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Transient ischemic attack of the brain as the first link in a decompensatory event chain of cerebro- and cardiovascular incidents in a 68-year-old female patient with radiological features of Fahr's syndrome – a case report

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ABSTRACT

Introduction: Transient ischemic attack (TIA) is a reversible episode of neurologic deficit, that symptomatically can be similar to stroke, but lasts shorter than 24 hours and does not cause any radiological changes in brain images. TIA is a major risk factor for subsequent ischemic stroke. Fahr's syndrome is a rare, genetically conditioned incidence of calcified deposits in basal ganglia and cerebral cortex.

Case report: A 68-year-old female patient was admitted to the Department of Neurology due to an incident of left limbs weakness and left mouth angle drop. The symptoms lasted 5 minutes. The initial brain CT revealed no acute ischemic foci, although the radiologist found characteristic features of Fahr's syndrome. Carotid ultrasound showed stenosis of right internal carotid artery. The patient experienced an ischemic stroke in 6th day from initial symptoms. 4 days later a myocardial infarction occurred as well.

Discussion: TIA is a major prognostic factor for an ischemic stroke with the greatest risk of incidence in the first week after initial symptoms. Proper imaging diagnostics and prophylactic treatment with aspirin and statins should be administered. Patients after TIA incident should stay under watchful neurological supervision during the first 3 months after the attack.

Keywords: Ischemic Attack, Transient; Stroke; Myocardial Infarction; Fahr's Syndrome

INTRODUCTION

Transient ischemic attack (TIA) is a reversible episode of neurologic deficit, that symptomatically can be similar to a stroke, but lasts shorter than 24 hours and does not cause any radiological changes in brain images. The entity is caused by local central nervous system hypoperfusion. In most patients the incident typically lasts not longer than 1 hour. TIA is a powerful warning sign of a possible ischemic stroke coming [1, 2, 3, 4].

The risk of subsequent stroke after TIA can be stratified by scales such as the California score, the ABCD score or fusion of these two – the ABCD² score. There are five independent predictors of ischemic stroke after TIA which are:

1. Age > 60 years old
2. Diabetes
3. Symptoms duration > 10 minutes
4. Symptoms of weakness

5. Symptoms of speech impairment [5].

Although transient ischemic attack can manifest as a mild syndrome, syncope, isolated dizziness or global amnesia are the most often caused by other conditions, rather than real TIA [2].

Fahr's syndrome is a rare neurological disorder, where abnormal calcified deposits in basal ganglia, thalamus, hippocampus, cerebral cortex and subcortical white matter appear. The disease is genetically conditioned, but other factors play an important role either, such as endocrine disorders or infectious diseases [6].

CASE REPORT

A 68-year-old female patient was admitted to the Department of Neurology due to an incident of syncope with left mouth angle drop and weakness of left limbs. The condition lasted 5 minutes and was not remembered by the patient. The patient claimed that the day before admission she was discharged from neurology department in other hospital, as she stated – without any neurologic deficits (but she did not present any documentation from that hospital). The patient was suffering from hypertension, hypoparathyroidism (caused by strumectomy performed due to hyperthyroidic goiter), coxarthrosis and she previously had an ischemic stroke of the left hemisphere and myocardial infarction in the past. Physical examination on the day of admission revealed unstable Romberg's test, walking impairment, restricted hips mobility and abnormal curvatures of the spine. There was no evident paresis, nor facial nerve was affected. Brain CT performed in the A&E Department did not show any evidence for acute ischemic changes; however there were diffuse chronic ischemic changes found. Moreover, the CT revealed calcified deposits in subcortical nuclei, cerebellar hemispheres and deposits in occipital cortex as well. The radiologist stated the gained image is typical for Fahr's disease. However, the syndrome was not further diagnosed because it was not the cause of patient's current state. Besides, because of the endocrine problems, an exclusion criterion of absence of metabolic disorders was not fulfilled. Apart from the CT, the patient had ABPM Holter and also a carotid ultrasound performed. The examination showed stenosis (>90%) of right internal carotid artery. The patient got stroke prophylactic treatment: aspirin, statins, infusion liquids and hypotensive drugs. A vascular surgeon was also asked to see the patient. Unfortunately, on the 6th day of hospitalization, before the appointment with vascular surgeon, the patient experienced left-sided hemiparesis. Emergency brain angio-CT showed no new ischemic foci, though because of the carotid stenosis, the patient underwent arteriography with neuroprotected percutaneous carotid stenting. After the intervention the patient felt better and the symptoms

