Case Report

Intracranial Calcification in a Case of Seizure Disorder

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A patient with repeated episodes of Seizures and elevated Blood Pressure for 2 days, was admitted in a primary care set up initially, followed by admission to our Institute where he was thoroughly worked up to find the possible etiology behind the presentation. Extensive investigations and imaging led to the conclusion that the patient had Idiopathic Intracranial Calcification after the possible secondary causes of Intracranial Calcification were ruled out.

[*J Indian Med Assoc* 2022; **120(1):** 50-1]

Key words **: Seizure, Idiopathic, Intracranial, Calcification, Genetic study.**

Case Report

A 50 year old male from Krishnanagar, West Bengal presented with the chief complaint of repeated episodes of Seizures for last 2 days and was admitted in a primary care set up from where he was referred to our Hospital for further management. On Admission, he had Seizures, manifested as generalised tonic clinic convulsions associated with frothing from the mouth and urinary incontinence. Patient was brought to the Hospital in an unconscious state. There were no definite identifiable precipitating factors for his Seizure at the time of Admission.

Patient is Hypertensive for the last 5 years but was on irregular medication for the same.

There was no history of fever, trauma, vomiting or previous such Seizure attacks. He had no known addictions, there was no history of any high risk behaviour.

His family history was non-contributory as well.

Patient was unconscious at the time of Admission and regained consciousness within a few hours, after conservative management. Pulse rate was 106/min and respiratory rate was 22/min on Admission. His BP was recorded to be 204/102 mm Hg.

SpO2 was 98%. His, Higher Mental Functions (HMF) could not be assessed at the time of Admission. Tone was reduced in all four Limbs and plantar reflex was found to be extensor bilaterally. Pupils were normal in size bilaterally and reacting sluggishly to light.

Examination of the other systems was grossly normal. When he regained consciousness, he was disoriented and restless.

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Received on : 16/11/2021

Accepted on : 28/12/2021

Editor's Comment :

- Idiopathic intracranial hypertension should be considered as an important, albeit rare differential in a patient with seizures when no other apparent secondary cause can be as certained. While there is no primary intervention, the patient can lead a normal life on symptomatic management. Keeping this differential in mind would help build an important database of the rarer secondary causes of seizures.

He was managed with supportive care and appropriate measures were taken to reduce his BP.

Initial lab investigations showed a Haemoglobin (Hb) of 13.8, normal Total Leukocyte Count (TLC), Differential Count (DC) and Platelets.

Urea, Creatinine, Sodium, Potassium and Liver Function Tests (LFT) were within normal range.

His Integrated Counselling and Testing Centre (ICTC) report was non reactive.

An urgent NCCT Brain was done at admission (Figs 1 & 2).

A Neuro-medicine consultation was taken and as per their advice the following tests were done :

Calcium - 1.27 mmol/l (1.15-1.33 mmol/L)

Magnesium- 2.1 meq/l (1.3- 2.2 meq/l)

Phosphate- 2.91mg/dl (2.5- 4.5 mg/dl)

Intact Parathyroid Hormone (iPTH) - 37.8 pg/ml (15-65 pg/ml)

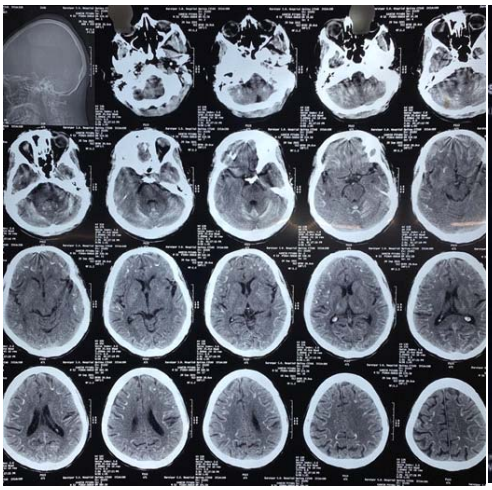
S Lactate - 0.8 microU/ml (0.5- 1 microU/ml)

MRI brain with contrast was done (Figs 3 & 4)

Patient gradually improved over the next 24-48 hours. He became oriented with a Glasgow Coma Scale (GCS) of 15/15. Power in all 4 Limbs was normal, tone normalised and bilateral plantars were flexor. No neuro deficit was found.

He underwent further tests like Magnetic Resonance Angiography (MRA) and Magnetic Resonance Venography (MRV) keeping a possibility of vessel calcification in mind. Both investigations were normal. A Toxoplasmosis, Rubella Cytomegalovirus, Herpes Simplex (TORCH) profile was done and was also negative. EEG, ECG and Carotid Artery Doppler were done and found to be normal.

