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Moya Moya's disease in a 20-year-old female patient – case report

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Introduction

Moya-Moya disease is a rare disease of unknown etiology that causes pathological occlusion changes in the large intracranial arteries - the internal carotid artery and the proximal parts of the anterior and middle cerebral arteries. The disease occurs most often among young Asian women, and its most intensive development falls on the first decade of life. In most cases, Moya-Moya occurs in the form of transient ischemic attacks, strokes or cerebral hemorrhages.

Case report

A 20-year-old woman was admitted to the Neurosurgery Clinic for the surgical treatment of Moya-Moya disease. Patient with paroxysmal paresis of left and right limbs several times since 2016. In CT, MRI and angio-MRI, as well as DSA, critical narrowing of the internal carotid arteries, obstruction of the anterior and central lights of the cerebral arteries, with small vessels around the central arteries and their branches marked - bilaterally, but mainly on the right. The changes create an image resembling "clouds of smoke" that is characteristic of Moya-Moya's disease. The patient was qualified for surgical treatment - revascularization of the cerebral cortex with the use of periosteal strips by multi-hole trepanation in the right fronto-temporo-parietal region. Periosteal stripes were introduced into the subarachnoid space to reproduce vessels of the cerebral cortex.

Conclusion

Because of the rarity of Moya Moya disease, it is very important to be alert vigilantly to avoid complications such as tetraparesis or mental retardation. Japanese surgery allows revascularization of the cerebral cortex without the need for an arterial bypass. The presented surgical method ensures good treatment effects and is preferred in young people due to the safety of the technique used.

Key words: Moya Moya disease, neurosurgery, hemorrhages

Introduction

Moya Moya is a rare vascular disease with unknown etiology. In order to pathological process that affects the vessels occlusive changes appear in them. Alterations occur in the arteries that make up the circle of Willis. We can observe narrowing or occlusion especially in distal part of internal carotid artery, proximal part of anterior cerebral artery and medial cerebral artery. This location is characteristic for the disease and the diagnosis is determined on its basis [1]. Moya Moya occurs mainly in the Asian population. Appears more often in women than in men [2]. The prevalence of the disorder is estimated at 6.03/100,000 population in Japan [3], 9.1/100,000 population in Korea [4] and 1.63/100,000 population in Taiwan [5]. In Poland, the morbidity for the disease was set at 0.089 / 100,000 cases based on 34 registered cases of the disease in 2013 [2]. There are two peaks of disease. The first reveals in the first decade of life, while the second occurs between the third and fourth decades of life [31].

The pathological process mainly involves the inner membrane of the arteries and leads to its thickening. The presence of lipid deposits was also found there. This leads to a narrowing of the light of the cerebral arteries what can cause ischemic changes in the brain. Chronic ischemia also stimulates angiogenesis processes resulting in the formation of a pathological vascular network [6]. Cerebral ischaemia leads to serious complications of the disease such as transient ischemic attacks, strokes, cerebral haemorrhage or even quadriplegia. ischemic

attacks are more common in pediatric patients, however intracranial haemorrhages are usually observed among adults [2].

There is no causative treatment of Moya Moya. Neurosurgical treatment is the only available method of therapy. There are several techniques for performing surgery. Direct and indirect methods are included. Direct methods such as superficial temporal artery–middle cerebral artery anastomosis (STA-MCA) are usually used in adults. Indirect methods such as EDAS (encephalo-duro-arterio-synangiosis) or Multiple burr hole surgery are more beneficial among paediatric patients [2].

Case report

We present 21 years old woman who was admitted to Department of Neurosurgery. Patient noticed the first symptoms of the disease in 2016. At the time of their appearance she was 18 years old. She reported paresis of the limbs on the right and left side of the body. The symptoms were paroxysmal and their intensity was increasing. Paresis afflicted both upper and lower limbs. According to anamnesis the patient did not suffer from any other diseases. In her family history there wasn't any case of Moya Moya. The patient underwent a series of imaging examinations including computer tomography (CT), magnetic resonance imaging (MRI), MR angiography (MRA) and Digital Subtraction Angiography (DSA). A significant degree of narrowing of the internal carotid arteries, large narrowing of the right and left central and anterior cerebral arteries and small vessels around the middle cerebral arteries have been found. On the basis of the obtained imaging results, the diagnosis was made. The changes were located on both sides, but on the right side they were more advanced. In addition to imaging tests, a number of diagnostic tests were also commissioned. Morphology, biochemical tests of the liver and kidneys, electrolyte levels, coagulation system tests, urine tests, thyroid hormone levels, glucose concentration were performed. The results of laboratory tests of the patient were mostly correct. The only deviations observed were a minor decrease in the haematocrit value (36,2%), slight increase in total cholesterol (219 mg/dl) and minimal prolonged prothrombin time (13,00 s). The value of the INR index was within the normal range. Neurosurgical treatment was performed to the patient. The woman underwent two operations - revascularization of the cortex using multiple burr hole technique. The first operation took place on 12/02/2018 and concerned changes located on the right side of the temporo-parietal-frontal region. The second intervention was performed on 11/12/2018 and affected changes located on the left side of the same area. The operations consisted of making trepanning 6 holes in this area to expose dura mater. After skin incision, strips of periosteum

connected to the skin flap were made. Subsequently neurosurgeons cut dura mater and then arachnoid mater. Afterwards doctors punctured the cortex and inserted dissected strip into the holes and placed on the cerebral cortex in order to start revascularization process. The patient was discharged home in good condition both after the first and second surgery. Further treatment consisted of taking 75 mg acetylsalicylic acid once a day. The patient was also advised to visit the neurosurgical outpatient clinic and repeat angio-MRI for 12 months.

