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Case Report

Seizure And Cerebellar Signs: A young lady with Hypoparathyroidism

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Abstract
Neurological emergencies are known to be difficult to manage due to lack of cure in most instances and the long lasting implications of the disease. However, a reversible cause if could be identified, can change the entire course of treatment and prognosis of these conditions. We are reporting a case of 16 years lady who presented to us with generalized tonic clonic convulsion (GTCS) and cerebellar signs. With proper investigations she was diagnosed as a case of idiopathic Hypoparathyroidism. Her successful treatment with calcium and calcitriol enriches our knowledge of hypoparathyroidism.

Key words: Hypoparathyroidism, Calcium metabolism, Metabolic disorder Seizure

Background

Hypoparathyroidism is an uncommon endocrinal disorder which can present with various neurological manifestations as a consequence of hypocalcaemia.

Patients may present with life threatening situations or remain asymptomatic throughout their entire life and diagnosed just incidentally. Seizure is one of the most common modes of presentation of Hypoparathyroidism in the Emergency, although neuropsychological presentations are also increasingly identified. The cause of seizures in Hypoparathyroidism is the hypocalcaemia itself, but rarely can also be due to intracranial calcifications.[1] Extra-pyramidal signs and cerebellar signs have rarely been reported and so too have been psychological disorders.[1,2] Interest lies in this case as successful treatment after prompt diagnosis cured her seizures. However cerebellar signs persist even after normalization of serum calcium, phosphorus which hints towards the complex nature of this disease.

Case report

A 16 year old female patient attended to the Emergency department of Burdwan Medical College with recurrent generalized tonic clonic seizure (GTCS) which was going on for the last 30 minutes. She was immediately started on Intravenous Lorazepam to control the status epilepticus.

She had suffered episodes of a similar nature one day back as well but it had not progressed to 'status' then. Her family members denied any previous history of seizure, head injury, fever or substance abuse. She was non diabetic, with normal menstrual history and did not have any previous psychiatric illness. After controlling the status and stabilizing the patient a thorough clinical examination was performed. She had a normal body habitus with an average build. She developed carpopedal spasm on sustained pressure by sphygmomanometer while measuring her BP. On reviewing the history with her parents once again, they mentioned that she had gradually progressive difficulty in walking with fine tremor and frequently would lose her balance during the last couple of months. Her Blood pressure was 160/90 mm of Hg, pulse 94/minute and sPO2 97% on room air. Cardiovascular and respiratory system, neck and cranium examinations were all unremarkable. There were no motor or sensory deficits.

After initial stabilization she was taken into ward and base line investigation were sent. In the ward a further neurological examination revealed truncal ataxia, dys-synergia, past pointing and intension tremor. Her tandem walk was impaired. After 12 hours, she suddenly developed another episode of Grand mal seizure for about 5 minutes which was controlled with another dose of intravenous Lorazepam. Thereafter she was also started on intravenous Levetiracetam.

Her routine blood tests were all normal (Table 1) except for hypocalcaemia and hyperphosphatemia. On getting laboratory result of hypocalcemia she was given intravenous Calcium glucuronate. Within 48 hours of hospital stay she became seizure free and signs of latent tetany subsided. She was started on oral Calcium carbonate 2g/day, along with Calcitriol 0.50mcg/day along with oral Levetiracetam from 3rd day of her admission.

Her iPTH level was 4.6pg/ml (11-54pg/ml). A screening non contrast computed tomography (CT) scan of brain showed bilateral hyper-intensity suggestive of calcifications involving the basal ganglia, cortex, cerebellum which were bilaterally symmetrical (Figure 1). Her urine examination and Radiological examination of her abdomen were normal.

