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CASUISTIC PAPER

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Basal ganglia calcifications is not inconsequential in pediatric cases

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ABSTRACT

Introduction. Basal ganglia calcification (BGC) in pediatric population is rare and is considered as a pathological finding. Various causes may be responsible for BGC including hypoparathyroidism, various infectious, toxicities or hereditary disorders.
Aim. We aimed to present a 8 year old boy presented with generalized seizure and bilateral small amount of globus pallidum calcifications on neuroimaging studies leading to the diagnosis of idiopathic hypoparathyroidism, which is a treatable cause of seizure.
Description of the case. A 8-year-old boy presented to our emergency department with generalized seizure for the first time in his life. There was no history of previous head trauma and his family history was unremarkable. Neurological examination revealed no pathological findings. Radiological imaging studies revealed only bilateral small amount of globus pallidus calcifications. He was referred to the pediatric endocrinology department for further evaluation of the hypocalcemic convulsion, where laboratory investigations revealed idiopathic hypoparathyroidism as the cause of hypocalcemic convulsion with exclusion of the other causes.
Conclusion. Even a small amount of BGC in pediatric patients may be the sign of primary hypoparathyroidism and should be evaluated with serum electrolyte levels for early diagnosis and for the prevention of multisystemic complications of hypoparathyroidism.
Keywords. basal ganglia calcification, idiopathic hypoparathyroidism, seizure

Introduction

Isolated basal ganglia calcifications (BGCs) on neuroimaging studies particularly on head computed tomography (CT) is a frequently observed phenomenon in elderly patients and is generally considered as an idiopathic age related incidental finding. Lentiform nucleus particularly globus pallidus is the most common location for these calcifications.[1] In addition to aging, idiopathic BGCs may also be primary familial in origin, which is known as Fahr disease. BGC in these cases typically presents in ages between 40 and 60.[2] However, recently it has been shown that it can also be observed in pediatric cases with chromosomal deletions on 8p11.[3] However, in pediatric patients, it is more common that BGCs occur as a result of secondary causes, including parathyroid disorders (hypoparathyroidism, pseudo-

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hypoparathyroidism), nephrotic syndrome, infections (like TORCH, brucellosis), various congenital disorders (like Cockayne syndrome), radiation or chemotherapy, toxicities (like hypervitaminosis D), which are all known as Fahr syndrome.[2,4-6] Nevertheless, BGC in pediatric population is rarer than adults and is considered as a pathological finding.

Aim

We aimed to show the importance of BGCs detected on pediatric patients and the necessity for further evaluation.

Description of the case

A 8-year-old boy presented to our emergency department with generalized seizure for the first time in his life. His mother defined the seizure lasting approximately 3 to 5 minutes with the features of generalized tonic clonic convulsion without accompanying a febrile disease. His seizure was not associated with loss of consciousness, postictal confusion or any focal deficits following the seizure episode. There was no history of previous head trauma and his family history was unremarkable. Neurological examination revealed no pathological findings. CT of head was performed at the emergency department to rule out any epileptogenic space occupying lesion but revealed only bilateral small amount of globus pallidus calcifications (Fig. 1).

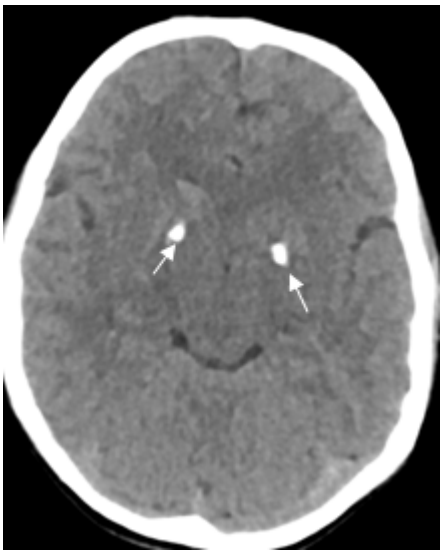

Fig. 1. Axial nonenhanced CT image of head shows bilateral symmetrical small GPCs (arrows)

He was evaluated by the pediatric neurology department and electroencephalography (EEG) showed normal findings. In addition, magnetic resonance imaging (MRI) was ordered to rule out any subtle structural abnormalities, but revealed normal findings except faint areas of increased signal intensity due to T1 shortening effect of calcifications (Fig. 2) and foci of signal losses on gradient echo (GRE) images (Fig. 3) at bilateral globus pallidus.

