Brainstem Involvement in a Patient With Fahr’s Syndrome

Dilcan Kotan1, Saadet Sayan2

ABSTRACT
In cases of Fahr’s syndrome, calcium and other minerals are stored bilaterally, and almost always symmetrically. These minerals are typically stored in the basal ganglia, thalamus, centrum semiovale, cerebellar dentate nucleus, and periventricular white matter. Here, we present a rare case of Fahr’s syndrome in a female patient with unusual calcifications in the central pons.

Keywords: Fahr’s syndrome, intracranial calcification, brainstem involvement

FAHR SENDROMU BİR OLGUDA BEYİN SAPI TUTULUMU

ÖZET

Anahtar sözcükler: Fahr’s sendromu, intrakranyal kalsifikasyon, beyin sapı tutulumu

Fahr’s syndrome is a rare disease with multiple psychiatric and neurological manifestations. The most common neurological signs are headache, vertigo, syncope, movement disorders, and seizures (1,2). Within the basal ganglia, the globus pallidus is the most frequent site of calcification, but deposits may be present in the putamen, caudate nucleus, internal capsule, dentate nucleus of the cerebellum, thalamus, and the periventricular white matter (2,3). In our first case, we considered secondary hypoparathyroidism as the causative etiological factor since there was also pons calcification as determined by the brain CT scan.

Case report
A 63-year-old female patient was admitted to the emergency room during the postictal period after having experienced generalized seizures in four-hour intervals. During the neurological examination, the patient was in the altered states of consciousness
Figure 1. Computed tomography showing calcification in the bilateral basal ganglia, thalamus, periventricular white matter (A), cerebellar dentate nucleus (B), and pons (C).

Discussion

Fahr’s syndrome is associated with a variety of other diseases but no specific etiologic agent has been identified yet. Suggested possible causes for this disorder include calcium metabolism, inflammatory, and vascular disorders. Other possible causes include tumoral conditions, encephalitis, systemic diseases, anoxia, radiation, genetic disorders, and various toxins (2,4). Our case was related to a parathormone metabolism disorder. Etiology was not directly correlated with image calcification patterns. Fahr’s syndrome rarely presents during childhood or adolescence, and the usual age of presentation is during the fourth to sixth decades of life (2). Our case was in her seventh decade and based on the clinico-radiological and biochemical findings, the diagnosis of Fahr’s syndrome due to secondary hypoparathyroidism was strongly indicative (6). Several bilateral symmetrical calcifications of the basal ganglia, thalamus, periventricular white matter, cerebellar nuclei have been described following Fahr’s original description in 1930. CT scanning is an easy test with maximum sensitivity, and allows the easy diagnosis of Fahr’s syndrome (3,4). Pontine calcification has rarely been reported in the literature; moreover, the reasons for focal calcium accumulations in the pons are currently unknown (3). According to the radiological findings, our case was the first described in which the pons involvement to be secondary to hypoparathyroidism. The treatment for Fahr’s syndrome was directed at the identified cause, particularly the hypoparathyroidism. In other cases, symptomatic or conservative therapy with clinical observation is the rule. The prognosis is variable and difficult to predict. Our case was medicated and followed in terms of Parkinsonism and secondary hypoparathyroidism. The patient had no electroencephalogram (EEG) abnormalities. The patient was started on treatments with levodopa, and received regular follow-up care through outpatient services.
References