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Case report

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Fahr´s disease presenting with epileptic seizures: a case report

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Abstract

Fahr is a rare neurological disorder that leads to abnormal, symmetrical, and bilateral calcification of the basal ganglia and other brain regions, seizures are one of many symptoms to lead to the diagnosis of this disease. We here report a 45-year-old male diagnosed as Fahr´s disease presenting with loss of consciousness and multiples seizures. A 45-year-old male with a history of an untreated depressive syndrome, smoking, and active alcoholism, admitted to the emergency department with a sudden loss of consciousness and multiples seizures. The Computed tomography scan showed a bilateral and symmetric deposit of calcifications in basal ganglia, the blood tests excluded any other anomaly that may lead to secondary intracranial calcification, the patient denied any similar cases in the family, the clinical, biological and radiological

findings were compatible with a sporadic form of Fahr´s disease. Seizures are ones of the rarest symptoms that can reveal a Fahr´s disease, the finding of bilateral and symmetrical calcification of basal ganglia in a patient presenting with seizures should be reminiscent of this illness after excluding any other pathological processes leading to secondary intracranial calcification.

Introduction

Fahr also called idiopathic basal ganglion calcification (IBGC), is a rare neurological disorder characterized by the abnormal, symmetrical, and bilateral calcification of the basal ganglia and other brain regions, it was described for the first time by Karl Theodor Fahr in 1930 [1]. According to a registry of Fahr´s disease, symptomatic individuals represent 67% of the studied population, with a higher incidence among men than women [2]. Patients usually present with chronic cognitive deterioration, later in the development of the disease, other symptoms and signs occur, especially extrapyramidal symptoms. Seizures are ones of the rarest presentation of this disease. The prognosis is very hard to predict, there is no correlation between the age and the course of evolution. We´re going to report a 45-year-old male that was diagnosed with Fahr´s disease presenting with multiples seizures. We review the literature to look for similar cases and we discuss the links between these two entities.

Patient and observation

We´re going to report the case of a 45-year-old patient, with a history of an untreated depressive syndrome, smoking and active alcoholism, owner of a food shop, who was admitted to emergency department because of a sudden loss of consciousness followed by multiple seizures, this in the absence of any triggering factor, such as trauma or drug use. During the interrogation, the patient denied any similar cases in the family or any past medication. The clinical examination finds a patient in a good general condition with stable vitals: blood pressure 146/70mmHg, pulse: 90 beats/min, respirations 18 breaths/min and normal temperature of 37°C, the sugar blood level was normal too (1,09 g/l), there were no other abnormalities to mention including the neurologic examination. The patient experienced another seizure during the examination which stopped after the administration of 2mg of midazolam. According to the family, the patient experienced recently a change in behavior with a tendency to anxiety, agitation and an impressing increase in alcohol consumption, with normal cognition and this was the first time for him to have seizures. Blood tests, which included blood cell count, electrolytes, calcium 103.1 mg/l, phosphor 33.2mg/l, and parathormone were strictly normal, except a little elevation of transaminase (GPT/GOT): 203 and 228 UI/L respectively, and GGT 174 UI/L due to severe alcoholism. In front of this clinical presentation, the patient underwent a non-contrast head computed tomography in order to explain the seizures, which objectified a bilateral and symmetric calcification of the basal ganglia (Figure 1). We could not perform a magnetic resonance imaging on the patient due to financial issues. However, his electroencephalogram showed no signs of epileptic activity. The patient was tested negative for HIV, hepatitis, and toxicology showed signs of alcohol in urine. In the light of the clinical and radiological shreds of evidence and the absence of any calcium-phosphorus metabolism disorder or any other plausible cause, the diagnosis of the Fahr´s disease was retained. The patient was put on anticonvulsant treatment “Sodium valproate”, stayed for 48h under surveillance in the intensive care unit as there is a risk of recurrence of the seizures, and then transferred to the neurology department. During this period the patient did not experience other seizures and a few days later the patient was discharged from the hospital. The patient was followed up afterward, no more seizures were observed, and the neurologists maintained the antiepileptic treatment in view of the possible risk of recurrence. However, the behavioral disorders have persisted so the patient was referred to a psychiatrist for further treatment.

