Article published online: 2022-09-14

Case Report

Chronic Subdural Hematoma Associated with Fahr Syndrome: A Clinical Association or Just a Simple Coincidence?

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Abstract

The Fahr syndrome (FS) is a rare degenerative neurological disorder (its prevalence is <0.5%). FS is distinguished by the presence of abnormal bilateral intracranial calcifications with a predilection for the basal ganglia, also presented by movement disorders such as parkinsonism, paresis, and speech disorders. Chronic subdural hematoma (CSH), which is typically the result of mild head trauma, is a regularly encountered condition in elderly. A 63-year-old man has referred to our clinic from another hospital with a history of mild head trauma approximately a month ago. At the time of admission, the patient's Glasgow Coma Scale point was 15 points. In the history, there was only mild ataxia and right-sided hemiparesis. The laboratory examination revealed no electrolytes level abnormalities and normal endocrinal test examinations. Computed tomography revealed bilateral calcifications of basal ganglia, dentate nuclei which were misinterpreted as intracerebral contusion; with CSH of left temporal and parietal region. The hematoma was evacuated by burr-hole drainage. The patient was discharged 5 days after the surgery. The pathophysiology of FS is still unrevealed. There are some suggestions such as secondary to local disturbance of blood-brain barrier or a calcium neuronal metabolism disorder. However, on the other hand, local blood-brain barrier disturbance would also take part in CSH pathology. We hypostasized that patients with the history of FS, who had mild head traumas, might prone to subdural collections. On the other hand, FS and CSH coexistence is very unusual. Neurosurgeons might keep in mind FS when bilateral calcifications are seen in a patient.

Keywords: *Blood-brain barrier, chronic subdural hematoma, coincidence, Fahr syndrome*

Introduction

The radiological examinations of cranium revealed that accidentally found intracranial calcifications are 0.3–1.2% of routine examination.[1] Though intracranial calcifications, which are incidentally detected on computed tomography scan of head, are mostly benign requiring no follow-up such as calcification of pineal body and choroid plexus.

The Fahr syndrome (FS), that is a form of degeneration, is one of the rare neurological disorder. This syndrome can be distinguished if there are abnormal bilateral intracranial calcifications, especially in the areas of the basal ganglia, lateral thalamus, and cerebral cortex. FS is also presented with movement disorders such as parkinsonism, dystonia, and chorea.[1]

In elderly patients, chronic subdural hematoma (CSH) is a common disease.[2] In the present case, we report a FS patient with the history of minor head trauma, who had CSH and bilateral intracranial calcifications, which are misinterpreted as intracerebral contusions. We hypostasized that there could be a clinical association between CSH and FS.

Case Report

A 63-year-old right-handed man has referred to our clinic from another hospital with a history of mild head trauma approximately a month ago. At the time of admission to our hospital, the patient was alert and oriented. Glasgow Coma Scale point was 15 points. Physical examination revealed only mild ataxia, right-sided hemiparesis, and right-sided hemihypesthesia at the time of admission. The laboratory examinations revealed normal electrolytes levels with normal endocrinological, coagulation, and hematological tests. In patient's history, there is no coagulopathy, no anticoagulant therapy including the use of aspirin, and no thrombocytopenia. In the evolution of computed tomography and magnetic resonance imaging of brain revealed that

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How to cite this article: Gurcan O, Gurcay AG, Kazanci A, Goker T, Eylen O, Turkoglu OF. Chronic subdural hematoma associated with fahr syndrome: A clinical association or just a simple coincidence?. Asian J Neurosurg 2018;13:90-2.

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Access this article online

Website: www.asianjns.org

DOI: 10.4103/1793-5482.224831

Quick Response Code:

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bilateral calcifications of basal ganglia, dentate nuclei, and cerebellum, which were misinterpreted as intracerebral contusion, with CSH of left temporal and parietal region [Figures 1 and 2].

The subdural hematoma was evacuated by burr-hole drainage. In the postoperative observation, the patient was alert and oriented. There were no additional deficits observed. The patient was discharged 5 days after the surgery performed. There was neither neurological deficit nor endocrinal deficit observed at the time of discharge.

Discussion

The FS that was first described by Karl Theodor Fahr in 1930 is an unusual degenerative neurological disorder whose prevalence is probably <0.5%.[3]

The FS identifies many conditions which result in metastatic or bilateral dystrophic calcification in the basal ganglia. For example, endocrinal disorders such as parathyroid disorders (hypoparathyroidism, pseudohypoparathyroidism, and hyperparathyroidism), toxic storage disorders (CO and Pb intoxication, hypervitaminosis D, and radiotherapy), inflammatory disease (cytomegalovirus infection, neurocysticercosis, and neurobrucellosis), hypoxic and vascular problems (calcified infarct and ischemic encephalopathy), metabolic and degenerative disorders (senility, mitochondrial encephalopathy, leukodystrophic diseases, idiopathic familial, and carbonic anhydrase deficit), and others.[1]

Both familial and sporadic cases have been previously reported. A locus on chromosome 14q has been identified as associated with FS.[4] FS may exhibit movement disorders such as parkinsonism, chorea, tremor, dystonia, and speech disorders and psychiatric conditions such as psychosis or dementia.[1] In our case, the patient revealed no sign of movement disorders. The patient does not have a family history of the FS. The onset age of clinical symptoms is usually between 30 and 60 years.[1] Our patient was 63-year-old. This age is also in the age group of CSH.

