

Weber syndrome – clinical-imaging correlations

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ABSTRACT

An 89 years-old female patient was brought in by the family for altered mental status and left-sided motor deficit. Medical history revealed high blood pressure for which the patient was receiving anti-hypertensive treatment. Upon examination, the patient presented altered mental status, dysarthria, dysphagia, left central facial palsy, ataxic tetraparesis with marked paralysis on the left side, bilateral Babinski signs, right-sided ophthalmoplegia with limitation of gaze in all directions, left-sided skew deviation, limited abduction and upgaze movements of the left eye, and bilateral ptosis (complete on the right side and incomplete on the left side). The clinical picture was thus compatible with a vertebrobasilar stroke. A possible diagnosis of Weber syndrome was suggested, given the left-sided hemiparesis and right-sided ophthalmoplegia. Both head CT and brain MRI confirmed this by demonstrating a right-sided thalamo-mesencephalic subacute ischemic stroke. No cardio-embolic source was identified on cardiac assessment. Consequently, the patient was discharged with antiplatelet therapy, statin and anti-hypertensive treatment. We provide a short review on Weber syndrome, emphasizing the correlations between the clinical pictures and imaging findings.

Keywords: Weber syndrome, crossed paralyzes, alternating hemiplegia

Abbreviations (in alphabetical order):

ADC	Apparent Diffusion Coefficient
CT	Computer Tomography
DWI	Diffusion-Weighted Imaging
ECG	Electrocardiogram
FLAIR	Fluid Attenuated Inversion Recovery
MRC	Medical Research Council
MRI	Magnetic Resonance Imaging

INTRODUCTION

Weber syndrome is determined by a lesion of the midbrain, which damages the corticospinal tract and adjacent rootlets of the oculomotor nerve. Consequently, the patient will present with contralateral paralysis of the face, arm, and leg, but with ipsilateral ophthalmoplegia (or ipsilateral third nerve palsy).

Weber syndrome is included in a group of clinical syndromes which are characterized by *contralateral* hemiparesis and *ipsilateral motor deficits of cranial nerves*. These syndromes are known as “crossed paralyzes” or “alternating hemiplegia” and are very helpful in localizing the lesion to a specific part of the brainstem [1,2].

CASE PRESENTATION

Our patient is an 89 years-old female, who was brought in by the family for altered mental status and left-sided motor deficit. Medical history revealed high blood pressure for which the patient was receiving anti-hypertensive treatment.

Neurological exam performed by the on-call neurologist highlighted altered mental status (expressed as somnolence and confusion), dysarthria, dysphagia, Parinaud syndrome with right-sided mydriasis, left central facial palsy, ataxic tetraparesis with slight deficit on the right side (4/5 MRC), but marked paralysis on the left side (1-2/5 MRC), and bilateral Babinski signs. Blood pressure was 248/110 mmHg, and the heart rate was 85 bpm. Subsequent assess-

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FIGURE 1. Right-sided complete ptosis. Slight left-sided ptosis. Upon elevation of the superior eyelid, the right eye appears deviated towards the right side. There is also anisocoria (right-sided mydriasis)