Discussion

Fahr also called idiopathic basal ganglion calcification (IBGC), is a rare neurological disorder characterized by the abnormal, symmetrical, and bilateral calcification of the basal ganglia and other brain regions with the absence of any systemic calcium disorder. The physiopathology is not clear. It´s suggested that calcifications in Fahr´s disease may be attributed to a metastatic deposition, secondary to local disruption of the blood-brain barrier (BBB), or a disorder of neuronal calcium-phosphorus metabolism [3]. Autosomal dominant inheritance in the familial cases was recently explained due to the identification of the responsible gene mutations [4], but sporadic forms continue to be described in the literature. The common sites of calcifications in Fahr´s disease are globus pallidus, putamen, caudate nucleus, internal capsule, dentate nucleus, thalamus, and cerebral white matter [5]. The clinical presentation is polymorphic, patients remain asymptomatic for a long period, the typical presentation starts in the 4th or the 5th decade of the life with usually a cognitive decline, psychiatric symptoms, movement disorders, seizures are reportedly ones of the rare symptoms that lead to the diagnosis of the Fahr´s disease [6]. It believed that they are maybe related to a dysfunction of the cortico-basal connections and their inter-hemispheric relation [6].

To our best knowledge, it was reported a few times [6-8]. Ongun *et al*. reported two cases of Fahr´s syndrome presenting with epileptic seizures, in both clinical cases, it seems to be related to secondary causes [6]. We must not confuse Fahr´s disease and Fahr syndrome which is due to a secondary cause such as idiopathic hypoparathyroidism, secondary hypoparathyroidism, hyperparathyroidism, cysticercosis, toxoplasmosis, HIV infection [9]. As far as for our patient, he had been well until the incident, the behavior changes are recent according to the family, he never experienced seizures before and he has not stopped drinking alcohol before the incident, which eliminates alcoholic withdrawal seizures. There are no similar cases in the family, but asymptomatic forms may exist, the laboratory tests showed a normal balance of calcium and phosphate ions which excludes a parathyroid disorder. The behavior troubles, the depressive syndrome, the age, and the CT scan images, in an addition to the absence of similar cases in the family, are all in favor of a sporadic form of Fahr´s disease. Treatment is usually symptomatic. Many trials have been conducted without any major outcome [10]. In our case, the anticonvulsant treatment led to the complete disappearance of the seizures until the last control.

Conclusion

To sum up, the Fahr´s disease is a rare neurodegenerative entity which is poorly explained, the diagnosis is based on a cluster of clinical, biological and radiological arguments, unfortunately, there is no specific drug to stop the progression. However, symptomatic treatments can be effective to control some of the symptoms at the early stages. It is important to report our case because the rarity and clinical polymorphism of this pathology generally makes it diagnosed at advanced stages. Seizures remain one of the rarest symptoms of this pathology, especially when they are generalized, which is the case of our patient, the possibility of Fahr´s disease should always be kept in mind when faced with convulsive seizures with bilateral and symmetric calcification of the basal ganglia, in the lack of another more relevant diagnosis.

Competing interests

The authors declare no competing interests.

Authors' contributions

AB examined the patient and drafted the manuscript. BH evaluated the neuroimaging findings and gave important clinical opinions. CC and MK participated in the design of the case report

and helped to draft the manuscript. All authors read and approved the final manuscript.

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We would like to thank the cooperation from the patient.

Figure

Figure 1: noncontrast CT of the brain, symmetric calcifications of basal ganglia can be observed in a axial and b coronal sections

