Abnormal pattern visual evoked response in carotid-cavernous fistula

Sir,

The visual evoked cortical potential (VEP) to pattern reversal stimulation is a sensitive indicator of optic nerve function, and has the advantage that there is a highly reproducible waveform across subjects.^[1] We report the clinical and electrophysiological findings in a patient with a carotid-cavernous fistula, which presented exclusively left-sided signs and symptoms specifically post-traumatic

proptosis. Carotid-cavernous fistulas (CCFs) are abnormal communications between the carotid arterial system and the venous cavernous sinus.^[2]

An eight-year-old male child presented with diminution of vision and protrusion of left eye since one and a half month. The patient was apparently alright about two months back when he sustained trauma to left eye after he had a fall on ground. On ocular examination of the left eye, visual acuity was 6/24, PR accurate, eyeball exophthalmatous (28 mm), protruded forwards, downwards and medially. On auscultation, bruit was present over eyeball and on carotid artery. Episcleral vessels were dilated, tortuous, and pulsating. Fundus exam also showed -vessels dilated and tortuous and disc edema. Intraocular tension was 20 mmHg. Examination of the right eye was normal with visual acuity 6/6, PR accurate, normal fundus and disc. Intraocular tension was 16 mmHg. Visual fields, color vision (Ishihara) were unremarkable in both eyes. Figure 1 shows the photograph of the patient on day of VEP examination, showing a proptotic left eye. MRI brain and orbit study technique revealed dilated and tortuous superior ophthalmic vein (5 mm) of left eye with enlarged left cavernous sinus with opacification in post-contrast studies. Evidence of proptosis, mild thickening, and enlargement of left eye muscles. This explains the restriction of ocular motility seen in the patient on the affected side. NECT and CECT orbit study revealed dilated tortuous enhancing vessels draining left cavernous sinus likely to be carotido-cavernous fistula. This MRI scan prompted the referring ophthalmologist to opt visual electrophysiology, and then pattern reversal VEP (PRVEP) was performed in an attempt to exclude significant underlying pathology and to provide re-assurance. The VEP testing was conducted in the neurophysiology unit of the department of physiology of our institute. Left eye PRVEP was found to be grossly

Figure 1: Photograph of the patient on the day of VEP examination, showing a proptotic left eye

abnormal with marked amplitude reduction, prolonged P100 latency, and prolonged P100 duration relative to that from the right eye. The value of P100 latency in LE was 111.5 msec, and the amplitude was 4.25 μ v. The PRVEP from right eye was within normal limits, and there was interocular waveform asymmetry. The value of P100 latency in LE was 99.8 msec, and the amplitude was 11.60 μ v. These neurophysiological findings, therefore, suggested clear cut left optic nerve dysfunction. VEP waveforms of left and right eye are depicted in Figures 2 and 3. The case was then referred for surgical treatment with detachable balloon, which later led to normalization of vision and returning of the prolonged latencies back to normal.

Clinical manifestations of CCFs frequently involve ophthalmologic abnormalities, and as a result, patients initially consult an ophthalmologist. Regarding its mortality/morbidity, as many as 90% of patients with direct carotid-cavernous fistulas (CCFs) may lose vision if not treated. Most often, CCFs are broadly classified as either direct or indirect. Direct CCFs account for 70-90% of all CCFs.^[3] Direct fistulas are often traumatic, and they characteristically have high rates of blood flow.^[4] Patients

invariably present with the classic triad of chemosis, pulsatile exophthalmos, and ocular bruit.^[5] Diplopia and visual loss also may result with these fistulas. Pulsating exophthalmos is seen due to dilated ophthalmic veins and swelling within the orbits. The hemodynamic characteristics of carotid-cavernous sinus fistulas include increased venous pressure in the orbit and signs of orbital congestion, such as proptosis, dilation of episcleral and retinal vessels, ocular hypertension, dilation of the superior ophthalmic vein, and enlargement of the extraocular muscles. The potentially sight-robbing vascular abnormality, known as the carotid-cavernous sinus fistula (CCF), can masquerade as conjunctivitis or other common ocular conditions if not managed in a proper way, which diminishes the chance for a speedy diagnosis. But, treatment success rates for these fistulas can approach 100% when the repairs are performed early by experienced specialists. Prompt diagnosis, however, remains a challenge. The diagnosis is often missed, leading to months of inappropriate therapy and, in some patients, vision loss due to treatment delay. Hence, the purpose of the VEP recording in such cases is to potentiate early diagnosis and planning of surgical intervention since timely intervention is mandatory to prevent morbidity or mortality. We presented an unusual case of a post-traumatic high flow carotido-cavernous fistula with abnormal visual electrophysiological manifestations that were exclusively present on the ipsilateral side. When PRVEP was performed on the patient, it depicted marked amplitude reduction, prolonged P100 latency, and prolonged P100 duration relative to that from the right eye. These neurophysiological findings, therefore, suggested clear cut left optic nerve dysfunction and aided in anterior visual pathway diagnosis. This non-invasive technique presents as an excellent alternative to invasive vascular studies such as angiography for the diagnosis and evaluation of carotid-cavernous sinus fistulas.

Figure 2: VEP wave forms in the right and left eye recordings

Figure 3: Superimposed VEP wave forms in the right and left eye recordings

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