Case Report

Case of an Intracranial Malignant Peripheral Nerve Sheath Tumor in the Setting of Pacer-dependent Heart Block

Abstract

Intracranial malignant peripheral nerve sheath tumors (MPNSTs) are an extremely rare entity with only a handful of cases reported in the literature. MPNSTs typically occur in the extremities and the trunk. The treatment algorithm includes, when possible, gross-total resection as these tumors are extremely aggressive. When these tumors occur intracranially, they are termed malignant intracerebral nerve sheath tumors. The diagnosis hinges on immunohistochemistry and pathological features and often the diagnosis can be delayed for this reason. In this setting, it is critical to utilize intraoperative navigation, thus necessitating the use of fine-cut magnetic resonance imaging (MRI). This report presents a patient who presented with symptoms of obstructive hydrocephalus secondary to an intracranial mass. The patient underwent a full and extensive metastatic workup that was ultimately negative. To complicate things, the patient was fully pacemaker dependent. In this report, we review the literature surrounding this type of tumor, along with a detailed presentation of the case mentioned including the difficulties of cardiac pacing in the setting of MRI.

Keywords: *Magnetic resonance imaging compatible, malignant peripheral nerve sheath tumor, pacemaker*

Jonathan Chica, Isaac Yepes, S. Shelby Burks[1], Ricardo Komotar[1], Roger Carrillo

Department of Cardiothoracic Surgery, University of Miami Hospital, [1]Department of Neurological Surgery, The Miami Project to Cure Paralysis, University of Miami Miller School of Medicine, Miami, Florida 33136, USA

Introduction

Intracranial schwannomas account for approximately 8% of all brain tumors in adults. They most commonly arise from the vestibular portion of the eighth cranial nerve. There is considerably less knowledge regarding their malignant counterpart.[1,2] Malignant peripheral nerve sheath tumors (MPNSTs) typically originate from nerves of the extremities and trunk or from preexisting neurofibromas. Those that arise from brain parenchyma are termed malignant intracerebral nerve sheath tumors (MINST).[3] These are extremely rare with only a few cases reported in the literature. We report a unique case of a patient with spontaneous MINST with a brief literature review.

Case Report

A 38-year-old Hispanic female with a past medical history of Hodgkin lymphoma during childhood treated with both chemotherapy and radiation to the chest, and pacemaker placement 3 years ago due to third degree AV block thought to be secondary to radiation therapy presented with a 1-week history of headaches, dizziness, and dysarthria. She did not present any other neurological signs or symptoms, and physical examination was otherwise normal. A CT of the brain without contrast was performed in the ER, showing a questionable mass in the superior aspect of the cerebellum at midline, measuring approximately 2.8 cm × 4.3 cm in AP by transverse dimension with surrounding vasogenic edema and mild prominence of the temporal horns without a midline shift [Figure 1]. Since the patient had an absolute indication for the magnetic resonance imaging (MRI) scan to guide surgery and further treatment options, she required complete removal of her pacing system, which included both the pacemaker and the leads implanted. The patient was evaluated by a cardiologist and was found to be fully pacer dependent. The patient underwent removal of pacemaker, laser lead extraction of the atrial pacing lead and ventricular pacing lead, and implantation of MRI compatible leads and pacemaker [MRI compatible pacer system; Figure 2]. In addition, the patient underwent an extensive oncological workup with no primary cancer found that could explain the etiology of the mass.

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Address for correspondence:
Dr. Jonathan Chica, Department of Cardiothoracic Surgery, University of Miami Hospital, 1295 NW 14th Street, Suite H, Miami, Florida 33136, USA.
E-mail: jonathankchica@gmail.com

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After workup was completed, the patient was taken to surgery with the goal of biopsy with or without gross-total resection. A sitting position infratentorial supracerebellar approach with a suboccipital craniotomy for resection of the mass was performed. Stereotactic navigation was used to confirm the location of the tumor. After the intraoperative pathology report confirmed that it was not lymphoma but, in fact, a spindle cell tumor, gross-total resection was achieved [Figure 3], and an external ventricular drain was left open at 10 cm of water above the ear. The pathology reported MPNSTs, grade 3/3, associated with abundant hemorrhage. Immunohistochemistry revealed tumor cells that focally express S100, CD34, and desmin. The patient's postoperative course was uneventful and was discharged postoperative on day nine.