As our Lab does not have provision for gene analysis, patient was informed about the constraint and advised to



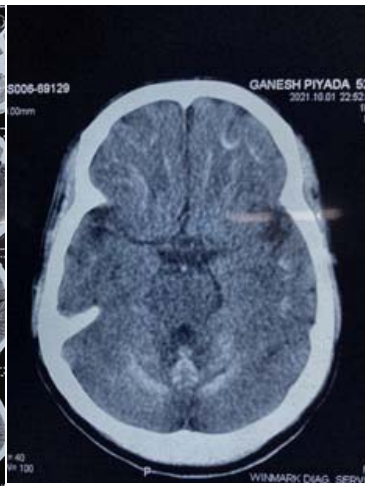
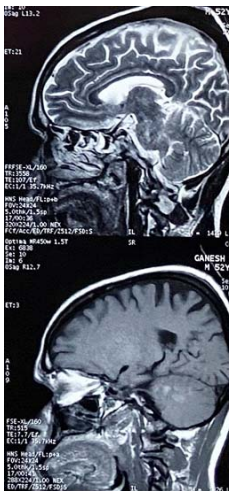
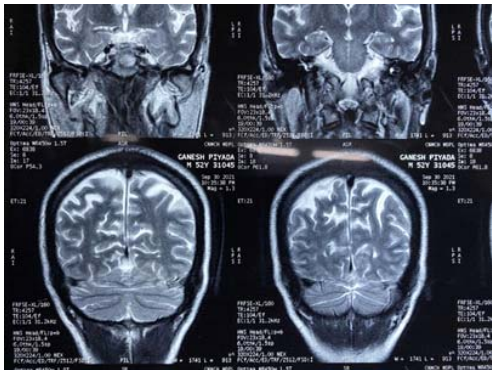


Fig 1 & Fig 2 — NCCT Brain shows diffuse parenchymal hyperdensities along the grey white matter junction and also in the cerebellar parenchyma





Figs 3 & 4 — MRI brain (plain) showing areas of calcification in the cerebellum and areas of apparent demyelination in the thalamus and pons as well as acute lacunar infarction in left frontal lobe.

follow up at a centre with provision for gene analysis for SCL20A2 and PDGFRB.

Patient did not have any further episodes of Seizures after Admission. He was treated with antiepileptics and his Blood Pressure levels normalised with anti-hypertensives. He was discharged with a diagnosis of Primary Brain Calcification (Idiopathic Intracranial calcification) with symptomatic seizures and was put on maintenance antiepileptics.

Discussion

Primary (Idiopathic) Intracranial Calcification, also known as Fahr disease among several other names refers to both familial and sporadic cases of calcification. The term encompasses the calcification of Bilateral Basal Ganglia, along with cerebellum and in some instances, Cerebral White Matter[1].

The onset of primary brain calcification is usually in the fourth to sixth decade[2]. The typical presentations include psychiatric and cognitive manifestations like rest and action tremor, Parkisonism, dystonia, choreoathetoid movements, myoclonus and tics. Rigidity and bradykinesia along with cerebellar signs are also seen[3,4].

Patient can present with Epilepsy, Syncope, Stroke, and stroke-like episodes. Up to one-third of patients may be asymptomatic, even at advanced age.

The most commonly mutated gene is SLC20A2 which accounts for around 40 percent of cases. The next most common gene involved is PDGFRB gene implicated in about 10 percent of cases[5]. In about 50 % of individuals the cause still remains unknown.

Diagnosis of the condition is by Neuroimaging techniques such as Computed Tomography (CT) of the brain (the most sensitive technique) and Magnetic Resonance Imaging (MRI). MRI has been reported to be normal or falsely suggestive of demyelination in a few patients with CT proven diffuse calcification[6]. The lack of biochemical abnormalities or infections like CMV and other TORCH infections, trauma, exposure to toxins and heavy metals makes the diagnosis of Idiopathic Intracranial Calcification very likely.

Further Genetic Testing for the implicated mutation helps narrow down the diagnosis.

Symptomatic treatment remains the only option till date and genetic counselling has been strongly advised, especially for individuals with Platelet-Derived Growth Factor (PDGF) mutation.

Conclusion

There are a myriad of secondary causes of Seizure Disorder. However, it should also be kept in mind that Idiopathic Intracranial Calcification, though rare, can manifest with the above presentation. Though the genetic analysis could not be done due to certain constraints, it can be assumed that when the other possible causes are excluded, this becomes the most likely possibility behind the seizure and the patient would need treatment with anti-epileptics for a good outcome and a normal life.

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