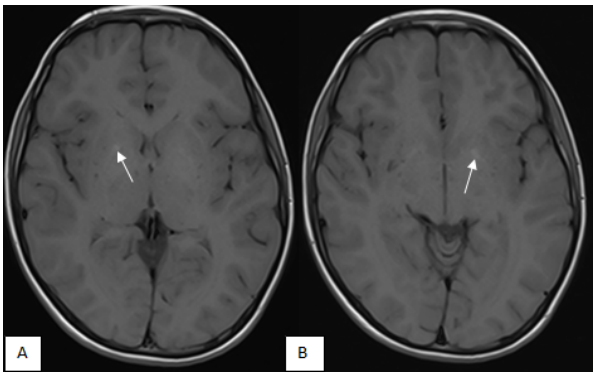

Fig. 2. Axial T1w MRI of brain show faint T1w shortening effect of calcifications on the right (A, arrow) and left (B, arrow) globus pallidum

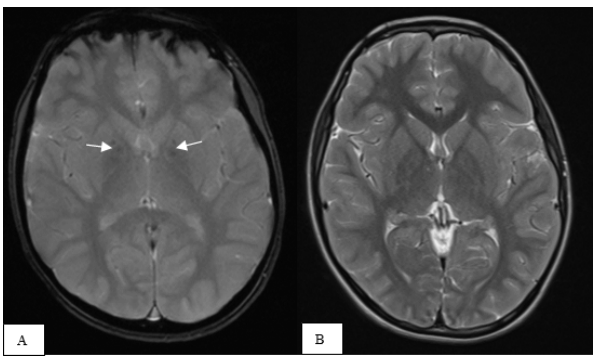

Fig. 3. Axial gradient echo (GRE) image and axial T2w image of the brain. Bilateral globus pallidus calcifications are seen as foci of signal losses on GRE image (A, white arrows). Note that these calcifications are not clearly visible on T2w images.

Initial laboratory results (Table 1) revealed normal hemogram and biochemistry except hypocalcemia (6.58 mg/dL) and hyperphosphatemia (8.23 mg/dL). The patient was referred to the pediatric endocrinology department for further evaluation of the hypocalcemic convulsion, where laboratory investigations revealed idiopathic hypoparathyroidism as the cause of hypocalcemic convulsion with demonstration of low level of serum parathyroid hormone (PTH) (12.2 ng/L). Hypomagnesemia, renal failure and vitamin D deficiency were excluded (Table 1). Renal ultrasonography was performed and ruled out nephrocalcinosis and nephrolithiasis. Ophthalmological examination was normal. DiGeorge syndrome was also excluded by echocardiography and chromosomal analysis.

*Informed consent was taken from the child's parents.

Table 1. The laboratory results

Parameter	Patient's laboratory results	Normal reference
calcium	6.58 mg/dL	8.8-10.8 mg/dL
phosphate	8.23 mg/dL	3-5.4 mg/dL
PTH	12.2 ng/L	15-65 ng/L
magnesium	1.75 mg/dL	1.7-2.1 mg/dL
creatinine	0.45 mg/dL	0.40-0.60 mg/dL
25-hydroxyvitamin D	24 ng/ml	20-40 ng/ml

