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IMAGING FINDINGS OF FAHR'S SYNDROME MAY VARIABLE

Fahr Sendromunda Görüntüleme Bulguları Değişken Olabilir

Murat Gültekin[1], Serkan Şenol[2], Burak Kütük[1], Tayfun Turan[3], Meral Mirza[1]
1 .Erciyes University, Faculty of Medicine, Neurology Department
2. Erciyes University, Faculty of Medicine, Radiology Department
3. Erciyes University, Faculty of Medicine, Psychiatry Department

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ABSTRACT

Although Fahr's syndrome (FS) was defined a long time ago, it is a rare disorder that may be overlooked in differential diagnosis. Neuropsychiatric disorders such as dystonia, parkinsonism, cognitive dysfunction, depression, schizoid disorders, obsessive-compulsive disorder and personality disorders are frequently seen. A female patient, 52 years old, was admitted to making differential diagnosis for secondary parkinsonism. There was revealed FS as cause of secondary parkinsonism. A male patient, 73 years old, was admitted due to manic like symptoms and motor agitation. The radiological findings of patient's was consistent with the FS. FS diagnosis is based on the evaluation of three main elements, including bilateral idiopathic non-atherosclerotic calcification of the basal ganglia, psychiatric symptoms, main movement disorders and neurological findings. In this paper, we report two FS cases whose standard magnetic resonance (MR) images were normal and who therefore were given a late diagnosis.

Key Words: Fahr's syndrome, computed tomography, magnetic resonance imaging

ÖZET

Fahr sendromu (FS) uzun zaman önce tanımlanmış olmasına rağmen, nadir görülen bir hastalık olduğundan ayırıcı tanıda gözden kaçabilmektedir. FS'de klinik semptomlar çok çeşitlidir. Distoni, parkinsonizm, zihinsel işlev bozukluğu, depresyon, şizoid bozukluk, obsesif kompulsif bozukluk ve kişilik bozuklukları gibi nöropsikiyatrik hastalıklar sık görülür. 52 yaşında kadın hasta parkinsonizm ayırıcı tanısı için başvurdu. Yapılan incelemede sekonder parkinsonizm sebebi olarak FS saptandı. 73 yaşında erkek hasta manik benzeri semptomlar ve motor huzursuzluk nedeniyle başvurdu. Yapılan incelemede FS ile uyumlu radyolojik bulgular saptandı. FS tanısı, bazal ganglionların bilateral idiyopatik non-aterosklerotik kalsifikasyonu, psikiyatrik belirtiler ve başlıca hareket bozuklukları olmak üzere nörolojik bulgular dahil üç ana unsurun değerlendirilmesi temeline dayanmaktadır. Bu yazıda; standart kraniyal manyetik rezonans (MR) görüntülemesi normal olan ve bu yüzden geç tanı alan iki FS olguyu sunuyoruz.

Anahtar Kelimeler: Fahr sendromu, bilgisayarlı tomografi, manyetik rezonans görüntüleme

INTRODUCTION

Although Fahr's syndrome (FS) was defined a long time ago, it is a rare disorder that may be overlooked in differential diagnosis. This often occurs especially when images specific to FS cannot be obtained. Patients with FS exhibit a wide variety of clinical symptoms. Neuropsychiatric disorders such as dystonia, parkinsonism, cognitive dysfunction, depression, schizoid disorders, obsessive-compulsive disorder and personality disorders are frequently seen (1). FS diagnosis is based on the evaluation of three main elements, including bilateral idiopathic non-atherosclerotic calcification of the basal ganglia, psychiatric symptoms, main movement disorders and neurological findings (2). In this paper, we report two FS cases whose standard magnetic resonance (MR) images were normal and who therefore were given a late diagnosis.