started to withdraw slowly. 4 days later, on the 10th day of hospitalization, the patient suffered from severe chest pain without changes in ECG but with troponin dynamics and was diagnosed with NSTEMI. Because of recurrent symptoms of chest pain the patient underwent emergency echocardiography, and subsequently the patient was referred for coronarography catheterization, which revealed two-vessel coronary artery disease. The patient underwent percutaneous coronary intervention with stenting of the circumflex branch of the left coronary artery. After the procedure patient felt better. During next few days she had a control carotid ultrasound and cardiologic consultation, and after 27 days from admission, she was discharged from hospital in good general state, with only slight, residual left-sided hemiparesis.

DISCUSSION

Transient ischemic attack is a major risk factor for subsequent stroke with 4% incidence within 2 days and 9% within a month from TIA. The first week seems to present the biggest risk for subsequent ischemic stroke [7, 8]. Studies show, that patients without risk factors included in California, ABCD or ABCD² scales do not experience stroke in 90 days following TIA, while those having all 5 factors had overall 90-day risk of 34%, with half of strokes occurring within 48 hours from initial symptoms [5]. In case of described patient, the ABCD² score was 4 (age >60 years old (1 point), BP \geq 140/90 mm Hg (1 point), unilateral weakness (2 points)) what means a moderate risk for stroke (2-day risk – 4.1%, 7-day risk – 5.9%, 90-day risk 9,8%). Literature data says that most patients indeed have moderate risk (up to 50%), but still nearly 20% are at high risk of stroke. Besides, 0.1% of patients with TIA are supposed to experience a myocardial infarction following the incident [9]. Although it is possible, that overall risk of stroke after TIA is a bit lower than predicted by the ABCD² scale, patients with TIA should be referred for further diagnostics in hospital, what is beneficial especially for those at high risk of stroke [7]. However, a Swiss survey shows, that awareness of the issue in physicians may be too low; although more than 90% will investigate the cause of a TIA, less than 40% would refer patient for urgent investigation [3]. The first line of treatment after TIA is aspirin and statins. Moreover, endarterectomy or carotid shunting make a great benefit to patients with TIA secondary to the stenosis of carotid artery [8,10].

Fahr's syndrome is a very rare (incidence < 1/1,000,000) entity, where calcified deposits in subcortical nuclei are found symmetrically. The disease is mostly transmitted as an autosomal dominant trait. In the described patient the brain CT was typical for the radiological criteria. However, the patient did not meet all of the other criteria. Normally the symptoms range from neurological disorders of extrapyramidal system through neuropsychiatric disorders and

cognitive impairment to Parkinsonism, chorea and tremor. Patient indeed was diagnosed with MCI (mild cognitive impairment) in psychological examination (but the onset in Fahr's disease is suspected in 3rd-4th decade of life) and had impaired walking, but she claims the problem started when coxarthrosis was diagnosed. Moreover, to diagnose a patient with a Fahr's disease, one should exclude other conditions that can influence the possible diagnosis like metabolic and endocrine disorders [6].

Managing the patient with transient ischemic attack require a watchful neurological supervision and the use of stroke prophylactic treatment (aspirin, statins), as well as the profunded diagnostics of the cause of the incident to prevent them from further cerebrovasuclar events.

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