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. 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.

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
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 μV. 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 μV. 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. Direct fistulas are often traumatic, and they characteristically have high rates of blood flow. Patients invariably present with the classic triad of chemosis, pulsatile exophthalmos, and ocular bruit. 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.

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Sir,

Malaria continues to be an important parasitic disease in tropics and contributes to morbidity and mortality. We report a case of *Plasmodium vivax* infection who presented with features of spontaneous extradural bleed in order to create an awareness of this unusual entity among practitioners.

A 22-years-old male lorry driver was referred from a private hospital to our emergency department (ED) for altered sensorium, while he was on treatment for 4 days for smear positive *vivax* malaria. He recently had a trip to northern India in the lorry. There was no history of trauma, fall from height, seizure, bleeding diathesis, substance abuse, or intake of any herbal preparations. Prior to referral he was given chloroquine, paracetamol, domperidone orally, and intravenous fluids for 3 days. However, fever persisted even after a course of oral chloroquine as per WHO recommendation. On examination in ED, he was drowsy, disoriented with Glasgow Coma Scale of 9 (E2, M5, and V2), febrile (102.5°F), dehydrated, pale, and icteric. There were no other bleeding manifestations. He was hemodynamically stable and maintaining adequate saturation in room air. He had diffuse abdominal tenderness with hepatosplenomegaly. His pupils were unequal with sluggish reaction to light and planters were bilaterally extensor. There was no neck rigidity. His investigations revealed hemoglobin of 6.0 gm/dl (normal: 12.5‑14.5), platelet count of 45,000/cumm (140,000‑400,000), serum creatinine of 1.8 mg/dl, total bilirubin 9.6 mg/dl (0.4‑1), International normalized ratio (INR) of 1.12, and activated partial thromboplastin time (APTT) of 30 s (24‑32). His remaining parameters were within normal limits. His HIV status was negative. Peripheral blood smear was positive for *Plasmodium vivax* gametocytes and ring stages with marked parasitaemia and rapid card test was done to rule out undetected mixed infection, which is based on detecting specific *Plasmodium* LDH antigen by using monoclonal antibody directed against iso‑forms of the enzyme. Noncontrast computed tomography of the cranium demonstrated left side extradural hematoma. He was started on intravenous artesunate 2.4 mg/kg at the time of admission, then at 12 and 24 h, and then, once a day for 5 days. In view of his deteriorating neurological status, he underwent prompt surgical decompression and 50 ml of dark red blood clots were evacuated. As he had high parasitic index, partial exchange transfusion (1350 ml) was carried out which reduced the parasitaemia. He had a favorable outcome, with remarkable clinical improvement in the immediate postoperative period. We did not start oral artesunate tablets because it also contains mefloquine, which can precipitate neuropsychiatric complications. The repeat peripheral smear at the time of discharge was free of malarial parasite and he was discharged on primaquine 15 mg/day for 14 days after screening for G6PD deficiency.

*Plasmodium vivax* infection though considered to be benign and self‑limiting disease, in recent past it has produced severe and complicated malaria.