Pregnancy-precipitated status epilepticus: A rare presentation of MELAS syndrome

Sir,

Seizures are sometimes seen in pregnancy. However, rarely do we come across an underlying etiology, which is at least partially treatable. We here report an infrequent cause of seizures precipitated by pregnancy.

A 29-year-old female, primigravida with 10 weeks amenorrhea, presented in status epilepticus caused by focal seizures involving the left side of the face and left upper limb with secondary generalization since one day. She had previous history of a generalized seizure three years ago (well controlled on carbamazepine 600 mg/day with good compliance) and right hemiparesis with recovery within 24h, two years ago. There was no history of fever, vomiting, substance abuse, vasculitis, poisoning or trauma; and no history of developmental delay, short stature or preexisting hearing, visual, cognitive or motor impairment before the pregnancy. There was no history of any neurological disease running in her family.

On examination, patient was drowsy, pulse-102/min, BP-130/80 with mildly dilated and sluggishly reacting pupils, hypotonia, sluggish deep tendon reflexes with bilateral extensor plantar response. There was no neck rigidity or focal deficit. The seizures were acutely controlled with intravenous lorazepam. Investigations: CBC, blood sugar, liver and kidney function tests, coagulation profile and EKG were normal. Blood gas showed high anion gap metabolic acidosis without hypoxemia. Urine pregnancy test was positive. HIV, VDRL, ANA were negative. CSF showed no cells, protein-60 mg/dl, sugar- 70 mg/dl, lactic acid- 40 mg/dl (normal-10.8-18.9). MRI Brain showed multiple, bilaterally symmetrical cortical and subcortical areas of signal...
alteration in both cerebral hemispheres, basal ganglia, brainstem and cerebellum, not restricted to any vascular territory, hypointense on T1 and hyperintense on T2 and FLAIR sequences. MR angiogram was normal. Overall picture suggested mitochondrial disease [Figure 1]. Muscle biopsy from right quadriceps showed normal-sized muscle fibers with peripheral nuclei, few atrophic fibers and increased fibers with internal nuclei in hematoxylin-eosin stain. Succinate-dehydrogenase stain showed abnormal mitochondria and Gomori-trichrome showed 13% ragged-red fibers suggestive of mitochondrial cytopathy [Figure 2]. There was no vasculitis and membrane proteins were normal. Nerve conduction studies were normal. Gene analysis could not be done. Patient was maintained on a higher dose of carbamazepine (1000 mg/day). No recurrence of seizures was seen till 20 weeks gestation after which she was lost to follow-up. The outcome of pregnancy could not be determined.

Mitochondrial myopathy, encephalopathy, lactic acidosis, stroke-like syndrome (MELAS) is associated with deficiency of Complex I and IV of mitochondrial energy-generating pathway. Though it is maternally inherited, often the patients present as sporadic cases. It is diagnosed by three invariant criteria: (1) stroke-like episodes before age 40 years; (2) encephalopathy characterized by seizures, dementia or both; and (3) lactic acidosis, ragged-red fibers or both.

Our patient satisfies the above criteria. She had history of seizures, stroke; evidence of myopathy, lactic acidosis in CSF- a characteristic feature; muscle biopsy and MRI consistent with diagnosis. Usually, the age of onset is in childhood. Seventy per cent of patients experience their first symptoms before age 15. The peculiarities of our case are: somewhat late onset of symptoms, rare association of MELAS with pregnancy and deterioration of symptoms in pregnancy.

Prior reports on MELAS with pregnancy are rare (we could find only three). Yanagawa et al reported a 31-year-old woman who developed myopathy and neuropathy during pregnancy. Her symptoms subsided spontaneously and she delivered. Similarly, in our patient also symptoms deteriorated during pregnancy. Pregnancy, a high energy requiring state, increases the stress on mitochondrial function and so aggravates mitochondrial diseases. Also, in pregnancy the serum levels of carbamazepine fluctuate, which might have led to precipitation of seizures. No alternative cause of deterioration (excessive exertion, infection, drugs) were found to explain the precipitation other than pregnancy.

This has important therapeutic implications. Phenytoin and barbiturates should be avoided in suspected mitochondrial cytopathies. Although new medications such as co-enzyme-Q and dichloroacetate have been described for mitochondrial diseases, their safety in pregnant women has not been established. Intensive perinatal care, with close cooperation among medical specialists is important for a successful outcome.

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References
Letters to Editor

A 30-year-old male was admitted after his motorcycle skidded causing his crash helmet to shatter. Apart from a transient loss of consciousness, the patient did not have any neurological deficits. On examination, he had stable vitals and a full Glasgow coma score. He was distraught as the helmet side-bolt anchoring the facial visor had penetrated his left frontal scalp [Figure 1].

A CT scan of the head revealed that the bolt was lodged against the outer table of the left frontal bone [Figure 2] with no intracranial injury. A surgery, the bolt, nut and a fiberglass helmet shard [Figure 3] were disimpacted from bone and removed. The wound was debrided and closed primarily under local anesthesia. The patient was discharged the next day and had a healthy wound at a follow-up of six weeks.

Moped and motorcycle riders are involved in more than half of all road traffic fatalities with helmetless riders being three times more likely to sustain head injuries than those with helmets. [1]

Crash helmets help reduce head-injury-related mortality by 71%. [2] It is to be emphasized in the strongest possible language that this crash helmet was instrumental in saving the patient from a potentially fatal outcome. The authors are unaware of any such case as has been recorded in this report. However [3], the 'cranial bolt' does indicate that performance tests for crash helmets may not exactly replicate ground conditions. This may result in unpredictable patterns of injury as in this case.

As the bolt, nut and a shard of the helmet were recovered in toto after surgery, it is hypothesized that the side impact smashed the helmet at its attachment with the face visor. This was then driven inwards. The sharp threads of the bolt pierced the scalp and lodged against the frontal bone where the projectile's kinetic energy dissipated. It is our suggestion that the nut-bolt assembly anchoring the face visor be replaced by a rivet system or else by simply reversing the direction of the bolt's existing out-to-in direction. This will bring the broad bolt head

References


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