CASE REPORT

# Unexplained neuropsychiatric symptoms in intensive care: A Fahr Syndrome case

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## Abstract

Fahr Syndrome is a rare disease where calcium and other minerals are stored bilaterally and symmetrically in the basal ganglia, cerebellar dentate nucleus and white matter. Fahr Syndrome is associated with various metabolic disorders, mainly parathyroid disorders.

The presented case discusses a 64-year old male patient admitted to the intensive care unit of our hospital diagnosed with aspiration pneumonia and urosepsis. The cranial tomography examination to explain his nonspecific neurological symptoms showed bilateral calcifications in the temporal, parietal, frontal, occipital lobes, basal ganglia, cerebellar hemisphere and medulla oblongata posteriorly. His biochemical test results also indicated parathormone-calcium metabolic abnormalities.

Fahr Syndrome must be considered for a definitive diagnosis in patients with nonspecific neuropsychiatric symptoms and accompanying calcium metabolism disorders in order to control serious morbidity and complications because of neurological damage.

**Keywords:** Fahr syndrome, Calcification, Intensive care.

## Introduction

Fahr Syndrome is a disorder associated with various metabolic derangements especially with diseases of the parathyroid gland.[1] Besides genetic etiology, developmental, metabolic, infectious, sporadic and other conditions may also cause the disorder. Clinically it is characterized with dementia and psychiatric symptoms in addition to parkinsonism, dystonia, tremor, chorea, and ataxia. Diagnosis is usually through a cranial tomography. Treatment is mostly symptomatic and based on the improvement of the calcium metabolism.[1] In this presented case, we share our intensive care approach to a patient with Fahr Syndrome who was admitted with aspiration pneumonia and urosepsis.

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## Case Report

The 64 year old male patient was seen in the emergency department of our hospital in November 2012 with difficulty in swallowing, inability to pass urine, fever and shortness of breath and involuntary body movements. He was admitted to the intensive care with diagnoses of aspiration pneumonia, urinary tract infection and sepsis.

The physical examination showed the respiration rate as 38/min., blood pressure 142/100 mmHg, pulse, 110 beats per minute, temperature 38.4°C and a dry oral mucosa. There was harsh vesicular breathing accompanied with rhonchi and crepitations in the left basal area. The patient had cystofix and his urine was pyuric. His chest x-ray showed bronchovascular clarification, non-homogenous infiltration symptoms in the middle and lower zones of the left hemithorax. The urine was leucocyte and nitrite positive.

Tremor, choreiform movements, dysarthric speech, Parkinsonism, dementia and depressive mood state findings were determined in the neurological examination of the patient, who was started on intravenous Meropenem 2x1gr daily for his infection.

It was discovered that there was diabetes, hypertension and kidney disease in his medical history. The neurologic complaints had started five years ago with rapid exhaustion, unbalanced walking with small steps and speech impediments. With the passage of time he started having seizures, tremors, advanced limitation of movements, inability to talk and forgetfulness.

**Table:** Laboratory values pre-treatment and 5th day of treatment.

| | Pre-treatment | 5th day of treatment |
|---|---|---|
| PTH (16-25pg/ml) | 1,9 | |
| 25 OH Vitamin D (5.2-47 ng/ml) | 3,2 | 3,78 |
| Total calcium (8.2-10.6mg/dl) | 7 | 8,08 |
| Ionized calcium (4.4-5.4 mg/dl) | 3,7 | 4,48 |
| Magnesium (1.6-2.6mg/dl) | 1,5 | 1,9 |
| Phosphorus (2.5-4.5 mg/dl) | 5 | 4,7 |

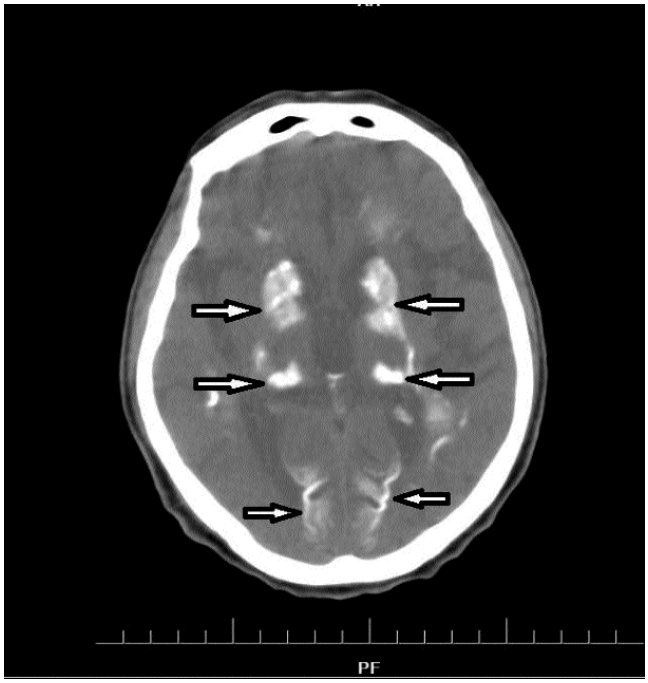


**Figure:** Bilateral calcifications in the thalamus, basal ganglia and cerebellum.

Treatment was started with Olanzapine 2x2.5 mg and sodium valproate 1x1000 mg but relief was not achieved.

