

Weber's Syndrome Due to Low-Grade Astrocytoma

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Abstract

Weber's syndrome is a classic midbrain stroke syndrome. We report an unusual presentation of Weber's syndrome caused by an astrocytoma (WHO grade II) and review the pertinent literature. To date, this represents with first case report of Weber's syndrome due to an astrocytoma.

Keywords

Weber's syndrome, Astrocytoma, Glioma, Tumor, Midbrain

Introduction

The midbrain comprises the most rostral portion of the brainstem. Within a small area, it contains the superior and inferior colliculi dorsally (tectal region) and the red nucleus, substantia nigra, oculomotor and trochlear nucleus more ventrally (tegmental region). Additionally, white matter tracts also span the midbrain such as the superior cerebellar peduncle, medial and lateral lemniscus, spinothalamic tracts and the motor tracts within the cerebral peduncles. The vascular supply comes from the posterior circulation, specifically from the basilar, superior cerebellar and posterior cerebral arteries [1]. Due to the complex nature of midbrain anatomy, pathology at this location can produce a wide variety of clinical syndromes.

The five classically described midbrain syndromes are Weber's, Claude's, Benedikt's, Parinaud's and Nothnagel's syndromes. Of these syndromes, the former three involve the tegmentum and the

latter two, the tectum. Weber's syndrome is characterized by a ventromedial midbrain lesion that affects both the oculomotor nerve and cerebral peduncle, resulting in an ipsilateral oculomotor nerve palsy with contralateral hemiparesis. It was first described by Sir Hermann David Weber in 1863 and is usually due to an infarct within the vascular territory of the paramedian branches of either the basilar artery or the posterior cerebral artery [1,2].

Interestingly, despite being highly regarded as a stroke syndrome, the incidence of stroke causing Weber's syndrome is actually quite rare [3]. There are even fewer reported cases in the literature describing alternative causes of Weber's syndrome. We present a case of Weber's syndrome that is due to a thalamo-mesencephalic astrocytoma. To our knowledge Weber's syndrome due to a glial cell tumor has not yet been described.

Case Presentation

A previously healthy 50-year old male presented with rapidly, progressive left sided upper and lower extremity hemiparesis, hemi-sensory loss and diplopia. Neurologic examination revealed an alert individual with normal pupillary responses, right ptosis, paresis of vertical and medial gaze of the right eye, approximately grade 4/5 strength of the left upper and lower extremities, and diminished pin prick sensation over the left face and left upper and lower extremities. Magnetic resonance imaging of the brain revealed a suspicious non-enhancing mass in the right thalamus and midbrain (Figure 1A, 1B). A stereotactic brain biopsy was performed. The pathologic examination demonstrated a hypercellular astrocytic neoplasm with cytological atypia that was without mitoses, endothelial hyperplasia, and necrosis, which was consistent with a diffuse astrocytoma (WHO Grade II). Co-deletion status of 1p19q was unable to be tested due to the paucity of tissue. There was sufficient tissue for isocitrate dehydrogenase 1 testing and it was not mutated by immunohistochemistry testing. The patient was subsequently treated with temozolomide (150 mg/m²) once daily for five consecutive days of a 28-day treatment cycle. By the completion of the first cycle he had clinical and radiographic progression and therefore therapy was escalated to receive o daily temozolomide (75mg/m²) plus radiation and bevacizumab 10mg/kg every two weeks.

Discussion

This case highlights an astrocytoma manifesting clinically as Weber's syndrome. To our knowledge, this is the first documented case of Weber's syndrome caused by a glial tumor. Currently, there is one previous

case report of Weber's syndrome as a result of a primary central nervous system lymphoma (diffuse large B cell) [4]. In comparison to our patient, the patient with CNS lymphoma also presented with rapid onset of hemiparesis. Initially the diagnosis was not clear and the patient was treated with IV dexamethasone and had gradual improvement. However, after steroid cessation his symptoms recurred and he additionally developed a contralateral oculomotor nerve palsy. Following biopsy, diagnosis and proper treatment, there was complete resolution of his symptoms [4]. In contrast, our patient's neurologic deficits have not improved despite initial chemotherapy.

