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Treatment of Moya Moya disease using indirect surgery technique

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Abstract

Introduction: Moya Moya disease is a rare health condition with unknown etiology. Because of the pathological process in the patient vessels we can observe narrowing or occlusion of distal part of internal carotid artery and proximal part of anterior and medial cerebral artery, which is characteristic for this disease. The disease occurs most often among asian women and it usually affects patients in their first decade of life or 3-4 decade.

Case presentation: Four year old female suffered from headaches, dizziness as well as progressive paroxysmal hemiparesis and speech disorder. The symptoms were recurrent and lasted for two years. MRI and MRA were performed and revealed pathological changes in arteries of scull base and pathological collateral arteries, characteristic for moyamoya disease.

Surgical treatment: Multiple burr hole surgery was performed. Holes were made in the left fronto-temporo-parietal area, peritoneal straps were inserted into the subarachnoidal area in order to start revascularization process.

Conclusion: Presented technique is preferred among children suffering from moya moya disease because of the safety of the technique and good treatment effects. There is no need to perform by-pass surgery which is also considered as big advantage of presented technique.

Introduction

Moya-Moya disease is a chronic condition of unknown etiology characterized by progressive narrowing of the cerebral vessels and as a result leading to the occlusion and triggering the growth of the vascular network of the collateral circulation. In adults, it is characterized by the presence of intracerebral hemorrhage, in contrast to pediatric patients who are diagnosed mainly with recurrent ischemic strokes. There is no pharmacological treatment of the disease so surgery is still the only way to stop the progress of this condition. For older children and adults direct techniques of revascularization are used, while in younger children indirect revascularization is preferred.

The aim of this surgical technique is to restore flow in the middle cerebral artery by creating anastomoses from the external carotid artery, mainly using the temporal artery, temporal muscle or dura mater (EDAS, EMS or a combination EDAMS). A more recent technique, invented by Japanese neurosurgeons and popularized in Europe by French neurosurgeons is a procedure of multiple trepanations.

Epidemiology

First reported in Japan, mainly found among the Asian population, however, it is observed throughout the world [1]. In Japan, it is the most common children cerebrovascular disease, which affects women almost twice as often as men [2]. The disease occurs in two age groups: pediatric patients in the first decade of life and adults in the 3-4 decade of life. The predominant symptom among pediatric population was ischemic stroke while in the adult population the most common syndrome was hemorrhagic stroke [3,4]

Case report

In February 2014 four year old girl suffered from headaches and dizziness, occurred after the collapse which resulted with paresis of the right side of the body. The CT scan revealed bilateral ischemic stroke in the range of anterior cerebral artery. Paresis partially withdrew. MRA and angio - CT made in February 2014 revealed a stenosis of significant degree of the distal segments of both carotid arteries with obstruction of the left and the absence of the left anterior cerebral artery and incomplete filling of the middle cerebral arteries with small vessels around the middle cerebral arteries and their branches (both sides , mainly on the left side). The image was characteristic for the Moya-Moya disease. In December 2015 the patient at the time of infection with chickenpox suffered from another episode of ischemic attack of that left hemisphere of the brain, manifested with paresis of the right half of the body and speech disorders. Patient admitted to the clinic in good general condition, neurological examination found slight degree of paralysis of the right hand. She was qualified for surgery - revascularization of the cortex using multiple burr hole technique in the left fronto-temporo-parietal area.

Trepanning holes were made in this area, four at the base of a skin flap and the other two holes in the upper row. In each of the holes dura mater was exposed, which was cut, then the arachnoid

matter was cut and after that the cortex was punctured. Strips of periosteum connected to the skin flap were prepared. Dissected strips were inserted into the holes and placed on the cerebral cortex. Discharged home in good general condition, neurological examination found slight degree of paralysis of the right-hand, same as the date of admission, walking, wound healing properly.

Discussion

This technique was invented by accident by the japanese neurosurgeon Endo in 1984. Bilateral frontal burr hole for ventricular drainage was performed and three months later the neovascularization process via burr holes was observed [5]. It is an effective way to supply ischemic regions, that are not covered by the superficial temporal artery by-pass.

This procedure has a lot of advantages and the most important is that it does not require to perform arterial by-passes. The effect of the treatment is not determined by the condition and size of patients arteries. During the surgery usually no cortical vessels are coagulated, which is beneficial for the patient. The procedure is simple, less invasive as compared to a direct revascularization and results with low risk of perioperative complication. On the contrary to the direct techniques this surgery does not expose the patient to the hyperperfusion syndrome. In some centers it is performed under local anesthesia and the results are similar as the direct revascularization technique. In studies presented by De Oliveira all patients were treated with indirect revascularization technique and improved clinically with no additional ischemic attacks [6].

Concussion

Although, Moya Moya disease is a very rare condition, it is increasingly recognized and connected with cerebral ischemia especially among pediatric population. Diagnosis is based on the characteristical clinical and radiological presentation - stenoocclusive changes in cerebral vessels

and collateral vessel development. The use of indirect surgical procedures is considered effective, safe and durable way of cerebral revascularization and should be a primary choice when it comes to treatment of this condition, especially in the pediatric patients.

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