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Malignant Peripheral Nerve Sheath Tumors or MPNSTs of Nasal Cavity: Case Report

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Abstract | **Case Report**

Malignant peripheral nerve sheath tumors or MPNST are rare tumors and the main complication of neurofibromatosis type1. MPNST is most common in young adults and middle-aged adults. They occur mainly in the roots of the limbs and trunk and less frequently in the head and neck. MRI is the examination of choice in this type of tumor. The diagnosis is histological. The main treatment is surgery and the adjuvant treatment is indicated to prevent any recurrence. We report a very rare case of MPNST tumor of the nasal cavity and we will underline through this work the aggressiveness and the unpredicted prognosis of this malignant tumor.

Keywords: Malignant peripheral nerve sheath tumors, MPNST, nasal cavity.

INTRODUCTION

Malignant peripheral nerve sheath tumors or MPNST are rare tumors and the main complication of neurofibromatosis type1. MPNST is most common in young adults and middle-aged adults [1]. These tumors are defined by the 2013 WHO classification as tumors malignancies developed either from a peripheral nerve; either from a benign tumor of the nerve sheaths; or in a patient with neurofibromatosis type 1.

Cervicofacial nerve tumors poses nosological and topographical problems and present a clinico-radiological and histological polymorphism. Clinical signs of von Recklinghausen's disease are very important to confirm the diagnosis [2].

MRI brings a better delimitation of the relationship between tumors and adjacent structures, especially when the local extension is complex. The treatment of choice is based on extended resection. MPNSTs has classically been considered radio-resistant and chemo resistant. Its evolution depends on the association with NF1 and the quality of the first resection [3].

The main aim of our report was to study the particularities clinical, radiological and therapeutic aspects of the malignant peripheral nerve sheath tumors located in the nasal cavities.

CASE REPORT

We report a case of a 45-year-old woman, operated in 2009 for a left nasal tumor, who presented with a whitish, firm, tender mass associated with unilateral nasal obstruction and purulent rhinorrhea, filling the entire left nasal cavity. Exo buccal examination revealed a left jugo-nasal mass with no inflammatory signs opposite, effacing the nasogenial fold, mobile in relation to the superficial plane and fixed in relation to the deep plane, measuring 4 cm in long axis (Fig 1). There was no hypoesthesia in the V2 territory or facial palsy. Cervical examination showed no cervical adenopathy. The patient underwent a biopsy which revealed a synovialosarcoma. The radiological workup consisted of a facial MRI (Fig 2), which showed a tissue process measuring 63mm with locoregional extension classified as T2N0.The patient was referred to the oncology center for neoadjuvant chemotherapy. Taking into account the reduction of the tumor volume by 50%, our patient was admitted to the operating room under general anesthesia; she benefited from a complete tumor removal by paralateronasal approach. The surgical specimen was sent for anatomopathological study, whose complement by immunohistochemistry revealed a high grade MPNST. The resection of the tumor was complete with healthy surgical margins. The patient was readmitted for adjuvant radiotherapy.

One year after radiotherapy the patient presented a massive tumor recurrence. The surgery was not possible at this stage and the patient was referred to palliative treatment.

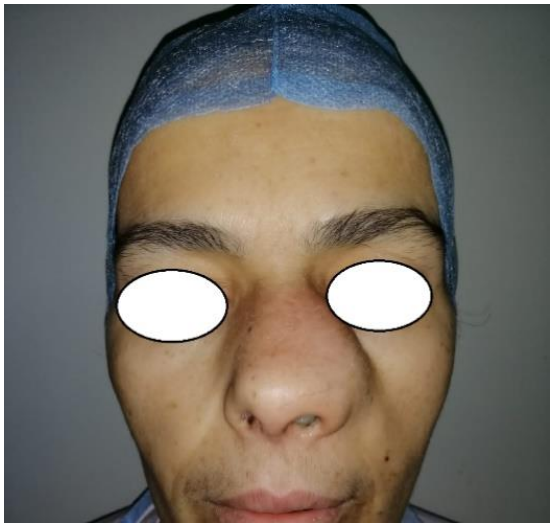

Figure 1: Picture of the patient showing a jugo-nasal mass

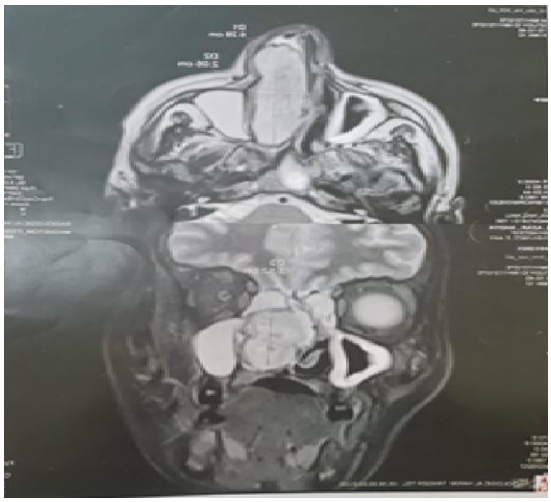

Figure 2: Facial MRI, axial and coronal sections show a tissue process filling the nasal cavity with a locoregional extension

DISCUSSION

Malignant peripheral nerve sheath tumors or MPNSTs are rare, usually appearing between the third and fourth decades [4]. Also known as malignant schwannomas, they represent 2% to 5% of soft tissue sarcomas [5]. There are many morphologic variants of more complex identification that may pose problems of differential diagnosis with other spindle cell lesions. Topographically, they occur mainly in the roots of the limbs and trunk and less frequently in the head and neck. They usually measure more than 5 cm and are whitish in color with areas of necrosis and hemorrhage [1]. MRI is the examination of choice in this type of tumor and shows an irregular, polylobed appearance with heterogeneous contrast [6]. The diagnosis is histological showing a dense, fasciculated proliferation of spindle-shaped elements with abundant cytoplasm, chromatic, tapered and elongated nuclei. In 75% of cases, necrosis is present, as well as mitotic figures [7]. Immunohistochemistry contributes effectively to the diagnosis. The most frequently used markers are the S100 protein, which allows the differentiation of these tumors from other differential diagnoses such as monophasic synovialosarcoma, leiomyosarcoma, whereas cytokeratin and desmin are rarely identified. However, despite these technical advances, MPNSTs tend to be underrecognized, both clinically and histologically, and the initial diagnosis is often missed [8].

In order to prevent local recurrence; it is paramount to perform a wide excision with healthy surgical margins. Neo-adjuvant or adjuvant radiotherapy improves local control and decreases the risk of recurrence but does not seem to have an effect on overall survival. The value of postoperative radiotherapy has been reported in high-grade MPNST, >5 cm in size, or when the limits of excision are invaded [2, 9]. Chemotherapy is appropriate in very advanced forms preoperatively or in inoperable forms. MPNST are often very aggressive and their high rate of metastasis and local recurrence justifies regular and prolonged surveillance.

The reported pejorative prognostic factors are: tumor size > 5 cm, positive surgical margins, local recurrence and rhabdomyosarcomatous differentiation, underlying neurofibromatosis.

CONCLUSION

Malignant peripheral nerve sheath tumors or MPNST are rare tumors, cervico-facial localization poses nosological and topographical problems. MRI is the imaging test of choice. The definitive diagnosis is established by histology. The treatment is based on a large excision of the tumor. Radiotherapy improves local control but the prognosis remains poor.

Conflicts of Interest: The authors declare no competing interest.

Authors' Contributions: All the authors participated in the treatment of this patient and in the redaction of this article.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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