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Clinical case

Acute hydrocephalus due to a primary malignant peripheral nerve sheath tumor of the cervicothoracic junction: A case report and review of the literature

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ABSTRACT

Background. – The estimated incidence of malignant nerve sheath tumors is 0.001% per year, and only 2–3% of those tumors involve the spinal nerves. We present a rare case of acute hydrocephalus caused by primary malignant peripheral nerve sheath tumor of the cervicothoracic junction.
Case description. – A 29-year-old previously healthy male patient, except for a history of two previous surgeries for ulnar nerve entrapment and progressive left upper extremity weakness, presented with acute onset somnolence. The CT and MRI revealed hydrocephalus and periventricular edema. The patient underwent ventriculoperitoneal shunt surgery. Postoperative MRI of the spine revealed a $6 \times 3 \times 3$ cm intra-extradural lesion at C7-T1 level and multiple metastases in other spinal segments. The patient underwent combined surgical excision and the tumor was diagnosed as a malignant peripheral nerve sheath tumor based on pathological and immunohistological findings. Radiation therapy and chemotherapy were initiated.
Conclusion. – Primary malignant peripheral nerve sheath tumor of the spine is a very aggressive tumor with a very high recurrence rate, significant potential for metastasis and very poor overall prognosis. They may present with features of more frequent diseases, such as peripheral neuropathies and may be overlooked as in our case. Thus, suspected cases should undergo a more detailed examination.

1. Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are soft tissue sarcomas of an ectomesenchymal origin and arise from major or minor peripheral nerve branches or sheaths of peripheral nerve fibers [1]. Although large MPNSTs of the thoracic or abdominal cavity may exhibit secondary spinal involvement, primary MPNSTs of the spine are very rare [2].

In this report, we present a case of primary MPNST in a male patient causing acute hydrocephalus and discuss the problems involved in the management of this tumor in conjunction with a brief literature review.

2. Case report

A 29-year-old, previously healthy, male presented to the emergency department with acute onset somnolence. He was also vomiting and his pupils dilated to 5 mm. Emergent computed tomography (CT) and magnetic resonance imaging (MRI) of the brain revealed an acute hydrocephalus (Fig. 1). Lumbar puncture opening pressure was 18 cmH_2O. Cerebrospinal fluid (CSF) analysis revealed transparent color with no coagulum, glucose level 58 (normal range: 40–80 mg/dL), protein 32 (normal range: 15–60 mg/dL) and cell count 2 lymphocytes. Lumbar drainage did not improve the symptoms. Therefore, emergent ventriculoperitoneal shunt insertion was performed. Surgery was uneventful.

The first postoperative neurological and clinical examination revealed that the patient had been suffering from gradually worsening left upper limb weakness during a one-year period and had undergone two left ulnar nerve decompression surgeries with a diagnosis of ulnar nerve entrapment. His medical and family histories were unremarkable in terms of any genetic disorders,

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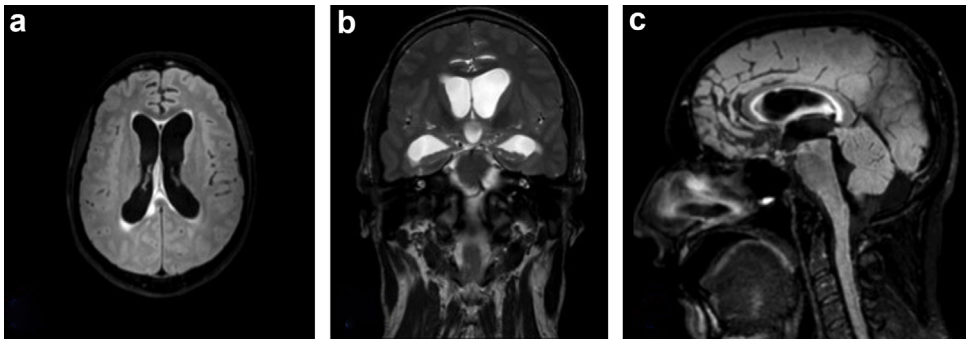

Fig. 1. Axial FLAIR (a), coronal T2-weighted (b) and sagittal FLAIR (c) magnetic resonance images showing ventricular dilatation and periventricular edema.

systemic diseases, medication or radiotherapies. The patient had no cutaneous stigmata of neurofibromatosis. The neurological examination revealed impaired finger extension, elbow extension, wrist flexion, flexion of the distal phalanges of the fourth and fifth digits, finger abduction and finger adduction associated with sensory loss in the middle finger and ulnar distribution of the left hand and forearm. The deep tendon reflexes of the biceps and brachioradialis muscles were all intact, but the triceps reflex was considered to be hypoactive. No other sensory abnormalities or muscle weaknesses were detected in other extremities. Laboratory values were within normal limits.

