Case Report

Distinguishing Weber from Pseudo Weber Syndrome: A Diagnostic Dilemma- A Case Report

Ahmad Akhtar Rashid[1], Hamza Khan [2], Akhtar Rashid [3], Rehan Gill [4]

[1]Third year MBBS Student, Sargodha Medical College
[2]Neuro Physician, DHQ Hospital Multan
[3]Consultant Physician, Al-Rashid Hospital Sargodha
[4]Radiologist, DHQ Hospital Sargodha

Author's Contribution
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[2,3] *Critical Review*
[4] *Analysis/Interpretation/Discussion*

Corresponding Author
Mr. Ahmad Akhtar Rashid,
Third year MBBS Studemt, Sargodha Medical College, Sargodha
Email: trino.aar@gmail.com

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Abstract

Introduction: Weber Syndrome is typically characterized by ipsilateral oculomotor nerve palsy and contralateral hemiparesis, commonly resulting from mesencephalon (midbrain) lesions. However, we present a unique case of a patient exhibiting classic Weber Syndrome symptoms that were ultimately attributed to distinct underlying causes, leading to a diagnosis of Pseudo-Weber Syndrome.

Case Presentation: The patient presented with left-sided hemiparesis and upper motor neuron facial paralysis, initially suggesting a clinical diagnosis of Weber Syndrome. Further radiological investigation revealed a left middle cerebral artery (MCA) infarct with internal capsule involvement as the primary cause of the hemiparesis and facial paralysis involving the contralateral lower half of the face. Additionally, the oculomotor nerve symptoms were traced to uncal herniation, rather than a mesencephalon(midbrain) lesion.

Discussion: This case underscores the importance of thorough radiological assessment in patients with Weber Syndrome-like presentations, highlighting the diagnostic challenge in distinguishing Weber from Pseudo-Weber Syndrome. Understanding the varied etiologies behind similar clinical presentations is crucial for accurate diagnosis and appropriate management.

Keywords: Weber Syndrome; Pseudo-Weber Syndrome; Oculomotor nerve palsy; Hemiparesis; Facial paralysis; Middle cerebral artery infarct; Internal capsule; Uncal herniation; mesencephalon lesion.

Introduction

Weber syndrome or superior alternating hemiplegia is a neurological condition characterized by ipsilateral oculomotor nerve palsy, contralateral hemiparesis and upper motor neuron cranial nerve palsies (most commonly facial nerve) typically resulting from lesions in the mesencephalon(midbrain).[1] This syndrome is well documented in clinical neurology with its hallmark features aiding in prompt diagnosis and treatment.

“Pseudo-weber" Syndrome, on the other hand, presents with the same symptoms as Weber syndrome but is a combination of different underlying mechanisms which can be a result of two different strokes, one in the midbrain and the other in the internal capsule, corona radiata or globus pallidus.[2] It can however also present itself after a large cortical infarct involving the anterior circulation. Infarct involving the anterior circulation also affects the corticobulbar fibers traversing in the internal capsule especially the genu of the internal capsule and can affect the fibers of cranial nerves traversing through it especially the corticobulbar fibers of the facial nerve.[3,4]

Anterior circulation strokes also produce contralateral hemiparesis either by affecting the primary motor cortex located in the precentral gyrus of the frontal lobe or by affecting the motor fibers traversing in the internal capsule’s posterior limb.[5]

About 10 per cent of ischemic strokes especially those involving the M1 part of the middle cerebral artery or the entirety of the middle cerebral artery can progress to malignant infarcts and cause mass effects.[6] One of the consequences of these mass effects is uncal herniation. Uncal herniation or trans tentorial herniation most often occurs after TBI, intracranial tumors and most commonly after intracerebral hemorrhage but can also occur as a result of mass effect after stroke involving MCA. One of the consequences of uncal herniation is impingement on the ipsilateral cerebral peduncle causing compression of the third nerve manifesting as ptosis, dilated pupil and a characteristic down and out lesion of the eye.[7]

The culmination of all these effects results in a clinical picture like Weber syndrome but underlying pathology and the temporal association of different symptoms make it an entirely different syndrome when compared to Weber. The only confirmatory test is radio imaging.

Case presentation

We present the case of a 40-year-old female patient who had a prior history of hypertension and type 2 Diabetes present in the emergency department in loss of consciousness. On examination, her left-sided pupil was dilated with concomitant ptosis and a down-and-out lesion of the left eye. There was also a loss of power in the right half of her body. Immediate treatment with intravenous mannitol started to relieve pressure from her brain. After the infusion of mannitol, her consciousness improved but she was exhibiting dysphasia. On clinical examination, a diagnosis of Weber syndrome was made considering her symptoms. A CT scan was ordered which was unremarkable and showed no lesion or infarct. An MRI scan was then ordered which showed no lesion or infarct of the midbrain but showed a massive left frontoparietal infarct in the MCA territory with midline shift with uncal herniation and involvement of the left internal capsule.

