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Malignant peripheral nerve sheath tumor of intracranial nerve: A case series review

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Abstract

Objectives: The incidence of malignant peripheral nerve sheath tumor (MPNST) is approximately 0.001%. Those involving intracranial nerves are even more exceptional. Little information is available concerning work up and management. Our objective is: (1) to review all cases of intracranial MPNST described in the literature, (2) to highlight the suspicion of intracranial MPNST, (3) to identify the gross pathology, the histopathology, the immunohistochemistry, (4) to discuss the differential diagnosis, the treatment, the recurrence rate, the follow-up, the incidence of metastasis and the prognosis.

Methods: We reviewed English, Spanish and French literature published from 1950 to date. We used the following Keywords: ''malignant peripheral nerve sheath tumor'', ''cranial nerve'', ''neurosarcoma'', ''malignant schwannoma'', ''neurofibroma'', ''malignant neurofibroma'' and ''nerve tumor''. We considered cases where MPNST involved an intracranial cranial nerve. The results yielded 20 relevant studies, in which 31 patient's records were transcribed. We also added our case to this series.

Results: We identified 32 cases of cranial MPNST including our case. The age ranged from 5 to 75 years old with most patients being in the 5th and 6th decade. Male to female ratio is 2.5:1. Most cases are developed sporadically (50%), 31% arise from a malignant transformation of schwannoma and 19% from a neurofibroma. Imaging findings were not specific. The cranial nerve VIII is the most involved (15/32), followed by the Vth (10/32) and the VIIth (5/32). 4 cases had neurofibromatosis type 1 and 2 had neurofibromatosis type 2. MPNST will strongly express protein S-100 and collagen IV-laminin. 13 cases were treated with radiotherapy for tumor recurrence and metastasis. In these cases the survival rate was better than the cases without radiotherapy. Fatal outcome occurred in 66% of patients whereas 19% were reported alive with or without complications. The seven cases reported to have metastasis were entirely to the spine. The mean time of recurrence or metastasis is 12.2 months.

Conclusion: MPNST of cranial nerves are very rare. In neurofibroma, even though MPNST is mainly associated to type 1, we should keep in mind its association to NF2. Mainstay of treatment is radical resection with adjuvant radiotherapy. Inaccessibility of cranial MPNST may explain the subtotal resection and thus the poor prognosis. Metastasis to the spinal cord is the most frequent one. A close postoperative follow-up is mandatory to eliminate recurrence.

Keywords: Malignant peripheral nerve sheath tumor; Neurofibroma; Neurosarcoma; Malignant schwannoma; Neurofibromatosis; MPNST; Plexiform

Contents

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1. Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a rare tumor accounting for 5% of soft tissue sarcomas [1]. Those involving intracranial nerves are even more exceptional. It is reported that MPNSTs occur de novo or in the setting of neurofibromatosis type 1 where they are usually a transition from a plexiform neurofibroma. Transition from a schwannoma is very rare [2,3]. Intracranial MPNSTs are one of the most aggressive tumors. Little information is available concerning work up and management. Literature is limited to isolated case reports and small series.

Our objective is to review all cases of intracranial MPNST described in the literature in order to highlight the suspicion of intracranial MPNST, to identify the gross pathology, the histopathology, the immunohistochemistry, to discuss the differential diagnosis, the treatment, the recurrence rate, the follow-up, the incidence of metastasis and the prognosis.

2. Materials and methods

We performed a MEDLINE database search for MPNST related articles. We reviewed English, Spanish and French literature published from 1950 to date. The electronic search was conducted using the Keywords: ‘‘malignant peripheral nerve sheath tumor’’; ‘‘cranial nerve’’; ‘‘neurosarcoma’’; ‘‘malignant schwannoma’’; ‘‘neurofibroma’’; ‘‘malignant neurofibroma’’ and ‘‘nerve tumor’’. We considered cases where MPNST involved an intracranial cranial nerve. We excluded cases involving neck cranial nerves and brain parenchyma. The results yielded 20 relevant studies; in which 31 patient’s records were transcribed. We also added our case to this series. The information from the reports was analyzed to characterize the clinical aspects; the radiological findings; the histopathology and immunohistochemistry; the treatment; the rate of recurrence; the follow-up; and the incidence of metastasis related disease.

2.1. Case presentation

A 62-year-old man with no significant medical history and non-contributory family history presented with a 2 years history of left hemifacial paresthesia and tingling. He describes also a paroxystic shooting pain in the 3rd branch of the trigeminal nerve.

He has to soften his diet because of difficulty masticating. He came to medical attention for worsened symptoms. The patient was referred to our otolaryngological department in a tertiary care center. Neurological exam was significant for sensory deficit in the mandibular territory of the left trigeminal nerve. A magnetic resonance imaging (MRI) was performed and showed an expansive enhancing lesion arising from the 3rd branch of the left trigeminal nerve. The lesion was unique and extends from the Meckel’s cavum to the infra temporal fossa through an enlarged foramen ovale (Fig. 1). A schwannoma was suspected. Surgical excision was performed jointly with the neurosurgery team. Tumor was accessed by subtemporal craniotomy and partial petrosectomy. The tumor was totally removed including

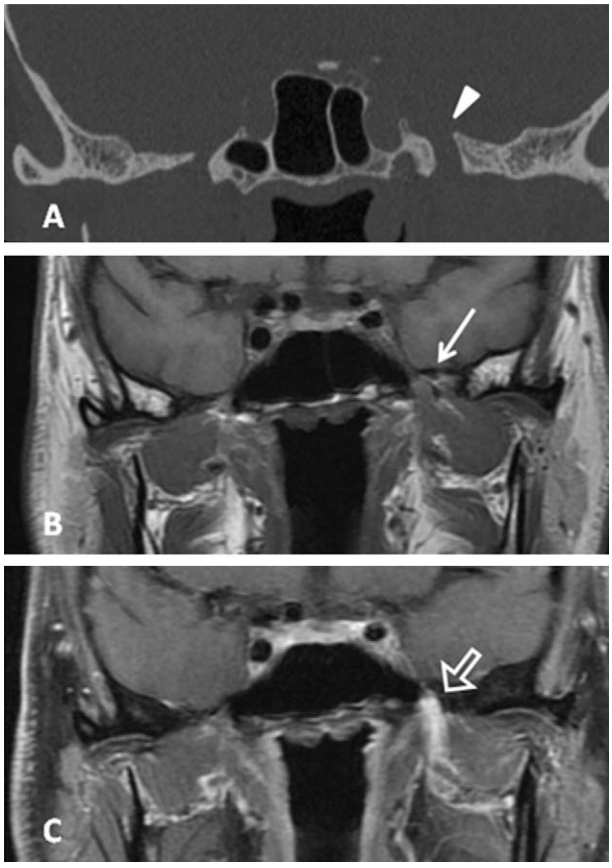

Fig. 1. Preoperative computerized tomography scan (A) showing in a coronal view the enlarged foramen ovale (arrow head) by the tumor. Coronal view of T1 weighted magnetic resonance imaging study ((B) without contrast and (C) with contrast) showing a hypodense tumor (white arrow) and a tumor enhancement (open arrow).