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Malignant nerve sheath tumor involving glossopharyngeal, vagus and spinal nerve with intracranial-extracranial extension and systemic metastases in a patient with type 1 neurofibromatosis: A case report

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ABSTRACT

INTRODUCTION: Intracranial malignant peripheral nerve sheath tumors are an extremely rare pathology with a high morbidity and mortality. Epidemiological, clinical and prognostic data are scarce and with little certainty in the literature. The aim of this paper is to report for first time in English literature, the case of a patient with type 1 neurofibromatosis, who presented a malignant peripheral nerve sheath tumor that involved the left glossopharyngeal, vagus and spinal nerves with intracranial and extracranial extension through jugular foramen and systemic metastases.

PRESENTATION OF CASE: A 37 years-old female patient with malnutrition and Villaretís syndrome. It was confirmed by brain magnetic resonance imaging and PET-CT the presence of a neoplasic lesion which was radiologically compatible with malignant peripheral nerve sheath tumor with systemic metastases. Partial surgical resection was performed; the patient postoperative course was without significant clinical improvement but with added peripheral facial palsy. The patient did not accept adjuvant management because of personal reasons.

DISCUSSION AND CONCLUSION: Behavior therapy is unclear due to the low frequency of the disease and the lack of case series, representing a challenge for the physician in its approach and a poor prognosis for the patient.

1. Introduction

Peripheral nerve sheath tumors (PNST) are histologically benign and slow growing lesions with an incidence of 0.001% in general population [1]. Schwannomas and neurofibromas represent the vast majority, being up to 10–12% of this benign neoplasm of connective tissue [2]. There is an increased risk of malignant transformation in genetically susceptible patients such as patients with type 1 neurofibromatosis (NF1), resulting in a malignant peripheral nerve sheath tumor (MPNST), which can occur rarely at any nerve root including cranial nerves. Current literature reports about 40 cases in the English-written journals and periodicals, the most frequently affected cranial nerves are in decreasing order of frequency the trigeminal and vestibulocochlear – facial complex [1,3–5] and in one case the lower cranial nerves complex through jugular foramen in a pediatric patient [6].

The objective of this manuscript is to report for the first time in the English-written literature the case of an adult female patient with NF1 who presented a MPNST involving the glossopharyngeal, vagus and spinal left nerves which extended from the left cerebellopontine cistern to C4–C5 levels via the jugular foramen, along with this primary lesion the patient presented systemic metastases.

2. Case report

2.1. Clinical history, physical examination and image

A 37-year-old female presented with the following relevant prior medical history: NF1 diagnosed eight years ago, neurofibroma resection at first left metacarpophalangeal joint six years ago.

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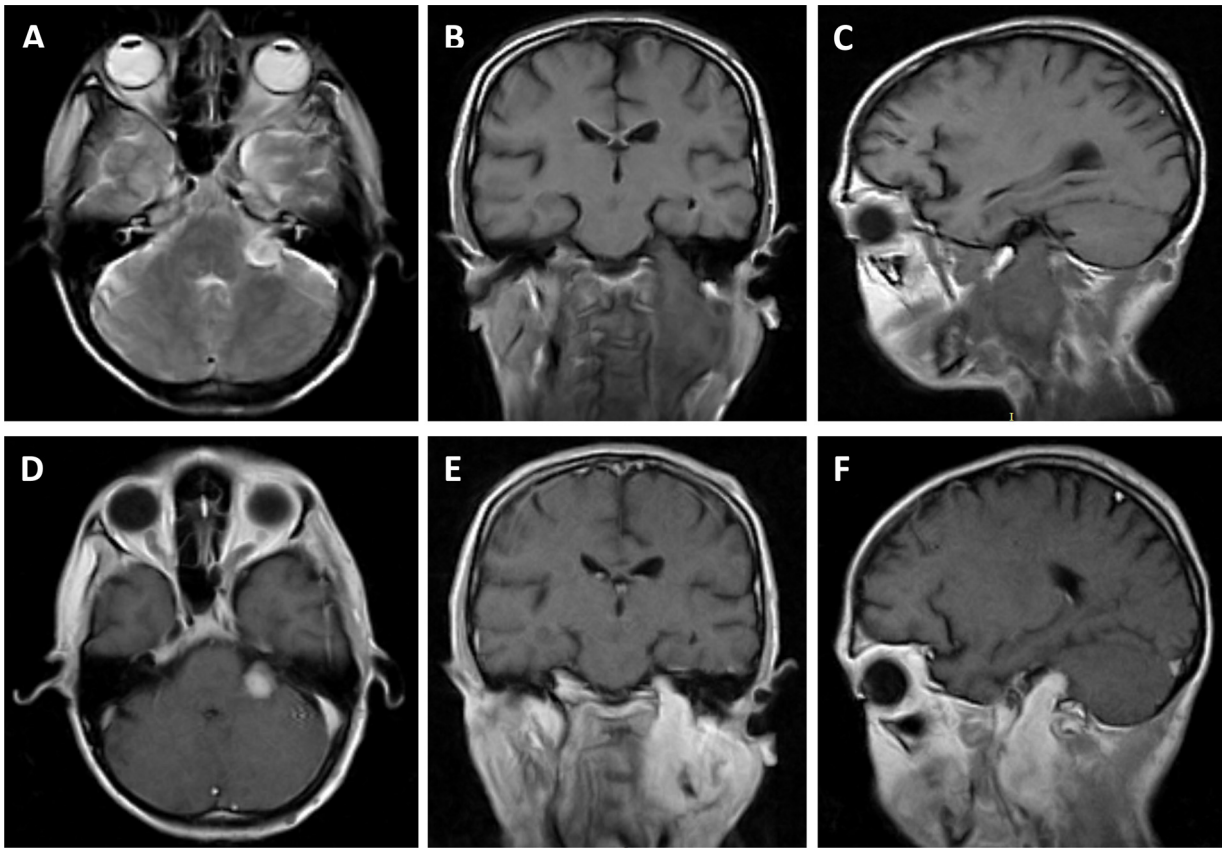

