

MIGRAINE ASSOCIATION WITH ATRIAL SEPTAL DEFECT- a case report

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Abstract

Migraine belongs to the group of primary headaches, in whose pathogenesis, despite the involvement of neurovascular, significant part, has the genetic factors, as well. The prevalence of migraine with aura in patients with atrial septal defect ranges from 11-22%. We present a case of a patient at the age of 28, with persistent headaches, 18 months backwards. Headaches were usually preceded by phosphorescence, blurred vision, buzzing in both ears, nausea, feeling of tingling of the right hemicranium. An incomplete hour later severe headaches appeared, followed by tingling and a feeling of weakness on the left arm and lower extremities. Neurological examination was with left sided paresthesias and hemiparesis. Routine biochemical examinations, hormone status, magnetic nuclear resonance of the brain and magnetic resonance angiography of intracranial vessels, cerebrospinal fluid analysis, and neurophysiological examinations was normal. At the transthoracic, and then transoesophageal echocardiography, was proved the presence of Atrial septal defect (ASD) in the central part of the interatrial septum. In the patient, an occlusion of ASD with ASD occluder was made, there was a reduction in the frequency of occurrence of migraine headaches. Controlled echocardiography was performed where the absence of the pre-established ASD occluder was observed in the aortic arch, where the removal of the device (occlusion) was done and the closing of the ASD with a direct suture. A pathogenetic mechanism that explains the association of ASD / PFO with migraine is interatrial communication, which may result in the occurrence of paradoxical embolism and humoral factors of the pulmonary circulation.

Key words: sporadic hemiplegic migraine, atrial septal defect.

Introduction

Migraine belongs to the group of primary headaches, in whose pathogenesis, despite the involvement of neurovascular, significant part, has the genetic factors, as well. According to various studies, the average prevalence of migraine ranges from 15-18% for women and 5-6% for men, with the highest number of cases registered at the age of 35-45 years. When it comes to the association of congenital heart disease with migraine, numerous studies suggest that patients with atrial septal defect (ASD) have two to four times more frequent migraine than the general population. Migraine with an aura is more common in women, while migraine without aura in men. The prevalence of migraine with aura in patients with atrial septal defect ranges from 11-22%, while that of a migraine without aura ranges from 12-28% [1,2]. Mechanisms for explaining the role of ASD in the pathogenesis of migraine are paradoxical embolism and the effect of vasoactive substances that are delayed in systemic circulation.

Case report

We present a case of a patient at the age of 28, hospitalized at the neurological department due to persistent headaches, 18 months backwards, and whose onset links them with experienced intense emotional stress. Given the preceding trigger, it was understood and treated as an anxiety-depressant state. Headaches were usually preceded by certain symptoms: phosphorescence, blurred vision, buzzing in both ears, nausea, feeling of tingling of the right hemicranium. An incomplete hour later severe headaches (diffuse, predilectionally right-handed) appeared, followed by tingling and a

feeling of weakness on the left arm and lower extremities, emphasised for the left. The headache lasted for several hours, rarely for several days, but the limb tolerance lasted for a longer time. Other hardships do not mourn, there is no positive family history of neurological or cardiovascular diseases.

From past illnesses, it is found that in children's age due to an auscultatory finding of systolic murmur, echocardiography was regularly monitored. The same period was proposed treatment abroad, which was not realized, most likely (anamnesic) because of the absence of anxiety and regular echocardiographic findings (the patient did not possess medical documentation). In childhood she was treated by hypochromic anemia when found to be a heterozygous carrier of beta-thalassemia.

Physical examination was arranged; cardiac action rhythmic, no adverse murmurs, arterial tension with values of 110 / 80mmHg.

Neurological examination with paresthesias of the right hemicranium and left limbs. Latent left-sided hemiparesis, with a positive Babinski sign.

Routine biochemical examinations (differential blood count, red cell sedimentation, glycemia, urea, creatinine, electrolytes, proteins, lipid, enzymatic status), and hormone status (thyroxine, thyreostimulative hormone-TSH, prolactin) were in reference limits. Neuroimaging studies-magnetic nuclear resonance of the brain and magnetic resonance angiography of intracranial vessels were neat. From other studies: cytochemical analysis and cerebrospinal fluid electrophoresis with normal findings. Neurophysiological examinations like: Electroencephalography (EEG), Electromyography (EMG), Evoked Potentials (visual-VEP, somatosensory-SEP) all with an orderly finding.

The anamnesic data with a typical onset, development and duration of headaches, backed up by neurological examination and findings from paracline examinations suggest that it is a migraine with aura (sporadic hemiplegic migraine), so that therapy with Topiramate has begun, with a gradual increase in the dose after 25mg at weekly intervals, to a maintenance dose of 100mg per day. This therapy was followed by a mild symptom reduction, but due to an underlying weight loss, which was an expected adverse drug effect, treatment with Topiramate was discontinued.

For the entire period, headaches were present, with periodic deterioration, but without a clear trigger on them. Due to the chronicity of headaches that functionally compromised the patient and the appearance of a mild depressive episode, antidepressant was added to the therapy. The patient regularly came to the scheduled checkups, which had always been verified by the persistent neurological deficit.