Later her attendant revealed that she developed hemiparesis in the right half of her body a few days before her loss of consciousness and pupillary involvement with ptosis (figure 1). Furthermore, on examination, she also had developed a left UMN lesion of the facial nerve (figure 2). These temporally different occurring symptoms although as a whole mimic Weber syndrome is not Weber syndrome. These are the results of her stroke which probably occurred given her history of diabetes and hypertension.[8,9] Her hemiparesis and facial nerve palsy were explained by the infarct of the left cortical area supplied by MCA and the infarct of the internal capsule. (figures 3). The third nerve involvement is explained by the uncal herniation which occurred as the consequence of the large stroke (figure 4).

She was started on the dual therapy of aspirin and clopidogrel.[10] Carotid Doppler and Holter monitoring ruled out any extracranial or thromboembolic cause arising from the heart (as a result of arrhythmia) or carotid arteries as a result of an embolus or any type of vasculitis.

She later returned for a follow-up after being discharged in about a week, where her symptoms didn't improve but she was maintaining consciousness. About two and a half months later, on her fourth follow-up, her ptosis improved, but the power in her affected limbs and facial palsy was the same.

Figure 1: Patient showing mild ptosis and slight down and out pupil on follow up.

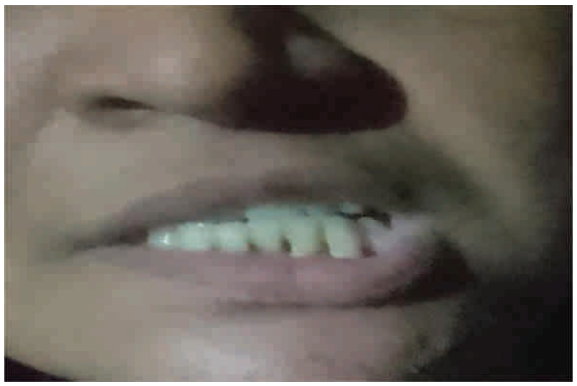

Figure 2: UMN facial palsy persisting on follow up-deviation of angle of mouth towards the unaffected side.

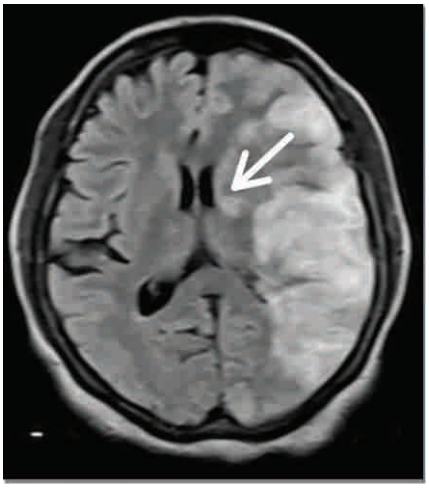

Figure 3: Hyperintensity present in internal capsule on FLAIR with a massive MCA stroke.

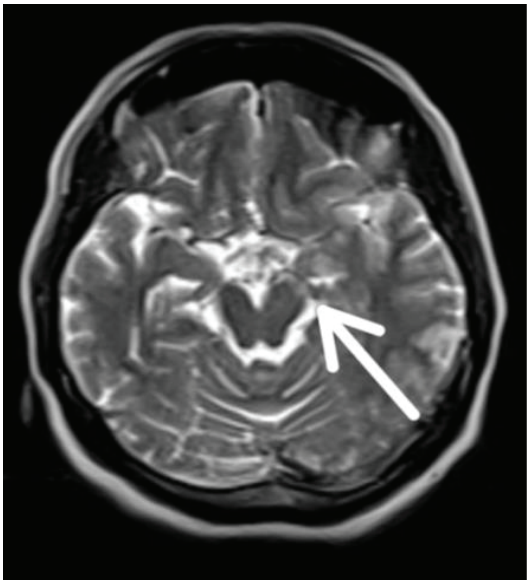

Figure 4: Uncal herniation on T2 axial showing compression on left peduncle of midbrain.

Discussion

We demonstrated a case of pseudo-Weber syndrome-essentially a culmination of different underlying pathologies which give the signs and symptoms of Weber syndrome. Clinicians on purely clinical grounds may label it as a pure Weber syndrome, and credit to them as any meticulous clinician would label it as Weber based on clinical evaluation.

The differentiating factor here is the temporally different manifestations of hemiparesis and OMN palsy which occur simultaneously in the case of Weber syndrome added to the loss of consciousness and the conflicting appearance on the MRI this syndrome is labelled radiologically and realistically as pseudo-weber syndrome. Although not a documented disease in literature at all nor a true disease in itself it is simply our own brain's way of tricking us into thinking otherwise. One must be meticulous and vigilant when diagnosing such cases taking into account the onset of the symptoms and the appearance on MRI.

Conclusion

This case report highlights the diagnostic complexity in patients presenting with Weber Syndrome-like symptoms. Through comprehensive radiological evaluation, the underlying causes were identified as a left middle cerebral artery infarct with internal capsule involvement and uncal herniation. This led to the diagnosis of Pseudo-Weber Syndrome rather than a midbrain lesion. The case underscores the necessity of thorough diagnostic workups to differentiate between similar clinical syndromes and ensure accurate diagnosis and appropriate management. Additionally, recognizing the importance of different temporal manifestations of symptoms is crucial in distinguishing Weber Syndrome from Pseudo-Weber Syndrome. Understanding the diverse etiologies and their temporal patterns that can present with similar neurological symptoms is essential for physicians to provide optimal care.

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