Discussion

Hypoparathyroidism is an endocrinopathy diagnosed with the laboratory findings of low serum calcium and high serum phosphorus levels associated with decreased or in some instances absent PTH production. Patients present with effects of hypocalcemia including muscle cramps, paraesthesia, seizure, teeth and nail problems. In addition, due to hyperphosphatemia and increased serum phosphorus-calcium products, ectopic tissue calcifications may occur on kidneys, the hearth, vessels, eyes and many other organs and tissues. Intracranial calcifications particularly occurs on basal ganglia and can be associated with epilepsy, cognitive impairment and parkinsonism.[7] The association between BGC and hypoparathyroidism and a correlation between the duration and the severity of the hypocalcemia with BGC were first described by Eaton et al. in 1939.[8] PTH disorders was also reported as the most common definable etiology for white matter and bilateral subcortical nuclei calcifications.[9] Intracranial calcifications of hypoparathyroidism may also involve thalamus, subcortical white matter and corona radiata.[10] In general the most common causes of hypoparathyroidism are iatrogenic and include thyroidectomy & parathyroidectomy or radiotherapy involving the cervical region. In the absence of these situations, in pediatric cases some rare hereditary conditions particularly Di George syndrome and polyglandular autoimmune syndrome type 1 should be considered and excluded.[7] Apart from hypoparathyroidism, vitamin D deficiency/resistance, renal failure and hypomagnesemia may also be the underlying cause and should be excluded in order to accurately manage the hypocalcemic complications. Pseudohypoparathyroidism, the resistance to the action of PTH should be differentiated with its high level of serum PTH. In our patient, low calcium level was associated with the low level of PTH and the high level of phosphorus, consistent with the diagnosis of primary hypoparathyroidism. Serum magnesium, vitamin D and creatinine level were in the normal range.

Conclusion

Since hypoparathyroidism is a treatable cause of hypocalcemic seizure and is associated with multisystemic consequences when untreated, BGCs in pediatric patients should be evaluated with serum electrolyte levels for early diagnosis of hypoparathyroidism and for the prevention of multisystemic complications. Even in small amount of BGC in pediatric patients, as in our case, may be the sign of primary hypoparathyroidism.

References

1. Rastogi R, Beauchamp NJ, Ladenson PW. Calcification of thebasal ganglia in chronic hypoparathyroidism. *J Clin Endocrinol Metab.* 2003;88(4):1476-1477.
2. Perugula ML, Lippmann S. Fahr's Disease or Fahr's Syndrome? *Innovations in Clinical Neuroscience.* 2016;13(7-8):45-46.
3. Morris M, Kwon R, Chen L. Pediatric Idiopathic Basal Ganglia Calcification and Spherocytosis With Chromosome 8p11 Deletion. *J Neuropathol Exp Neurol.* 2020;79(2):238-241.
4. Dähnert W. *Radiology Review Manual.* Wolters Kluwer; 2011.
5. Pistacchi M, Gioulis M, Sanson F, Marsala SM. Fahr's syndrome and clinical correlation: a case series and literature review. *Folia Neuropathologica.* 2016:54(3): 282-294.
6. Amisha F, Munakomi S. *Fahr Syndrome.* Treasure Island (FL): StatPearls Publishing; 2020.
7. Marcucci G, Cianferotti L, Brandi ML. Clinical presentationand management of hypoparathyroidism. *Best Pract Res ClinEndocrinol Metab.* 2018;32:927-939.
8. Eaton LM, Camp JD, Love JG. Symmetric cerebral calcifications, particularly of the basal ganglia demonstrate roentgenographically; calcification of the cerebral blood vessels. *Arch Neurol Psychiatry.* 1939;41:921-942.
9. Manyam BV, Walters AS, Narla KR. Bilateral striopallidodentate calcinosis: clinical characteristics of patients seen in a registry. *Mov Disord.* 2001;16:258-264.
10. Saleem S, Aslam H, Anwar M, et al. Fahr's syndrome: literature review of current evidence. *Orphanet J Rare Dis.* 2013;8:156.