Fig. 1. Preoperative brain MRI showing a hyperintense neoplastic lesion on T2 sequence at left cerebellopontine cistern (A) with extension to C4-C5 level through jugular foramen (B and C), it enhances at gadolinium administration (D–F) and shows a low signal intensity in the center (E and F).

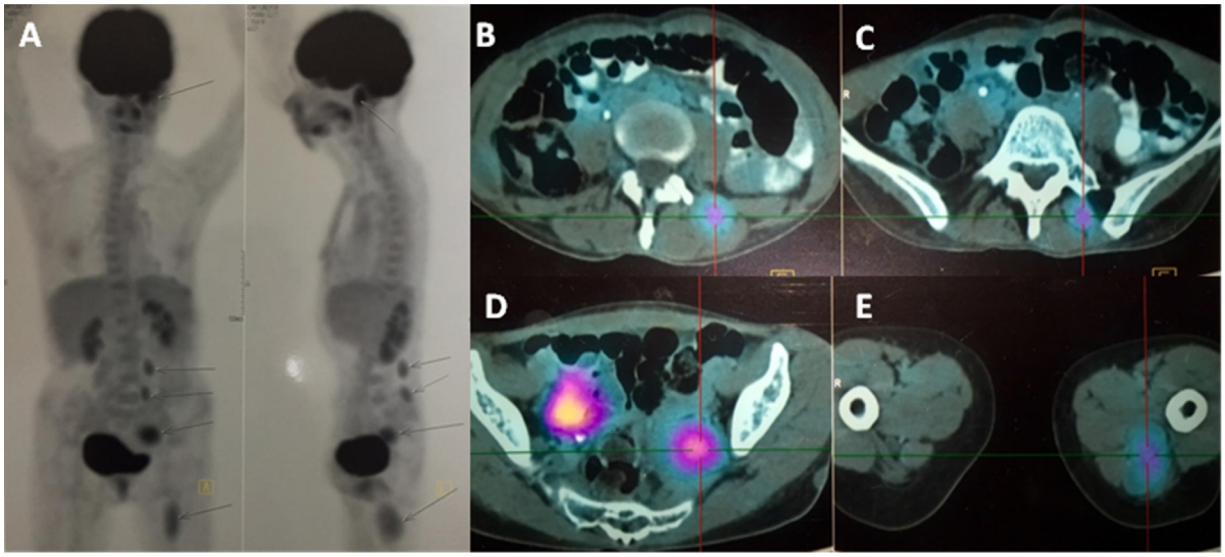

Fig. 2. PET-CT. Coronal, sagittal and axial sections showing metastatic lesions (arrows) at paravertebral (A–C), pelvic (A and D) and left thigh levels (A and E) with increased metabolism of 5-fluorodeoxyglucose.

The patient sought medical attention through his primary care physician (PCP) with a chief complaint of one-month history of hoarseness accompanied with nonproductive cough, she was treated with a 7-day antibiotic course with no successful results, reason why the PCP referred the patient to otolaryngology where she was diagnosed as chronic pharyngitis and put on a 2-month antibiotic course without improvement, further analysis of the case was done by the otolaryngologist, including laryngoscopy which revealed a left vocal cord paralysis. Magnetic resonance imaging (MRI) of the brain and cervical spine were ordered and the patient was ultimately referred to the neurology and neurosurgery department of our hospital.

At hospital admission she referred a history of fatigue, weakness, dysphagia to solids and weight loss of 8 kg in the last 6 months.