At the transthoracic, and then transoesophageal echocardiography, was proved the presence of Atrial septal defect (ASD) in the central part of the interatrial septum, with dimensions of about 12 mm and a minimum shunt, but without an evidence of volume load on the cardiac atriums and chambers.

In the patient, an occlusion of ASD with ASD occluder (umbrella 16 mm) was made; post operatively with a regulated course. The control echocardiography was with an orderly finding, with no signs of residual shunt, regular contractility and kinetics of cardiac cavities. Over the next few months, there was a reduction in the frequency of occurrence of migraine headaches, as well as spontaneous subjective sensations and tenderness of left-sided limbs. After one and a half years of surgery, frequent occurrence of headaches appeared, it was again reported on a neurological examination (in the meantime antiaggregation therapy due to pregnancy was stopped, but it ended with spontaneous abortion). The headaches were diffuse this time, after the description besides migraine, there were pains of tension type. Controlled echocardiography was performed where the absence of the pre-established ASD occluder was observed (the same found in the aortic arch). The patient was immediately transferred to the Cardiac Surgery, where the removal of the device (occlusion) was done and the closing of the ASD with a direct suture. After surgery and rehabilitation, the patient is in a stable condition with periodic appearance of migraine headaches but in the absence of neurological deficits. Control neuroimaging and EEG examinations were neat. She is currently without therapy.

Discussion

Migraine belongs to the category of primary headaches (together with a tension type headache, cluster headache and other trigeminal autonomic cephalions). In most of the cases of migraine attack (usually about 24 hours), vegetative or affective symptoms precede it. This stage is called prod. Aura is a phase of focal neurological symptoms that usually last about one hour (visual, sensory, speech disorders). After withdrawal of the symptoms that cause the aura, there is a headache, which is typically unilateral, pulsating, associated with photophobia, nausea, vomiting. Without treatment, the headache is retreated for up to 72 hours and there is a phase of resolution that usually ends with a dream. A mystery of the above description, which relates to a typical migraine with aura, are cases of migraine complications: migraine status (headache duration, with a strong intensity of more than 72 hours) and migraine infarction (with confirmed ischemic zone in the relevant area of the brain) [3].

By the mid - 20th century, vascular factors were considered as a major factor in the pathophysiology of migraine: vasoconstriction of brain vessels during the aura, with subsequent vasodilatation, resulting in pain attack. However, lately more and more emphasis has been put on neurovascular theory. According to it, the central event is the so-called cortical spreading depression: decreased electrical activity of the brain and reduced blood flow; activation of trigeminal nerve and nuclei in the brainstem; precipitation of vasodilatation.

Release of vasodilator substances in the extracellular space of the neocortex (hydrogen ions, potassium, arachidonic acid, nitric oxide) occurs. This leads to depolarization of the trigeminal endings, activation of the nucleus of the trigeminal nerve and other cores in the brainstem and dilation of the meningeal arteries. Neuroimaging studies have proven modulation of the trigeminovascular nociceptive input in dorsal mesencephalon, periacetval gray mass, dorsal nucleus raff, and locus pores locus. People with congenital heart disease have almost two to four times more frequent migraine than the general population. After transciterary closure of interatrial communication in a significant proportion of patients, an improvement in the condition of partial or complete withdrawal of migraine complaints occurs [4].

A pathogenetic mechanism that explains the association of ASD / PFO with migraine is interatrial communication, which may result in the occurrence of paradoxical embolism, but the influence of humoral factors (most of which are vasoactive substances such as nitric oxide, quinine, serotonin) that avoids degradation due to circumvention of the pulmonary circulation, which is a kind of filter [5]. This is particularly true for patients with an atrial septal defect with right-left shunt. As for the prevalence of migraine with an aura in individuals with Persistent Foramen ovale (PFO), it ranges from 13 to 50%, as opposed to 4% in the general population.

Sporadic hemiplegic migraine is an extremely rare clinical condition (according to the classification of the International Headache Society) and belongs to the subgroup of migraine with aura. Compulsory investigations to be made in these cases are neuroimaging and lumbar puncture, in order to exclude pseudomegran, which is characterized by occasional transient neurological outbreaks and lymphocytic pleocytosis in the cerebrospinal fluid.

In the early childhood (most likely) the patient was diagnosed with ASD (without medical documentation), which was confirmed by transoesophageal echocardiography; the same occluded with ASD occluder. After the intervention, a reduction in the frequency of occurrence of migraine attacks was observed.

The mechanism explaining the migraine attacks are paradoxical (micro) emboli because of the existence of communication between the two atriums, but the role of vasodilator substances that do not normally occur in the systemic circulation should not be ignored. Their influence is especially on individuals with ASD and right-left shunt, because they circumvent the lung filter. People suffering from migraine had 2-4 times more frequent changes in white matter (detected on magnetic nuclear resonance) than others [6]. This is especially true for people with migraine with an aura [7].

With respect to the association of white matter lesions (WMLs) with right-left shunt there are studies that negate the association [8], but also studies in which the right-to-left shunt relationship with changes in small blood brain vessels that are probably due to paradoxical embolus [9,10].

Despite proven association of migraine with congenital heart disease, however, according to the recommendations of the American Association for Headache, it is not necessary to screen for the existence of atrial and / or ventricular septal defect in all patients with migraine.

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