The cranial tomography showed calcifications in both cerebellar hemispheres, cerebral lobes, basal ganglia and posterior medulla oblongata (Figure).

The case was evaluated as Fahr Syndrome associated with hypoparathyroidism in the light of the laboratory, clinical and radiological findings (Table). In addition to the antibiotherapy and non-invasive ventilation support for treating the pneumonia and urosepsis, his previous treatment of Olanzapine 2.5mg once daily and Sodium valproate 500 mg twice daily were continued on the suggestion of the Neurology Clinic. Calcium carbonate 100 mg 2x1, calcitriol 0.5mcq 1x1, vitamin D3 ampoule 300,000 IU 1x1 treatments were started as replacement for the patient, who had hypocalcaemia, PTH and vitamin D deficiency. As no change was expected in PTH levels in early stages, no measurements were taken (Table).

On the 7th day, as the patient no longer needed the non-invasive ventilation support, he was discharged to the palliative care center and offered neurology polyclinic control.

## Discussion

Genetically, Fahr disease is known to have an autosomal dominant inheritance, and families with the disorder have been reported.[1] No individuals with similar neuropsychiatric symptoms or who had been diagnosed with Fahr Syndrome were encountered in the family history of our patient.

Besides calcium metabolism disorder, hypoparathyroidism or pseudo hypoparathyroidism, various factors caused by genetic, developmental, metabolic, infectious, sporadic, anoxic and other conditions have been reported.[2,3] Our patient also showed calcium metabolism disorder (hypoparathyroidism, vitamin D deficiency) as verified by the laboratory results. Literature also cites cases where there are no abnormalities in the laboratory tests, and the diagnosis depends solely on the imaging methods.[4]

It is not yet clear whether the calcium deposits are the result of a disruption of the blood brain barrier or whether they are caused by neuronal calcium metabolism disorder.[2] It has been reported that calcium accumulation in Fahr Syndrome starts approximately 3 decades before clinical symptoms appear.[5] Although the symptoms usually start at the 4th to 6th decade, as with our patient, child cases have been rarely reported.[4,6,7]

If the calcifications are dense, extrapyramidal and cerebellar symptoms may arise. In chronic hypoparathyroidism, cognitive disorder, speech impediments, emotional lability, mental and sensation disorders, aches, irritability, cataracts, papillae oedema may be seen.[2,5,7,8] Cases are also known in literature where the patient presents with speech impediment and slowing down of thought, with no other neurological-systemic anomalies, who were diagnosed with imaging methods.[4] Our patient also displayed choreiform movements, gait disorder, tremor, dysarthric speech, slowing down of thought and movement, dementia and depression. Literature also mentions cases where classic symptoms are not observed.[9]

There is also a case that presented with an epileptic seizure, where, although treated for hypocalcaemia, the seizures persisted,[8] our patient's medical records also showed a history of seizures. After anti-epileptic treatment, no seizures were observed during the follow-up.

Tomography is superior to craniography and MR in determining calcified areas, however, it is believed that magnetic resonance is a valuable imaging method in definitive diagnostics.[4] As sufficient cooperation of our patient was not possible due to abnormal extremity movements and dementia, MR imaging was not performed. Diagnosis was based on laboratory results and

tomography findings.

In a definitive Fahr diagnosis, metabolically-infectious diseases, neurological disorders, subarachnoid haemorrhages and psychiatric events that might cause calcifications must be kept in mind. In our patient, symptoms started approximately five years ago, and he was admitted to our unit due to morbidity caused by progressing neurological deficits. These deficits were aspiration pneumonia secondary to dysphagia, urine flow disorder and urinary tract infection developed in association with residue. As a result of the evaluation of the patient upon admittance to the intensive care unit, the identification of the calcifications in the brain tomography planned as the imaging method upon the verification of neuropsychiatric symptoms, and the establishment of calcium metabolism disorder through the laboratory results, the diagnosis was confirmed.

Most Fahr syndrome cases progress symptomatically and they must be followed even when they are asymptomatic.[1] Gür, et al., discharged a Fahr syndrome patient that applied with complaints of tetany after his clinical findings improved following replacement of calcium and calcitriol and suggested polyclinic control.[9] Yürekli, et al., diagnosed a case presenting with dementia, and observed a reduction in the dementia symptoms after treatment.[10] Our patient also displayed neurological deficits due to widespread calcifications.

Replacement of calcium and vitamin D eliminates the metabolic anomaly, and clinically delays negative progression.[3] Together with the regulation of the calcium metabolism, various agents were tried for the treatment of Fahr syndrome, including Nimodipine which did not give satisfactory results. However, it was shown that disodium etidronat alleviated the symptoms.[2] Support treatment is generally directed to affected brain areas and symptoms. Asymptomatic family members must also be investigated.

## Conclusion

In conclusion, although Fahr syndrome is not a newly identified disorder, it is often not brought to mind, or overlooked because it is so rare. We are of the opinion that Fahr Syndrome must be considered in the definitive diagnosis of patients displaying nonspecific neuropsychiatric symptoms accompanied by calcium metabolism disorders in order to prevent high morbidity neurological damage.

**Disclosure:** No.

**Conflict of Interest:** No.

**Funding Sources:** No.

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