The incidence of CSH is approximately three per 100,000 people, but it is more frequent among the elderly. CSH is a regularly encountered condition in neurosurgical practice.[2] CSH is typically the result of mild head trauma. There are multiple risk factors of CSH including coagulopathy and anticoagulant therapy including the use of aspirin, thrombocytopenia, and head injury. Our patient had not a history of usage of medications prior to admission. On the other hand, alcoholism, liver cirrhosis, hematologic disease, and chronic renal failure are also familiar causes of CSH.[5] In our patient history, he had a mild head trauma approximately a month ago but no other risk factors have been evaluated such as alcoholism or chronic disease.

We performed burr-hole drainage for treating the patient so as the population suffering from CSHs includes many elderly patients with co-morbid medical conditions though less invasive surgical techniques such as burr-hole drainage or twist drill craniotomy with closed-system drainage have become the initial procedures of choice.[2]

The pathophysiology of the FS is still unrevealed. There are some suggestions that the intracerebral calcium deposit is a kind of metastatic deposit type, secondary to local disturbance of blood-brain barrier, or to a calcium neuronal metabolism disorder.[1] In our patient, we did not observe calcium metabolism disorder. However, on the other hand, local blood-brain barrier disturbance would also take a part in CSH pathology. FS and CSH coincidence is very unusual, so it is difficult to hypostasize that patients with the history of FS, who had mild head traumas, are prone to subdural collections. Clinicians must keep in mind who are taking care of FS that intracranial hematomas and FS coexistence is very unusual in neuroscience practice, but there are some reports of cases mentioning this coexistence.

As we review English literature for FS and intracranial hemorrhages; we found the case report of Al-Jehani *et al*.

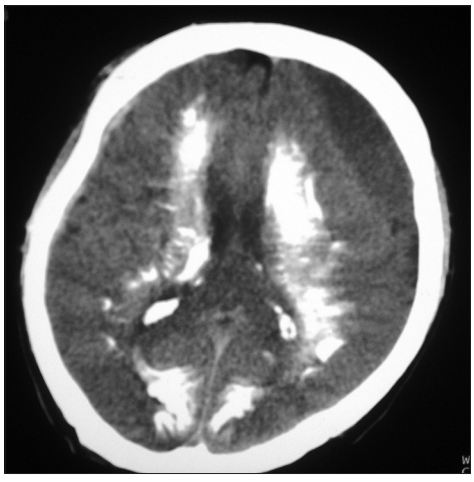

Figure 1: Computed Tomography of patient bilateral intracerebral calcifications and left parietal side subdural hematoma (axial)

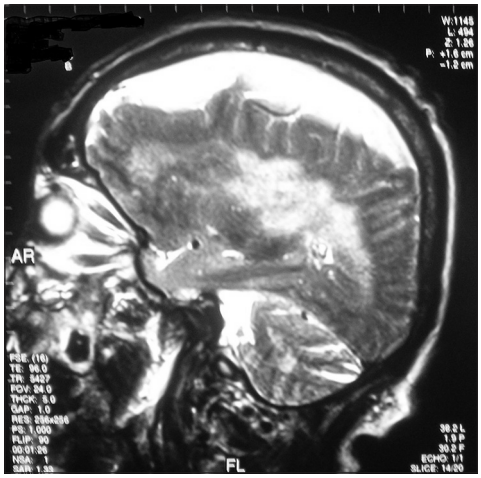

Figure 2: Magnetic resonance imaging of patient (sagittal)

that 54-year-old woman, who had subarachnoid hemorrhage from the possible source of right posterior communicating artery aneurysm, had FS and had been operated for the aneurysm,[3] and a case report of Swami and Kar that FS coexisted with mid-brain hemorrhage of a 45-year-old man.[6] Eroğlu *et al.* also reported a patient with FS coexistence with cerebral aneurysms.[7]

In the literature, there are several reports that emphasize familial inheritance of familial form of Fahr disease.[8-10] Kotan and Aygul reported a familial Fahr disease revealing that the disease was an autosomal dominant trait.[8] Bobek and Nowak also reported a familial form of FS but they were unable to suggest a probable type of inheritance.[9] In our case, there was no familial history of FS.

In a patient with the history of mild head trauma, when intracranial calcifications observed in computed tomography, can be misinterpreted as intracerebral hematoma or contusion. To our knowledge, our case is the first report in English literature that chronic subdural hematoma and FS occurs together. When we review the literature, it is not yet possible to link FS to intracranial hemorrhage or CSH. The connection between FS and intracranial hemorrhage is possibly incidental. Further reports and studies are needed to state the relationship between FS and intracranial hemorrhages. Tough the FS and CSH coexistence is very unusual in neuroscience practice, we might keep in mind FS when bilateral calcification seen in the basal ganglia in a patient with CSH.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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