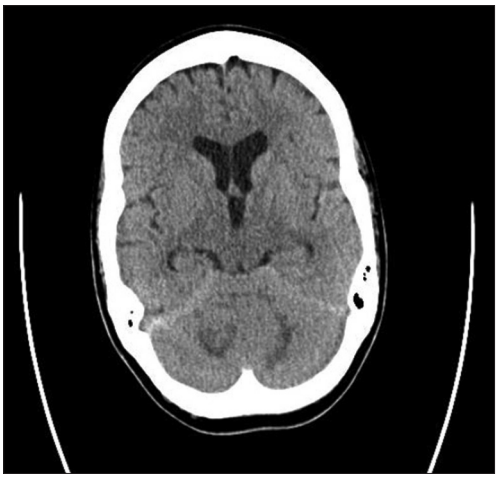

Figure 1: Original plain computed tomography scan obtained upon presentation demonstrating a mass located in the cerebellum with associated, obstructive hydrocephalus

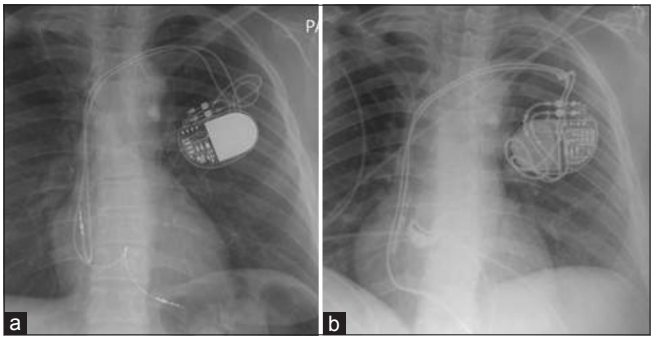

Figure 2: Posteroanterior chest radiograph demonstrating pacemaker. (a) Corresponds with nonmagnetic resonance imaging compatible device whereas (b) the new, magnetic resonance imaging compatible pacemaker in place

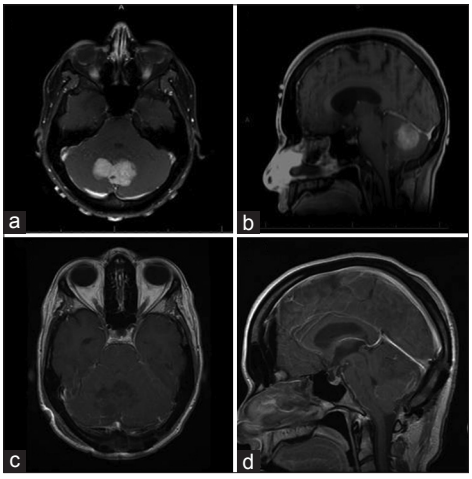

Figure 3: Demonstrating axial and sagittal pre-and post-operative imaging. All sections are postgadolinium contrast. (a and b) Representing preresection, whereas (c and d) are postresection

Discussion

Primary MPNST are extremely rare tumors. They are termed as MINST due to their histological, immunohistochemical, and ultrastructural similarities to MPNST but are distinguished by their intracerebral location.[4] Although some general characteristics of these tumors have been described, preoperative diagnosis is very challenging due to its rarity in the central nervous system and because of the absence of pathognomonic signs on radiologic images. Often, these challenges lead to a delay in proper treatment of the patient.[5] The cellular source of these tumors remains debatable, with some suggesting an origin from Schwann cells. Since Schwann cells do not normally exist within the central nervous system, others have suggested pluripotent mesenchymal cells.[3,6] In addition to patients with neurofibromatosis type 1, others predisposed to MINST include those with a history of ionizing radiation exposure,[3,7] such as our patient who had a history of childhood lymphoma that was treated with radiation. Baehring *et al*.[8] report that in 3 of their 54 patients studied, MPNST arose within the field of radiation therapy administered for previous malignancies (Wilms' tumor, Hodgkin's disease, chordoma) after an interval of nine to 36 years. This case becomes even more unique due to the history of radiation-induced heart block that required the patient to have a pacemaker implanted. Scenarios in which patients have noncompatible MRI devices can pose some problems when such imaging is needed for proper guidance and management. Noncompatible MRI pacemakers can have interactions with an external magnetic field. The most intuitive potential interaction of implanted devices with an external magnetic field is the possibility for movement and dislocation of the device. Current induction is the most feared potential interaction when conducting wires are placed in an MRI scanner. Another important potential interaction is the possibility of heating and tissue damage where the lead tip contacts tissues. Finally, implanted cardiac devices may provide unnecessary therapies or fail to provide necessary therapies when placed in the MRI scanner.[9] Because of this, it was imperative for the patient to have an MRI compatible pacer system. This allowed for the use of intraoperative, frameless navigation to enhance accuracy.

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Conflicts of interest

There are no conflicts of interest.

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