CASE

A 52-year-old female patient was admitted to the our clinic with a diagnosis of idiopathic Parkinson's disease (PD) that had been present for five years. In her anamnesis, it was learned that she had been monitored in various foreign centers, had undergone many cranial MRIs and her PD diagnosis remained unchanged. In addition, she had experienced difficulty in walking had, delusions, heard voices, had feelings of suspiciousness and, introversion and sometimes agitation symptoms over the last year. She had been examined for these complaints and was being treated with the diagnosis of psychotic depression. Her examination revealed widespread parkinsonism symptoms and negativism. Her cranial MR report was normal. (Figure 1a, 1b). Her present findings suggest that the patient is showing symptoms of secondary parkinsonism. Therefore, her past medical records were reconsidered. All her cranial MR reports were found to be normal. However, it was noted in her computed tomography (CT) report between 2011 and 2013 that she had bilateral calcification in the globus pallidus. (Figure 2), and this was not noticed. A new CT was requested to see the prevalence of calcification (Figure 3a, 3b). SWI images showed signal changes compatible with bilateral calcifications in the globus pallidus. Endocrine disorders including parathyroid hormone and calcium metabolism, infectious agents and other disorders were not detected in the differential diagnosis of the patient. As a result,the patient was diagnosed with idiopathic basal ganglia calcification in the etiology of FS.

A 73-year-old male patient was admitted to our clinic with manic like symptoms, euphoria, spending a lot of money and irritability. Also, it was learned that he had more difficulty in making logical decisions than before. His neurological examination revealed no abnormality. were detected in his psychiatric examination. It was learned that he had undergone thyroidectomy surgery because of goiter history three years previously. Moderate cortical atrophy was observed in his cranial MR report. (Figure 4a, 4b). Due to the thyroidectomy history of the patient and subsequent development of neuropsychiatric symptoms, a cranial CT was requested. Bilateral basal ganglia millimetric punctate calcification was observed in CT (Figure 5a, 5b). Also, all his the endocrine tests were found to be normal and other disorders were not detected in the differential diagnosis of the patient.

Corresponding Author: Murat GÜLTEKİN, Erciyes University, Faculty of Medicine, Neurology Department **Email:** gultekin@erciyes.edu.tr

Figure 1a, 1b: T1-weighted and T2-weighted brain MR images are normal.

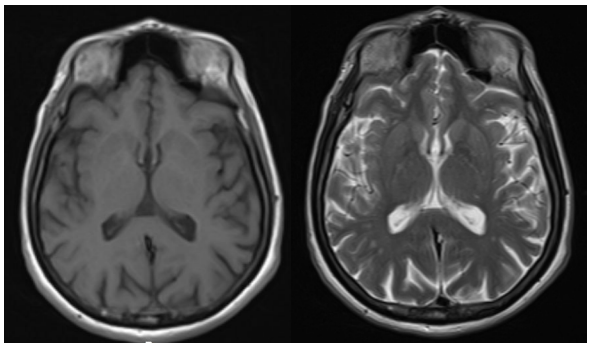


Figure 2: Bilateral calcifications in the globus pallidus are present in the CT image

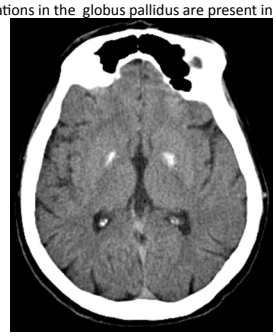
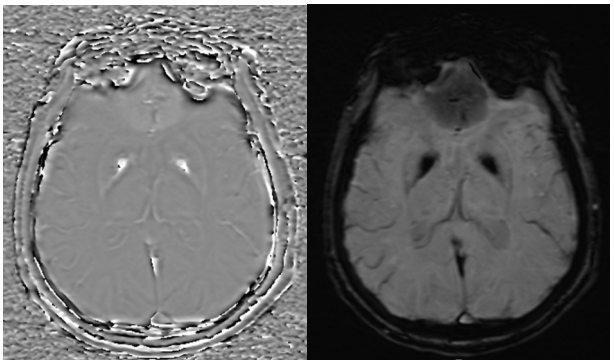


Figure 3a, 3b: SWI phase and magnitude images show signal changes compatible with calcifications.