Discussion

Moya Moya as a rare health conditions is very difficult to diagnose. Most often the first symptoms of the disease are its complications. Taking into account the most common occurrence of the disease in children or young people, it can significantly decrease quality of patient's life or even lead to their death. The prognosis of Moya Moya is difficult to estimate. The disease can be slow and steady, however course of the disease can be aggressive and turbulent and quickly lead to neurological deterioration [7,8]. The incidence of symptoms is associated with age [9]. In children, the dominant symptoms are ischemic symptoms, and the main one is transient ischemic attack [10]. Other usual symptoms in this group include intellectual decline, seizures, and involuntary movements. Involuntary movements such as chorea, dystonia, and dyskinesia probably are caused by hypoperfusion in the basal ganglia or cerebral cortical areas [14]. Vascular changes in the cerebral circulation such as microaneurysms or false aneurysms in collateral vessels can cause intracerebral haemorrhage or subarachnoid haemorrhage [11]. These symptoms are more common in adult population [2]. What is more, location of haemorrhage caused by Moya Moya disease is different than due to primary intracerebral hemorrhage. Intracerebral hemorrhage related to the disease were mostly intraventricular, lobar and in the putamen [12]. Headaches can also be a manifestation of the disease, but their etiopathology remains unclear [13]. According to research conducted at the Medical University in Lublin, the most common symptoms were hemiparesis. Tetraparesis was much less frequent. In addition to the previously mentioned symptoms, dysphasia, epilepsy and numbness of the limbs have also been found [2].

The presented patient did not suffer from any other diseases, however Moya Moya can be associated with other disorders. Such a condition is then called moya moya syndrome [11]. Genetic, hereditary disorders such as Neurofibromatosis [15,16] , Down syndrome [17] or Noonan syndrome [18] may accompany illness. Furthermore some of haematological disorders for example essential thrombocythemia [19] hereditary spherocytosis [20] , protein C deficiency [21] , and protein S deficiency [22] are also related with Moya Moya. The illness

sometimes also occurs with connective-tissue diseases: Systemic lupus erythematosus [23] , antiphospholipid antibody [24] , and livedo reticularis [25]. The other health conditions such as Infectious or chronic inflammatory conditions, metabolic diseases and vascular injuries may also appear with Moya Moya [11]. Artheriosclerosis is reported in Japan and Taiwan as a most common associated disease [26,27]. Thyroid disorders such as Graves disease and Hashimoto disease are considered to be significantly related to Moya Moya [6]. In addition, association between Moya Moya and Hashimoto disease was reported as a risk factor of bad result of revascularization surgery [28].

The main aim of surgical revascularization is to is to avoid brain ischemia by improving cerebral flow [29]. Our patient underwent revascularization of the cortex using multiple burr hole technique. This method is classified as indirect surgery technique. This kind of surgery is considered as a primary choice for neurosurgical treatment, especially in the young patients. The one of most important advantages of this technique is no need to create arterial by-passes. This method enables revascularization of ischemic regions that would not be possible through superficial temporal artery by-pass. A condition and dimension of patient's arteries does not influence on the outcome of treatment. What is more this technique does not require using coagulation. Multiple burr hole technique is more simple, less invasive , has a lower risk of complications and does cause the hyperperfusion syndrome in comparison to the direct techniques [1]. Other indirect methods such as encephalomyo synangiosis (EMS) or encephalo-duro-arterio synangiosis (EDAS) are also used in the treatment. They are classified according to the different tissues covering the brain [29]. Their effectiveness is probably conditioned by the tendency to spontaneous angiogenesis in patients with Moya Moya [30]. These methods have numerous advantages despite the fact that the more time is needed to improve cerebral blood flow [29]. Direct techniques include bypass. The operation consists in creating anastomosis between superficial temporal artery (STA) and cortical arteries. As mentioned above, this kind of surgery requires an appropriate vessel diameter and may develop post-operative hyperperfusion syndrome [29].

Conclusions

Due to the rarity of Moya Moya disease, it is very important to remain vigilant to avoid complications such as tetraparesis and mental retardation. Moya Moya can occur with other illnesses, which may indicate a diagnosis. The operation performed in the presented patient enables revascularization of the cerebral cortex without the need to establish arterial bypasses.

This surgical method ensures good treatment effects and is preferred in young people due to the safety of the technique used.

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