References

1. Fahr TH. Idiopathische verkalkung der hirngefasse. Zentralbl Allg Pathol. 1930;50: 129-133. **Google Scholar**
2. Manyam BV, Walters AS, Narla KR. Bilateral striopallidodentate calcinosis: clinical characteristics of patients seen in a registry. Movement Disorders. 2001;16(2): 258-264. **PubMed**| **Google Scholar**
3. Malik R, Pandya V, Naik D. FAHR Disease - a rare neurodegenerative disorder. Movement Disorders. 2001;16(2): 258-264. **Google Scholar**
4. Hsu SC, Sears RL, Lemos RR, Quintáns B, Huang A, Spiteri E *et al*. Mutations in SLC20A2 are a major cause of familial idiopathic basal ganglia calcification. Neurogenetics. 2013;14(1): 11-22. **PubMed**| **Google Scholar**
5. Lester J, Zuñiga C, Díaz S, Rugilo C, Micheli F. Diffuse Intracranial calcinosis: Fahr disease. Arch Neurol. 2006;63(12): 1806-1807. **PubMed**| **Google Scholar**
6. Ongun N, Degirmenci E, Erdogan C. Fahr´s syndrome presenting with epileptic seizure: Two case reports. North Clin Istanb. 2016;3(1): 71-74. **PubMed**
7. Rangaswamy null, Ranjith V, Vikas L, Santosh R. Fahr´s Disease with seizure presentation. J Assoc Physicians India. 2016;64(8): 85-86. **Google Scholar**
8. Chouaib N, Rafai M, Belkouch A, Bakkali H, Belyamani L. Fortuitous discovery of Fahr´s syndrome after seizures. Rev Neurol (Paris). 2015;171(12): 894-895. **PubMed**
9. Harrington MG, Macpherson P, McIntosh WB, Allam BF, Bone I. The significance of the incidental finding of basal ganglia calcification on computed tomography. Journal of Neurology, Neurosurgery & Psychiatry. 1981;44(12): 1168-1170. **PubMed**| **Google Scholar**
10. Loeb JA, Sohrab SA, Huq M, Fuerst DR. Brain calcifications induce neurological dysfunction that can be reversed by a bone drug. Journal of the Neurological Sciences. 2006;243(1-2): 77-81. **PubMed**| **Google Scholar**

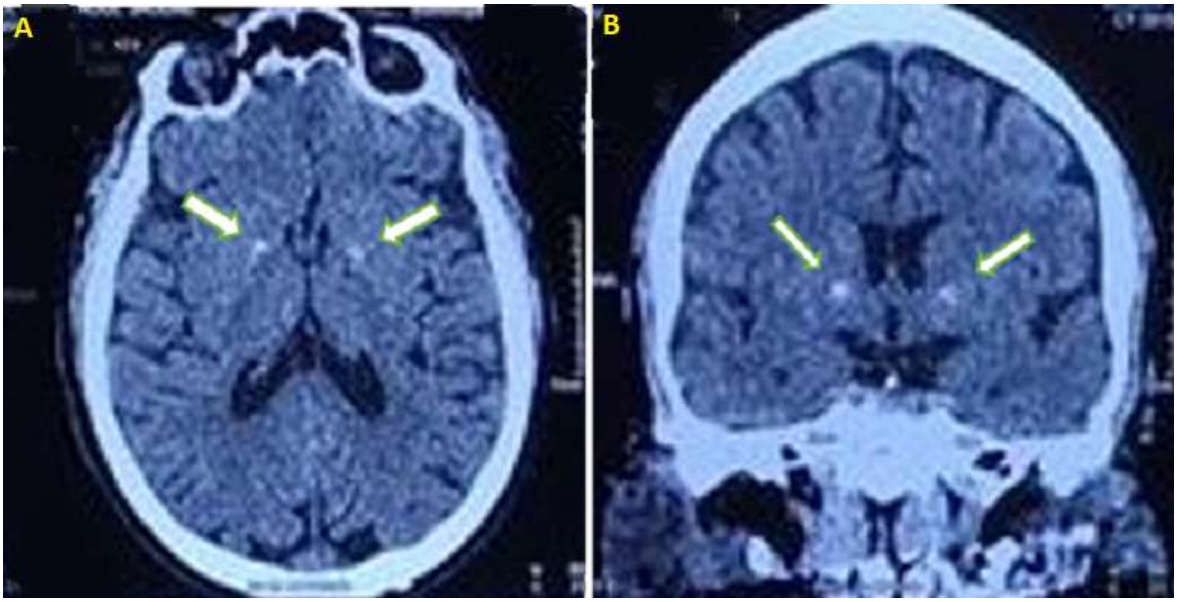

Figure 1: noncontrast CT of the brain, symmetric calcifications of basal ganglia can be observed in a axial and b coronal sections