DISCUSSION

Cranial MR and CT images of FS may vary depending on the progression of the disease and intensity of the calcium deposit (Table). Susceptibility-weighted imaging (SWI),which is superior to standard MR sequences in the detection of blood products, iron and calcification within the brain is a 3D gradient-weighted MR sequencing method, that has been recently introduced and consists of phase and magnitude images. Therefore, it has been used in the differential diagnosis of neurodegenerative diseases. It is highly effective in displaying calcification (3). Although the cranial MR images of our patients were normal, a significant signal increase in SWI MR images was diagnostic (Figure 6a, 6b). In the literature, there are many reports of FS cases in which the cranial (MR) images were normal and patients were diagnosed late (4, 5, 6). In routine MR images, imaging may vary for these areas of calcification which can contain zinc, magnesium, iron, mucopolysaccharides or blood components in routine MR images. Therefore, it is possible to establish areas of calcifications within the brain more clearly with SWI sequencing in addition to MR scans (3).

In conclusion, standard MR images of FS may vary according to disease phase depending on the progression of the disease and intensity of the calcium deposits. Therefore combined MR-SWI is a more ideal option to clearly show calcium accumulation and for the differential diagnosis of possible metabolic inflammatory conditions.

Conflict of Interest

The authors declare no conflict of interest.

Figure 4a, 4b: T1 and T2 moderate cortical atrophy is observed in brain MR images.

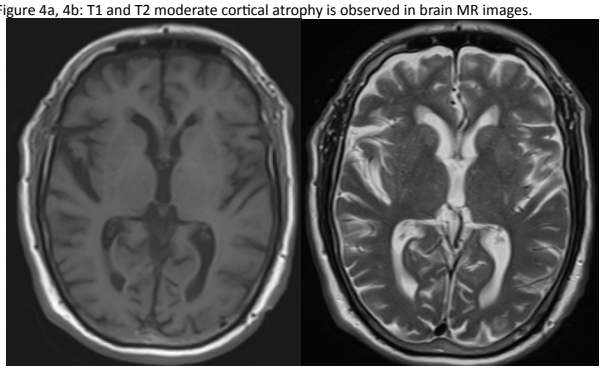


Figure 5: Bilateral millimetric punctate basal ganglia calcifications are observed in CT image.

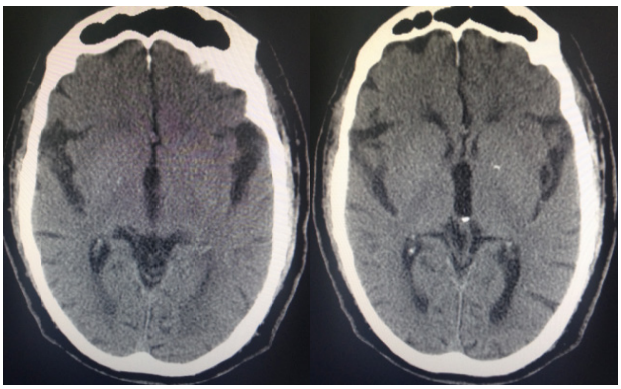


Figure 6a, 6b: SWI phase and magnitude images show signal changes compatible with calcifications.

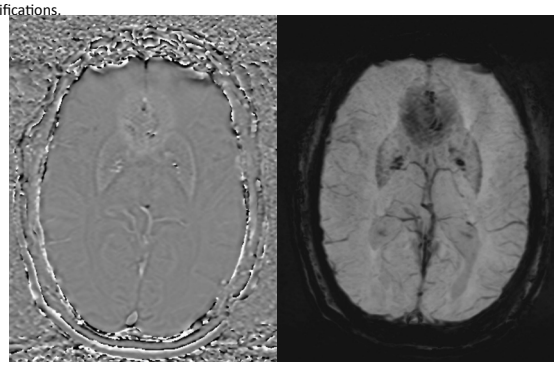


Table: MRI and CT features of Fahr's disease

CT	MR-T2A	MR-T1A	MR-SWI FAZ	MR-SWI MAGNITUD
Hyperdense	Hypointense	Hypointense	Hyperintense	Hypointense
	Normal	Hyperintense		
		Normal		

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