CASE REPORT

Diseases that masquerade neurology

Whenever I am asked to see a child and if the child happens to suffer from a seizure or an encephalopathy (altered sensorium) everyone including my conscious mind wants to start some magic treatment at the earliest to reverse the symptoms. I want to look like a hero! I want the parents to feel that I made their child well, I cured him. This is not unlike what the sages and prophets did centuries ago, and all of us have grown up on such stories! Each one us wants to be God, may be for a brief moment. This urge to act like God must be ignored while examining and treating a patient and this realisation took some time to dawn on me. This case illustrates how it is equally important to let the disease evolve before jumping to intervene.

Presentation

A 13 years old boy presented with an acute history of repeated vomiting (about 4 times in 6 hours) and a single episode of seizure. He had presented to the hospital with a left hemibody clonic seizure (jerking of left upper and lower limbs). After a dose of midazolam and fosphenytoin, the seizure subsided. There was no history of fever. He was a normal school going child with no significant past history. There was no family history of seizure.

In all likelihood, this appeared to be an acute symptomatic seizure*. Since the seizure was preceded by vomiting, it was unlikely to be first episode of seizure in case of epilepsy.

(*An acute symptomatic seizures is an event occurring in close temporal relationship with an acute CNS insult, which may be metabolic, toxic, structural, infectious, or due to inflammation.)
Following the seizure, the vital parameters of the child was normal. After about 12 hours, he had no abnormal neurological or systemic findings. He was ambulant with normal speech and cognitive functions.

Now it remained for me to find out what insult led to the seizure and how was it that he had recovered from the systemic or neurological insult in such a brief time and with only symptomatic treatment. Hypoglycemic or hypocalcemic seizures are not common at this age; hypo or hypernatremia usually occur secondary to any underlying disease which was not the case in this boy. A vascular insult causing a seizure should definitely show presence of neurological deficits or altered sensorium beyond 24 hours of presentation, while this child had recovered completely in 24 hours.

Since clinical medicine has now taken a backseat, it is obvious that I did not bother to use my neurons any further and instead ordered for neuroimaging!

Investigations

Complete blood count and serum electrolytes were normal

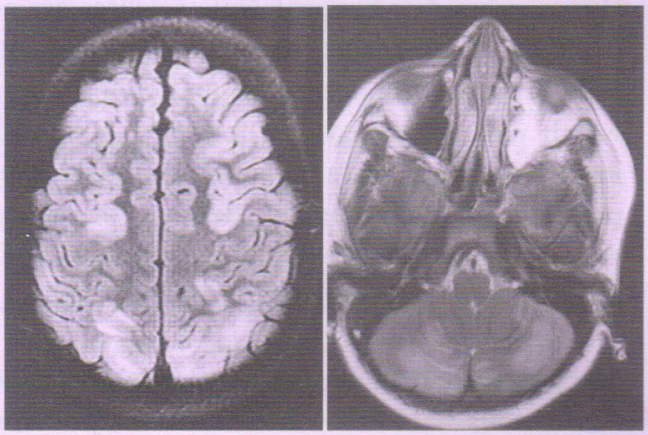
MRI (Magnetic resonance imaging) of brain including post-gadolinium study was performed. Multifocal lesions were present in cerebellar white matter and juxtacortical white matter which were hyperintense in T2weighted (long TR i.e repetition time and long TE i.e time to echo) images and FLAIR (fluid attenuated inversion recovery) and hypo to isointense in T1weighted (short TR and short TE) sequences. These lesions did not show evidence of reduced diffusivity or contrast enhancement.

Multiple lesions and such brief symptoms, now we had only two differentials: Clinically isolated syndrome (CIS) which is an idiopathic inflammatory demyelinating disease (IIDD) not unlike ADEM (acute disseminated encephalomyelitis) or PRES (posterior reversible encephalopathy syndrome).

Figure 1 - A and B.

A. Flair Axial images show juxtacortical multifocal hyperintensities

B. T2W Axial images show cerebellar white matter hyperintensities



About PRES

It is a clinico-radiological syndrome characterized by a headache, seizures, altered mental status and visual loss and characterized by white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain predominantly. Failure of cerebral autoregulation with vasogenic edema is a plausible explanation for this transient leucoencephalopathy. Hypertension and chemotherapy are most commonly associated with PRES. In hypertension associated or drug-induced PRES, the effective therapy includes withdrawal of offending agent, immediate control of blood pressure, anti-convulsive therapy and if required, temporary renal replacement therapy.

Back to our case

This 13 year old had no underlying disease. His neuroimaging features did not favour the possibility of demyelinating lesions. His clinico-radiological diagnosis was PRES. But what could have led to PRES and which investigations would help us find the underlying etiology or systemic disease that manifested as PRES ?

The patient started giving us the clue. His blood pressure readings gradually went up. Two subsequent readings at 4 hour interval were 142/98mmHg and 134/106mmHg respectively. Here you would expect me to involve the Cardiologist or Nephrologist! Well I remembered what my teachers in Pediatrics had taught me and that is to check the urine in a child who is hypertensive, so I decided to get an urinanalysis done first.

Urine routine/micro/culture: RBCs :30-40/hpf; Blood :2+; Protein :1+; Culture Analysis : No growth. C3 level - 30 mg/dl (range 80-200 mg/dl); ASO – 474 IU; USG KUB (ultrasonography of kidney urinary bladder).

Hypertension, hematuria and low C3 levels in a 13 year old boy meant post streptococcal glomerulonephritis (PSGN). PSGN in his case had decided to manifest as PRES!

The child was gradually taken off antiepileptic drugs and handed over to a nephrology colleague for further management and care.

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