As regards the neurological examination, a lesion affecting the middle trunk and mainly the lower trunk was suspected. As weakness and myatrophy of the intrinsic muscles of the hand are also seen in Pancoast-Tobias syndrome, a CT scan of the chest was performed and the diagnosis of a pancoast tumor was excluded. The MRI of the spine revealed a lobulated intra-extradural mass lesion ($6 \times 3 \times 3$ cm) at C7-T1 neural foramen, which extended into the paravertebral costoclavicular area and causing pressure on left brachial plexus. The lesion showed heterogeneous enhancement after gadolinium administration (Fig. 2). Staging workup was negative for extraspinal metastases.

Although the spinal lesion was radiologically reported as a benign peripheral nerve sheath tumor, surgical removal was planned, as malignancy could not have been ruled out. Following laminectomy of C7 and T1 vertebras, a longitudinal incision was made on the left side of the exposed dura. A fleshy, tan-white mass was observed extending into the foramen. Intraspinal part was gross-totally resected for decompression and diagnosis. An anterior approach was performed for the paraspinal component during the same session. Near total excision of paraspinal component was achieved (Fig. 3).

Microscopic examination of the surgical specimen revealed spindle cells arranged in a whorling pattern with irregular nuclei, cyst formation, and nuclear palisading with hypercellularity. The specimen showed immunohistochemical positivity for S-100 (Fig. 4). These findings confirmed the diagnosis of MPNST.

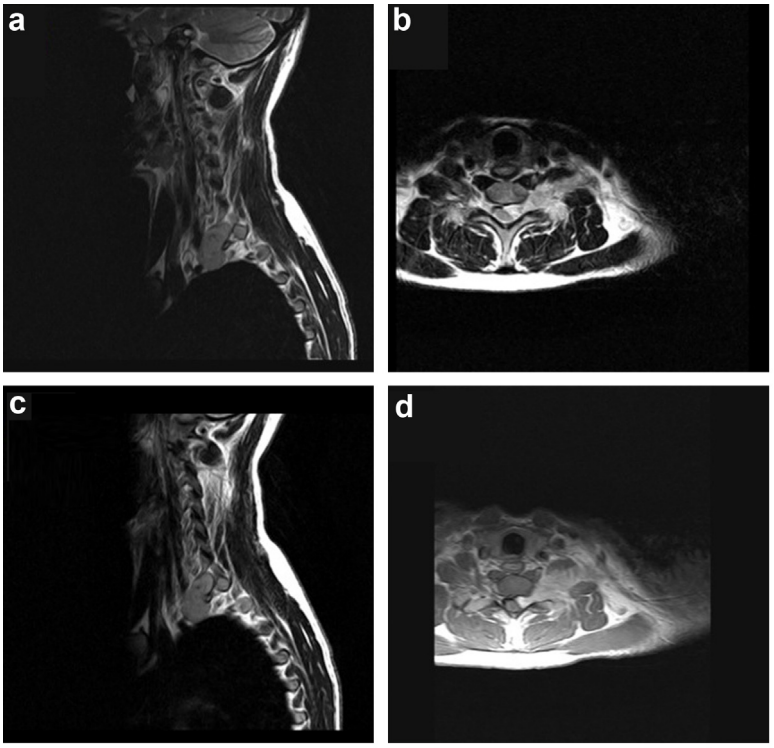

Fig. 2. Pre-operative magnetic resonance images of brachial plexus. Sagittal (a) and axial T2-weighted (b) images showing a lobulated intra-extradural mass lesion at C7-T1 neural foramen, extending into paravertebral costoclavicular area and causing pressure on left brachial plexus. Enhanced sagittal T2-weighted (c) and axial T1-weighted (d) images showing heterogeneous enhancement.