, she had three daughters and one granddaughter with NF1.

On physical examination, she was in good general condition. Concerning her neurofibromatosis, she had both sessile and pedunculated tumors throughout the body, in subcutaneous and skin tissues, with different sizes. The mass was located in the occipital region and had approximately 15cm in width and length, with central ulceration and purulent discharge (Fig. 1). Computed tomography scan revealed a large and irregular mass, without invasion of the bone.

An incisional biopsy was performed and histopathological examination revealed a high density of cells with fusiform and pleomorphic aspect (Fig. 3). Immunohistochemical findings were negative for CD57, CD34, desmin, smooth muscle actin and positive for S-100 (Fig. 3). These findings were compatible with MPNST.



Figure 2. Postoperative aspect.

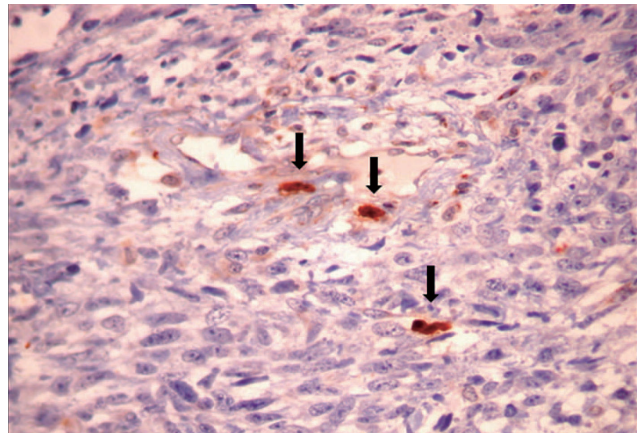


Figure 3. Nuclear and cytoplasmic immunostaining for S100 (arrows). Note the high cell density and pleomorphism.

She underwent a complete resection of the tumor with reconstruction of the scalp with flaps from the surrounding tissue (Fig. 2). The tumor measured 15.5×14.5×10.5 cm and weighed 1.05 pound. Frozen section biopsy showed tumor-free margins.

The patient had a good postoperative evolution. Notwithstanding the tumor size, no metastasis was found

and she was discharged to oncologic follow-up one week after surgery.

DISCUSSION

Although malignant degeneration of a plexiform neurofibroma is a fairly common finding in NF1, MPNST is a rare condition when located either on the scalp or on the neck^(2,3,4).

The lifetime risk of developing MPNST is about 10% and is well known that is an aggressive malignancy, with an five-year event-free and overall survival rate of 19 and 28 percent, respectively^(5,6).

The main cause of malignant transformation of neurofibromas is still under investigation, but there have been some evidences that suggest mutations in the NF1 tumor suppressor gene and impairment in the expression of proteins involved in signaling pathways of cell division, apoptosis and DNA repair as p16 and p53^(1,3,7).

Despite a growth rate that varies from months to years, patients with NF1 has a tendency to larger tumors at diagnosis^(4,8,9). Most of the times, the first presentation of malignant transformation is significant chronic pain or rapid growth of a nodule within a previous plexiform neurofibroma. In this report, the rapid growth and location on the scalp was atypical, as well as the tumor dimensions, causing symptoms like daily headache and neck pain. Although there have been previous descriptions of MPNST with similar dimensions, they have required a longer period to reach a giant size (two years)⁽²⁾. Fukushima et al described a Malignant Schwannoma with 210 mm in the largest diameter. This tumor was also ulcerated and had a poor evolution due to metastasis 10. Agrawal et al used the term “double-head” to describe a huge mass on the posterior side of the scalp in a 25-year-old woman, diagnosed as a Benign Schwannoma 4.

In the present case, the lesion also showed signs of infection, with central ulceration and purulent discharge, which certainly contributed to the symptoms of depressed mood and social isolation. The rapid growth and the delay between the onset of symptoms and the

medical care were responsible for the dimensions achieved by the tumor, as well as the infection. To the best of our knowledge, this is the second report of a MPNST of the scalp with such giant dimensions.

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