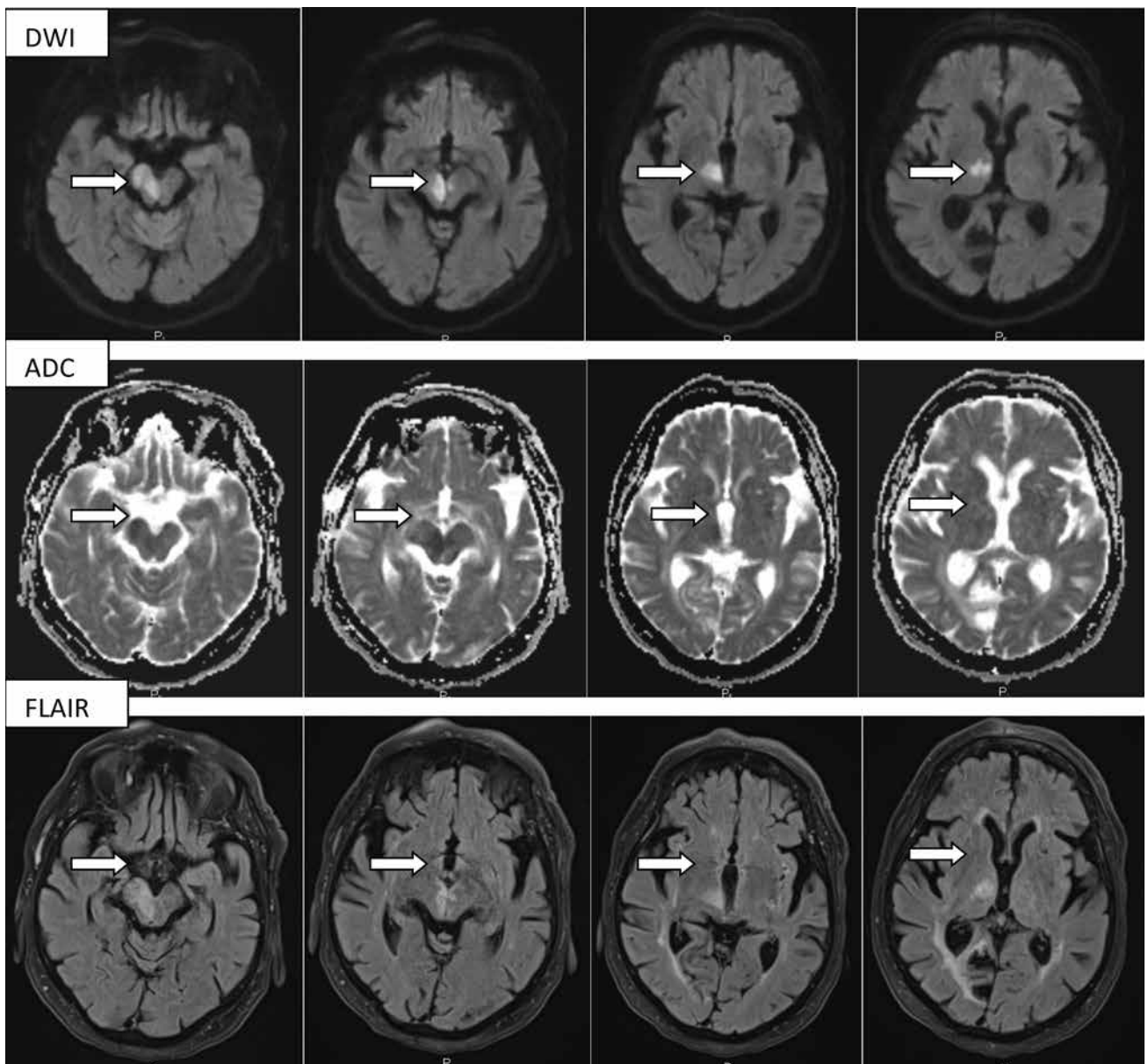


FIGURE 2. Brain MRI. Axial DWI, axial ADC and axial FLAIR sequences. (White arrows) Acute ischemic lesion involving the right-sided midbrain and ipsilateral thalamus. The lesion shows diffusion restriction (high DWI signal with low ADC signal) and appears hyperintense on FLAIR

ments performed by other neurologists revealed right-sided ophthalmoplegia with limitation of gaze in all directions, but also left-sided skew deviation, limited abduction and upgaze movements of the left eye, and bilateral ptosis (complete on the right side and incomplete on the left side) (Figure 1).

A possible diagnosis of Weber syndrome was suggested, given the left-sided hemiparesis and right-sided ophthalmoplegia.

Both head CT and brain MRI were performed and demonstrated a right-sided thalamo-mesencephalic subacute ischemic stroke (Figure 2).

