

CASE REPORT

Idiopathic brain calcification accompanied by intracranial hemorrhage, a challenging diagnosis

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ABSTRACT

Acute subdural hematoma is a neurosurgical emergency. Emergency and a serious trauma with high mortality rates. Idiopathic brain calcification, also called bilateral striopallidodentate calcinosis, is rare disease characterized by symmetric cerebral calcifications. Calcifications are demonstrated on computed tomography (CT) usually taken for other reasons. CT findings in these cases may be confused with subarachnoid hemorrhaging. A 79-year-old female patient was admitted to the emergency room presenting with seizure and confusion. She had a history of atrial fibrillation for which she was using anticoagulants. On physical and neurologic examinations, she was confused with limited orientation and cooperation. Her Glasgow Coma Scale score was 10. Her serum hemoglobin was 9.2 g/dl and her international normalized ratio was 5.02. Her cranial CT revealed bilateral striopallidodentate calcinosis accompanied by an acute subdural hematoma on a bilateral chronic basis and this case was evaluated in light of literature.

Key words: idiopathic brain calcification, intracranial hemorrhage, warfarin overdose, Fahr's syndrome.

INTRODUCTION

"Idiopathic brain calcification" is characterized by idiopathic calcifications in the basal ganglia, cerebellar dentate nucleus and centrum semiovale. It is usually familial and autosomal dominantly inherited, although sporadic and autosomal recessive forms have been reported. It was first described by Delacour in 1850 then by Fahr in 1930. So the term Fahr's disease is used interchangeably for this condition. Clinical findings usually is evident after 30 and include dementia and mood disorders in addition to parkinsonism, dystonia, tremor, chorea and ataxia. Due to frequent use of brain CT and similar imaging patterns, it may be overlooked or confused with intracranial hemorrhages.

CASE REPORT

A 79-year-old female patient was admitted to our clinic due to confusion developing after an epileptic seizure. The patient, previously diagnosed with atrial fibrillation, Alzheimer's disease, chronic renal failure and Fahr's disease was using warfarin, donepezil and olanzapine. On her neurologic examination, she was confused, orientation-cooperation were limited, her Glasgow Coma Scale score was 10, her pupils were bilaterally myotic and her direct and indirect light reflexes were bilaterally positive. She had ecchymosis on the anterior aspect of her right forearm. Laboratory findings were as follows: serum hemoglobin: 9.2 g/

dl, international normalized ratio: 5.02, total calcium: 6.6 mg/dl, parathyroid hormone: 10.09 pg/ml (Normal: 7-55 pg/dl), urea: 27.9 mg/dl, creatinine: 0.8 mg/dl, glucose: 96 mg/dl, sodium: 140 mEq/l, potassium: 3.9 mEq/l, phosphorus: 5 mg/dl and magnesium: 2 mg/dl, thyroid, liver function test, urinary calcium were within normal range. A cranial computed tomography (CT) was obtained due to the suspicion of a possible intracerebral hemorrhage as she had been using warfarin. On cranial CT imaging, multiple calcifications were observed in the cerebral white matter, ganglia, bilateral thalamus, hippocampus and cerebellar dentate nuclei which were evaluated in favor of idiopathic brain calcification (Fig. 1).



Figure 1, Cranial CT imaging. Yellow arrows: bilateral chronic subdural effusions, red arrows: acute subdural hematoma on a bilateral chronic basis, green arrows: multiple calcifications

In addition, hyperdensities consistent with subdural

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hemorrhage and chronic subdural effusions were seen in both hemispheres. The clinical, laboratory and radiologic findings were evaluated as bilateral subdural hemorrhage accompanying brain calcification. The patient was consulted with the neurosurgery clinic and the treatment and follow up of the patient were planned with the associated clinics.

DISCUSSION

Calcifications are usually bilateral in idiopathic brain calcification which is also known as familial idiopathic basal ganglion calcification. Striopallidodentate calcifications are usually seen with calcium-phosphorus metabolism disorders. However, in some cases without any abnormalities in serum calcium and parathyroid hormone levels, extensive calcifications can be seen in brain parenchyma as the result of genetic disorder.^{1, 2} Bilateral symmetrical cerebral calcifications are most commonly seen in idiopathic, familial or calcium metabolism disorders. Direct radiographies may rarely show severe calcifications. Cranial CT is the gold standard for diagnosis as even small calcifications can be demonstrated.³ Hounsfield or CT numbers is a useful tool in differential diagnosis. On magnetic resonance imaging, while calcifications may be hypo- or hyperintense in T1-weighted images, they are seen as hypointense in gradient-weighted images.⁴ In our case, the symmetrical calcifications on CT imaging were consistent with idiopathic brain calcification (Fig. 1). At the same time, this case also had acute bilateral subdural hematomas developed on a chronic basis. These two diseases rarely coexist and few reports have been published in literature. Over time, the use of anticoagulants grows with increases seen in diagnosis of atrial fibrillation.⁵ Most authors believe that serum calcium, phosphorus and parathyroid hormone level should all be within normal level in idiopathic brain calcification (Fahr's disease)⁶, however, others indicate an association with abnormal calcium level and parathyroid hormone is present in some cases (Fahr's syndrome).⁷

Warfarin-related intracranial hemorrhage is a rare but important problem (yearly 0.2-3%). The mortality rate may be as high as 50% in intracranial hemorrhages.⁸ Acute subdural hematoma is also a cause of cranial hemorrhage. The real incidence of acute subdural hematomas is unknown due to its rapid resolution and redistribution. The spontaneous resolution and redistribution of acute subdural hematoma is rare and its mechanism is not well understood.⁹ Cerebral calcifications are usually detected before the onset of symptoms which usually begin in 4th and 6th decades of life. Although rare, pediatric cases have been reported in literature.¹⁰ For patients admitted to the emergency room with head traumas, the discrimination between hemorrhage and calcification should be made by evaluating localizations and measuring densities on CT images (Hounsfield number). Thus,

for the patients with head trauma, in the absence of any known cause, this condition should be considered in the differential diagnosis when calcifications are detected in the basal ganglia, cerebellum and supratentorial region on cranial CT or MRI.

CONCLUSION

Idiopathic brain calcification is a rare and usually familial cause of basal ganglia calcifications. It is an intracranial pathology that can be easily overlooked, since it is frequently asymptomatic. The coexistence of idiopathic brain calcification and intracranial hemorrhage is quite rare and should be discriminated in clinical practice.

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