Table 1: Showing data from routine blood tests

Parameters	**Value**	**Reference**
Random Sugar	130mg/dl	-
Hb	14g/dl	11-15g/dl
TLC	9000/cc	4000-11000/cc
Urea	28mg/dl	25-40mg/dl
Creatinine	1.3mg/dl	1.1-1.5mg/dl
Na	140mEq/l	135-145mEq/l
K	4.0mEq/l	3.5-5.0mEq/l
Cl	95mEq/l	92-110mEq/l
Ca	**5.6mg/dl**	**8.0-10mg/dl**
Mg	2.5mg/dl	1.8-3.0mg/dl
PO4	3.0mg/dl	2.6-4.6mg/dl
Bilirubin (Total)	1.2mg/dl	1.1-2.0 mg/dl
ALT	30 IU/L	< 40 IU/L
AST	32 IU/L	< 40IU/L
ALP	50 IU/L	30-100 IU/L
Protein (total)	7.0 g/dl	6-8 g/dl
Albumin	4.0 g/dl	3.5-4.5g/dl
pH	7.4	7.35-7.45
pCO2	40	35-45
pO2	90	80-100
HCO3	24	22-30
SpO2	95% On Room air	
TSH	2.3 pIU/ml	0.5 - 5.5 pIU/ml

She was discharged after seven day with normalization of serum calcium and phosphorus level. On her last follow up in outpatient department two months later she is still seizure free but has some cerebellar signs.

Discussion

Seizure disorder is one of the most common causes of emergency admission where aetiology remains obscured most of the time. Finding a reversible cause not only gives the hopes of cure but also provides a window into the systemic nature of the disease presenting with seizure. Hypoparathyroidism is being increasingly identified as an endocrinal entity with varied neurological manifestation. Most seizures are generalized tonic clonic in type and respond well with treatment.[1] Hypoparathyroidism may be acquired or hereditary.

When no obvious cause is identified it is termed as Idiopathic.[1] Inadvertent removal of parathyroid tissue, especially neck surgery or thyroid surgery and some rare infiltratitive diseases cause acquired hypoparathyroidism. Isolated developmental anomaly of the parathyroid glands is rare, and when present, is also associated with defects involving other endocrine systems or muco-cutaneous manifestations.

Both varieties usually manifest in the first decade of life though exception do occurs. Interestingly post surgical hypoparathyroidism manifests seizure less commonly compared to idiopathic variety. Our patient presents in emergency at her late teens with successive episodes of generalized seizure. After carefully exclusion of acquired causes she was diagnosed as idiopathic hypoparathyroidism.

Hypocalcaemia dominates the clinical presentation of hypoparathyroidism. Circumoral numbness, parasthesia, carpopedal and laryngeal spasm, tetany and seizures are the most common presentations.[1] Parathyroid has been rightly termed as the “Pandora’s box” of neurology.[4] Neurological manifestations range from seizures to raised intracranial tension, papilledema, psychosis, extra pyramidal symptoms and cerebellar manifestations.

Cerebellar calcifications secondary to hypoparathyroidism have also been reported.[2,3] This young female after initial stabilisation showed cerebellar signs subtly in form of intention tremor and ataxia.

Computed tomography (CT) of brain should be carried out in all hypoparathyroid patients with neurological manifestations. Apart from physiological causes, hypoparathyroidism, pseudohypoparathyroidism, infectious, metabolic disorders and some genetic diseases can cause of pathological calcifications in the brain.

Wide-spread intracranial calcifications beyond basal ganglia have also been reported.[2,5] In spite of extensive extra pyramidal and cerebellar involvement symptoms attributed to them have been reported very rarely.[2,5] Our patient had extensive involvement of basal ganglia, cortex as well as cerebellum (Figure 1).Though no extra-pyramidal signs were evident clinically, she had subtle cerebellar sign which were evident on clinical examination. There is increasing evidence that

Basal ganglia calcification in idiopathic hypoparathyroidism can progress even after strict control of serum calcium level.[6] A recent prospective study found that a low calcium to phosphorus ratio (hyperphosphatemia), and a history of convulsion are significantly associated with this calcification progression.[6] Vitamin D or 1,25 (OH)2 D3 (calcitriol) with high dose of calcium salt remains the treatment of choice in idiopathic hypoparathyroidism. Our patient has been treated with both as per recommendation in literature. Although this combination helps balancing the calcium-phosphate level, but it does not revert the hypercalciuria which can lead to renal stone. Subcutaneous recombinant parathyroid hormone replacement has been studied as a new promise without skeletal complications or nephrolithiasis but it is yet to be fully understood.[7] After 2 months follow-up our patient stays seizure free with maintenance of serum calcium and phosphate levels, but cerebellar signs are still evident on careful examination. We are reporting this case as an important reminder that common disease can present in varied manifestations and clinicians should be cautious whenever confronting in real scenario.

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Figure 1. Plain CT brain showing bilateral calcified foci