Weber’s Syndrome with Vertical Gaze Palsy
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INTRODUCTION
Weber’s syndrome is a fascicular syndrome of the oculomotor nerve, commonly caused by the occlusion of the blood vessels that supply the midbrain and results in necrosis of midbrain tissue, involving oculomotor nerve fascicle and the cerebral peduncles. There is resultant third nerve palsy/paresis and contra lateral hemiplegia. Sir Herman David Weber described the first case of Weber’s syndrome, in the 19th century.1 Since then, many such cases have been reported, but cases of Weber’s syndrome with vertical gaze palsy are rare.

CASE REPORT
In April 2007, a 47-year-old female presented in the emergency department with sudden onset of weakness on right side of the body including facial weakness. There was outward deviation of her left eye and an inability to look in up and down gaze. The patient was fully conscious and well-oriented in time and space.

Her best-corrected visual acuity was 6/12 in both eyes. Intraocular pressures were normal. Slit-lamp biomicroscopy showed no abnormality. On fundus examination, there was non-proliferative diabetic retinopathy in both eyes.

On gross examination, she had left exotropia with mild hypertropia. There was mild ptosis in the left eye with compensatory lid retraction in the right eye. She had anisocoria, which increased in bright illumination. Size of pupil in moderate illumination was 3 mm and 4 mm; in right and left eye respectively. In bright illumination, it was 2 mm in right eye and 4 mm in left eye. Extraocular movements showed restriction of vertical gaze associated with left medial rectus paresis and inferior rectus palsy. Convergence could not be appreciated because of left medial rectus restriction. Bielschowsky three-step test revealed hypertropia due to left inferior rectus palsy. Visual fields were normal.

Systemically, she had uncontrolled hypertension and diabetes. Neurological examination revealed right hemiplegia, while sensory and cerebellar systems were intact. MRI was performed which showed multiple T1W hypointense (Figure 1) and T2W hyperintense foci in both thalami extending downwards into the left side of the midbrain (Figure 2 and 3). These areas indicate midbrain and thalamic infarcts.

The patient was managed with the joint involvement of the medical, neurological and medical departments. Her blood pressure and blood sugar was controlled and ocular examination including fundus examination for diabetic retinopathy was suggested after every 3 months. At her first follow-up, there was slight improvement in extraocular movements but no improvement in the vertical gaze palsy. She had been on physiotherapy for hemiplegia with extremely slow recovery.

DISCUSSION
The midbrain comprises of two lateral halves called cerebral peduncles.2 Each of these is further divided into an anterior part, the crus cerebri and a posterior part, the tegmentum. The tectum is the part of the tegmentum posterior to the cerebral aqueduct. The oculomotor nucleus is situated in the central grey matter close to the median plane, just posterior to the medial longitudinal fasciculus. The fibres of the oculomotor nucleus pass anteriorly through the red nucleus to emerge on the medial side of the crus cerebri. Oculomotor palsies at the level of the midbrain are either nuclear or fascicular. For a nuclear palsy, there should be bilateral ptosis and contralateral superior rectus involvement. As the ptosis in this patient was unilateral, nuclear lesion was excluded. Fascicular lesions of the third nerve produce symptoms and signs that are strictly ipsilateral. The common causes of fascicular oculomotor palsy are

ABSTRACT
Weber’s syndrome with vertical gaze palsy is rarely reported in literature. We present a case of a 47-year-old female who developed sudden onset of left exotropia, right sided hemiplegia and vertical gaze palsy. Magnetic resonance imaging (MRI) showed multiple infarcts involving both thalami and extending caudally into the midbrain. This case presents the diverse clinical picture following midbrain infarcts.

infarction, demyelination and tumours. In vascular causes, the occlusion of branches of the posterior cerebral artery in the most common. Due to the relative anatomic separation of the third nerve fascicle within the midbrain, fascicular lesions often cause partial rather than complete third nerve palsy as seen in this case. It is said that fascicular lesions are best recognised “by the company they keep”. In this context, many classical syndromes of third nerve palsy have been are described; Benedict’s (with contra lateral ataxia and intention tremor), Weber’s (with contra lateral hemiplegia), Nothnagel (with cerebellar ataxia) and Claude (with characteristics of both Nothnagel and Benedict’s). Weber’s syndrome, as seen in this case, occurs when there is damage to the oculomotor fascicle, along with the crus cerebri.

Association of Weber’s syndrome with vertical gaze palsy can be explained by the fact that vertical eye movements are generated in the rostral interstitial nucleus of the medial longitudinal fasciculus which also lies in the midbrain, dorsal to the red nucleus. Dorsal midbrain syndrome and Steele Richardson Olszewski syndrome are two important syndromes of vertical gaze palsy. In dorsal midbrain syndrome, there is light near dissociation, lid retraction, convergence retraction nystagmus and large pupils associated with vertical gaze palsy. In Steele Richardson syndrome, there is vertical gaze palsy, which later on results in globe palsy, extra pyramidal rigidity and gait ataxia. All these signs were absent in this patient who had vertical gaze palsy with Weber’s syndrome.

According to Bogousslavsky et al., there are three types of midbrain infarcts; upper, middle and lower. Upper dorsal midbrain infarcts result in vertical gaze disturbances. Middle infarcts involve the third nerve palsy with contralateral hemiplegia. Inferior infarcts are associated with fourth and fifth nerve palsies. Both upper and middle infarcts were present in this patient, which resulted in a mixed type of clinical picture. In a similar case reported by Chin Shah, Weber’s syndrome was associated with vertical as well as horizontal gaze palsy. In this case, horizontal gaze palsy was absent.

This patient also had thalamic infarct. Thalamic infarcts occurring simultaneously with the midbrain infarcts can be explained by the fact that a branch near the top of the basilar artery supplies both para median regions of the thalamus and rostral midbrain. When this branch is occluded, it leads to combined infarcts of the thalamus and midbrain.

Although ocular third nerve palsy is the commonest of isolated nerve palsies involving the eye and there are a number of studies showing the association of vertical gaze palsy with third nerve palsy; to our knowledge, only one case of Weber’s syndrome with vertical gaze palsy has been reported so far.

REFERENCES


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