The patient had a normal ECG. Cervical Doppler ultrasound indicated bilateral carotid atherosclerosis, without hemodynamically significant stenosis. Abnormal blood parameters included elevated homocysteine, elevated D-dimers, and positive lupus anticoagulant. On transthoracic echocardiography the patient presented aortic disease (with large stenosis and moderate regurgitation), aortic atheromatosis and possible thoracic aortic aneurysm.

During hospital stay, the patient received antiplatelet treatment with Aspirin 150 mg qd, statin therapy with Atorvastatin 40 mg qd, anti-hypertensive treatment (Amlodipine 10 mg qd, Metoprolol 25 mg bid and Olmesartan/Amlodipin 40 mg/5 mg bid), Vitamin B1 (100 mg bid), Vitamin B6 (50 mg bid) and Cerebroslyn 10 mL bid. Blood pressure values normalized. Kinesitherapy was started, however the patient remained bedridden, with left-sided hemiparesis 1-2/5 MRC, dysarthria, right-sided third oculomotor palsy, and limitation of upward gaze and abduction movements with the left eye.

Since no clear cardioembolic source was identified, the patient was discharged with antiplatelet treatment (Aspirin 100 mg qd). Additional recommended medications included Atorvastatin 40 mg qd, Metoprolol 25 mg bid, Olmesartan/Amlodipine 40 mg/5 mg bid and Vinpocetinum 10 mg tid. Details about the secondary prevention of cardiovascular risk factors were explained to the family, with emphasis on blood pressure monitoring and control.

DISCUSSIONS

Weber syndrome, also known as *superior alternating hemiplegia*, was first described by the German-born English physician Hermann Weber in 1863 [3,4]. It is usually caused by occlusion of the paramedian branches of the basilar or posterior cerebral arteries, leading to infarction in the ventromedial midbrain (oculomotor nucleus and cerebral crus)[5]. Consequently, it manifests as ipsilateral third nerve palsy and contralateral hemiplegia [6].

If the substantia nigra is also involved, there can be contralateral parkinsonian rigidity as well.

The frequency of Weber syndrome appears to be low, as only 5 cases were described in a series of 4816 patients with cerebrovascular disease (corresponding to a prevalence of 0.1%) [7]. While classically described as ischemic in origin, Weber syndrome is not pathognomonic for ischemic stroke, and can also be caused by other lesions, such as midbrain hemorrhage [8].

The oculomotor dysfunction can manifest either as complete oculomotor paralysis with mydriasis, or there may be pupillary sparing. Rarely, Weber syndrome can also present with supranuclear gaze palsy, if there is involvement of the ipsilateral rostral interstitial nucleus of the medial longitudinal fasciculus – which is considered the supranuclear control center of the vertical gaze [9,10]. This is usually seen in patients with thalamo-mesencephalic strokes, as was the case of our patient.

The differential diagnosis of Weber syndrome includes Benedikt syndrome, also known as *paramedian midbrain syndrome*. Benedikt syndrome is very similar to Weber, but additionally involves the red nucleus, and consists of (1) ipsilateral oculomotor nerve palsy, (2) contralateral hemiparesis and (3) contralateral cerebellar ataxia/Holmes tremor and/or contralateral choreo-athetosis [11].

Weber syndrome due to ischemic stroke can be caused by cardiac embolism, in situ thrombosis, small vessel disease (of penetrating branches) or artery-to-artery embolism. It is important to note that midbrain infarctions are 10 times more likely to associate infarction of adjacent structures than to occur in isolation [12].

Of interest is a recent paper by Marx et al, who analyzed 308 patients with acute brainstem infarctions, in order to determine the prevalence of the classical crossed brainstem syndromes. Apart from Wallenberg syndrome (14 patients), all other classical crossed syndromes were identified in only 1 or 2 cases, or even not at all. Only one case showed Weber's syndrome. However, more than 20% of patients presented with "different, so far unnamed crossed symptom combinations" [13].

CONCLUSIONS

Although rare, Weber syndrome is one of the classical crossed brainstem syndromes. A careful neurological examination is recommended in order to determine the presence of other brainstem signs, and this should be correlated with brain imaging (especially MRI) for identifying the exact location of the lesion.

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