Brainstem gliomas account for only 1-2% of all intracranial gliomas in adults [5]. This may partially account for why glial tumors rarely manifest as Weber's syndrome. To our knowledge, the explanation for this decreased posterior fossa predilection in the adult brain tumor population is not understood. Moreover, most descriptions of mesencephalic glial tumors involve the tectal region rather than the tegmentum, and thus it would make sense that tectal syndromes are more frequently associated with brain tumors [6]. Finally, an additional explanation why brain tumors rarely present as Weber's syndrome may be because brain tumors are not restricted to a particular vascular territory, unlike ischemic stroke.

In conclusion, this case provides an additional example of a brain tumor mimicking a traditional midbrain stroke. It serves as a reminder to cast a broader differential diagnosis when Weber's syndrome is encountered.

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Figures

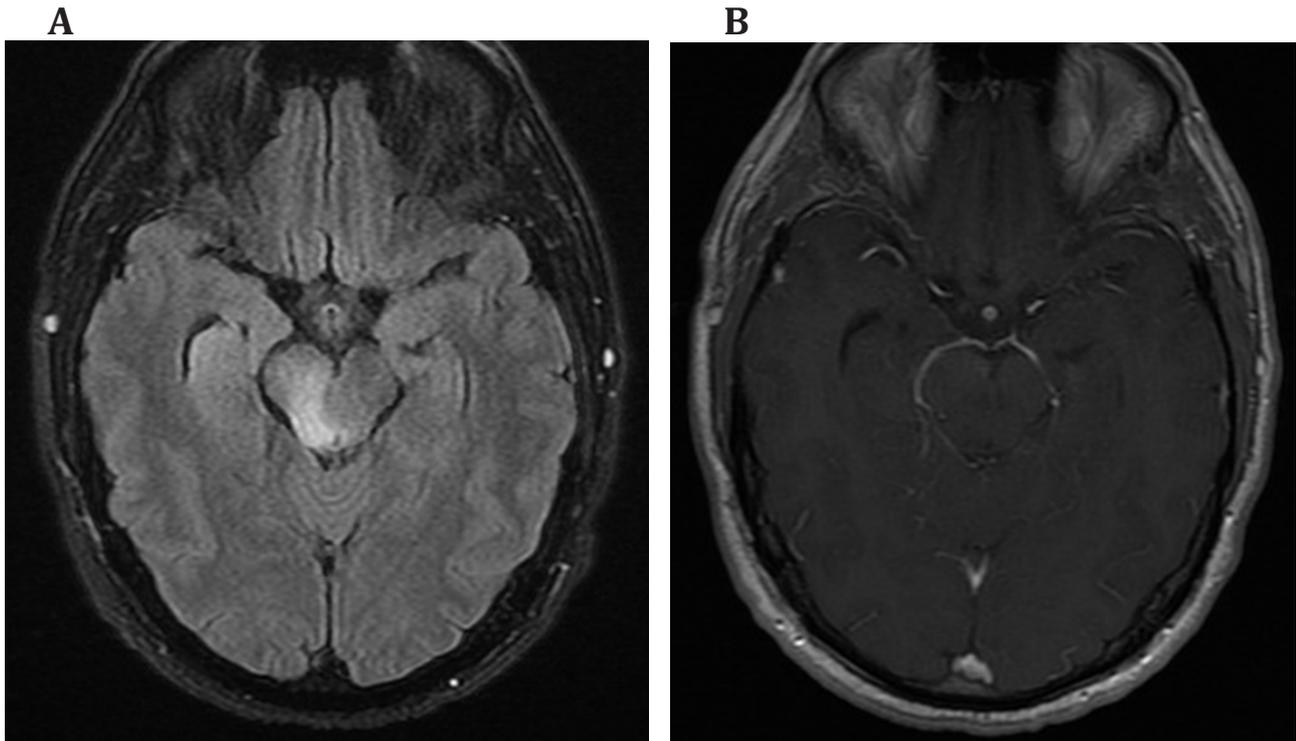


Figure 1: Non enhancing right mesencephalic mass prior to biopsy. (A): Axial fluid-attenuated inversion recovery (FLAIR) MRI showing hyperintense mass in right midbrain. Pathology was consistent with an astrocytoma (WHO grade II). (B): Axial T1 MRI with gadolinium demonstrating no enhancement of mesencephalic mass.

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