Restless legs syndrome in a person with Fahr's disease

Esra E. Okuyucu, MD, Tugba Tunc, MD,
Sinem Karazincir, MD, Taskin Duman, MD.

Bilateral striato-pallido-dentate calcinosis (Fahr's disease) is known as bilateral symmetric calcification, mainly of the basal ganglia and the dentate nucleus of the cerebellum.[1] The clinical features can be varied. The diagnosis is established by CT or MRI of brain and ruling out calcium metabolism abnormalities and developmental defects. Restless legs syndrome (RLS) is a common neurological sensorimotor disorder that encompasses an idiopathic form of genetic or unknown origin and symptomatic forms are due to many causes. The current understanding of the pathophysiology of RLS suggests the involvement of iron metabolism and dopaminergic dysfunction.[2] We present a patient with RLS associated with striopallidodentate calcification.

A 52-year-old right-handed female patient presented with a 3-year history of episodic, uncomfortable feelings in her legs while sitting or lying down especially in the evening or at night. When she experienced these uncomfortable sensations, she urgently needed to move her legs. Because of these unacceptable leg movements, she had been unable to sleep at nights for 3 years. Her mother had a similar history, with problems and difficulty in sleeping at nights. Neurological and physical examinations were normal. There was no extrapyramidal soft sign and neuropsychiatric abnormalities. Routine blood and urine tests, thyroid hormone level, blood ferritin and iron level, folic acid and vitamin B12 levels, blood ceruloplasmin levels, blood and urine copper levels were normal. Serum calcium, phosphorus and parathormone levels were also normal. Cerebrospinal fluid examination disclosed normal protein and glucose content with no cells. A search for antinuclear antibodies, anticardiolipin antibodies, lupus anticoagulant, anti-HIV, anti-Epstein-Barr Virus, and anti-cytomegalovirus antibodies were negative. The EEG and nerve conduction studies were normal. Sleep examination by polysomnography showed in the early part of the night there were jerks known as periodic limb movements (PLMS). Radiological and echographic screening and blood tests for occult neoplasm were negative. A CT showed bilateral calcifications in the basal ganglia, thalamus, caudate nucleus, centrum semiovale, and dentate nucleus of the cerebellum (**Figure 1**). On MRI, calcified lesions produce various signal intensities. The T2-weighted images showed slightly hyperintense lesions in the bilateral basal ganglia, thalamus, centrum semiovale, and hypointense lesions in the caudate nucleus. The T1-weighted images showed hyperintense lesions in the bilateral basal ganglia, thalamus, and caudate nucleus (**Figures 2a & b**). The diagnosis of RLS was made in view of her typical symptoms. She was prescribed cabergoline 2 mg to be taken at night. She has now experienced an improved situation, regarding both the symptoms of RLS and sleeplessness.

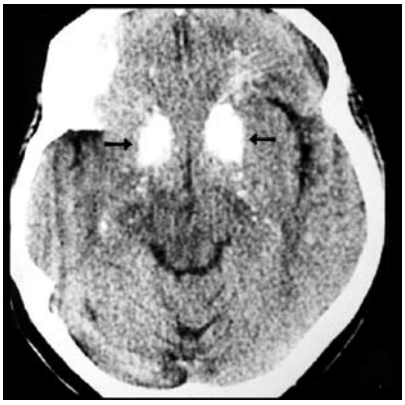

Figure 1 - An unenhanced axial CT scan shows calcified lesions in, basal ganglia, caudate nucleus, and centrum semiovale.

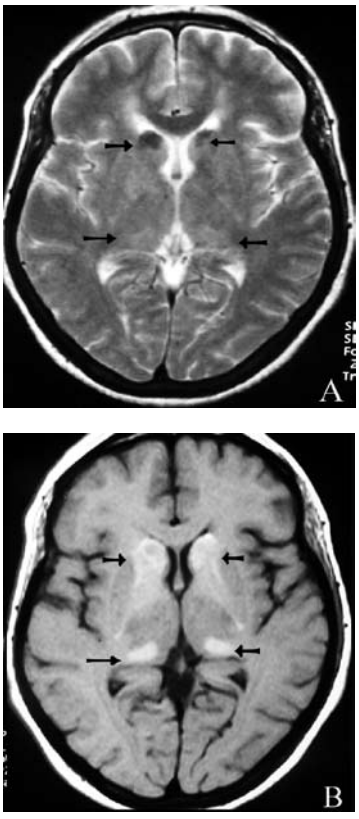

Figure 2 - **A)** Axial T2 weighted MRI shows slightly hyperintense lesions in the bilateral basal ganglia, thalamus, and hypointense lesions in the caudate nucleus. **B)** Axial T1 weighted MRI shows hyperintense lesions in the bilateral basal ganglia, thalamus, and caudate nucleus.

The features of Fahr's disease can be varied, and the diagnosis is established by obtaining a CT or MRI scan of the brain and ruling out abnormalities of calcium metabolism defects. Hypoparathyroidism is the major differential diagnosis. Serum calcium and parathormone levels help us to distinguish between idiopathic and secondary brain calcification. Causes of secondary striopallidodentate calcinosis can be endocrinologic, developmental, connective tissue disorders, or toxic.[3] The differential diagnosis of intracerebral calcification must include rubella encephalitis, toxoplasmosis, cytomegalovirus infection, HIV infection, tuberculoma, tuberous sclerosis, and brain tumors. The EEG and nerve conduction studies are usually normal in Fahr's disease.[4,5] The RLS is a common CNS disorder with a significant impact on resting and sleeping. The diagnosis of RLS mainly depends on the patient's history. Iron deficiency is the most common cause of symptomatic RLS. The diagnosis can be confirmed not only by the response to dopaminergic medication but also by polysomnography. Polysomnographic recordings have revealed the presence of PLMS in most of the patients presenting with symptoms of RLS. In our patient, we ruled out secondary RLS by laboratory tests and electrophysiological examinations. She had all 4 criteria that were established by the International RLS Study Group, and dopaminergic therapy was able to reverse the daytime and nighttime symptoms.[3] For a differential diagnosis, we used cranial MRI scan. According to the image on cranial MRI and CT, we understood that the main diagnosis of this patient was bilateral striopallidodentate calcification. The secondary striopallidodentate calcification and differential diagnosis of intracerebral calcification was ruled out by laboratory findings. Bilateral striopallidodentate calcification can share the features of RLS. The response to dopaminergic medication played a significant part in management. Both syndromes tend to be idiopathic. Whether RLS is the beginning of the disease or it is simply an incidental associated disorder is not clear. Only long follow up will clarify this matter.

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From the Departments of Neurology (Okuyucu, Duman) and Radiology (Karazincir), Faculty of Medicine, Mustafa Kemal University, Antakya, Hatay, and the Department of Neurology (Tunc), Ankara Research and Training Hospital, Ankara, Turkey. Address correspondence and reprint requests to: Ass. Professor E. Esra Okuyucu, Department of Neurology, Faculty of Medicine, Mustafa Kemal University, Antakya (Antochia), Hatay, Turkey. Tel. +90 (326) 2551405. Fax. +90 (326) 2144977. E-mail: esraokuyucu@yahoo.com

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