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CASE REPORT

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Arteriovenous Malformation of the Vein of Galen: A Case Report

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Abstract

The article presents literature data and the authors' observations of the course of the vein of Galen aneurysmal malformation in a one-day-old newborn infant. This clinical case is of practical interest since the vein of Galen aneurysmal malformation is a rare congenital anomaly of cerebral vessels. The results of arteriovenous malformation treatment in recent years have improved considerably, but many diagnostic and curative aspects in children require the development of new approaches to addressing this problem. (**International Journal of Biomedicine. 2021;11(1):58-60.**)

Key Words: arteriovenous malformation • the great cerebral vein • children • diagnosis • treatment

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Introduction

Brain diseases caused by damage to the vascular system, are one of the major problems of modern neurology and neurosurgery. This is due to the prevalence of these pathological conditions in the population: the severity of clinical manifestations, high mortality rate, and a large percentage of disabled people. Often cerebrovascular diseases in newborns and young children are combined with congenital infections, birth hemorrhages, cerebral and non-cerebral developmental defects, and birth injuries. At the same time, in children, the defects of brain vessels, their course, treatment, and prognosis differ from those in adults, especially in very young children.

Anatomy. The great cerebral vein is a large-diameter short vein, draining the internal brain veins and two basal veins into the straight sinus. The formation of the aneurysm of the great cerebral vein occurs during the period from the sixth to the 11th weeks of intrauterine development.[1-6] At the same time, embryos first develop arteries, then veins.[7,8]

According to the angiostructural peculiarities, arteriovenous malformations (AVMs) of the vein of Galen [9] are divided into 2 types:

Choroidal type – the true AVMs of the great cerebral vein, when the feeding arteries are directly connected (have fistula) to the wall of the aneurysmally dilated great cerebral vein;

Mural type – aneurysmal dilations of the great cerebral vein caused by true cerebral AVMs or dural arteriovenous fistulas draining into the normal but dilated great cerebral vein.

Clinical manifestations of the vein of Galen aneurysmal malformation (VGAM) often depend on the child's age, his/her pathophysiological features, and the type of VGAM.[4] In newborns, VGAM can occur in the form of asymptomatic cardiomegaly and mild cyanosis to severe cardiac failure with the gradual development of hydrocephalus due to the occlusion of the cerebral aqueduct characterized by the presence of the convulsive syndrome. In children under 1 year of life, after the newborn period, the most frequent manifestation of the disease is the occlusive hydrocephalus and delay in psychomotor development.[5] In older age groups, two forms

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of VGAM can be observed – apoplectic and pseudotumor-like with tumor symptoms of subtentorial localization. In the case of cranial auscultation, a pulsating noise, synchronous with the heart beat,[2] is sometimes heard. The pathological process in patients with VGAM is progradient, seldom remitting. The course of the disease should be predicted with caution.

Diagnosis and treatment. The diagnosis of VGAM in the fetus is carried out according to the ultrasound and MRI data, the latter being the method of choice. In other age groups, MRI, MRA, and CT are used. The MRA allows better visualization of the venous (venous drainage) and arterial (feeding pedicle) anatomy, as well as better assessment of the condition of the brain, which is suffering from recurrent ischemia and hemorrhage. As a rule, CT detects only pathological formations, and when the contrast agent is introduced, the image density of pathological formations significantly increases as a result of the entry of the contrast agent into the dilated vascular bed.[2,4]

Currently, the only effective method of treating VGAM in newborns and children of other age groups is surgery: endovascular occlusion of AVM feeding vessels as well as direct surgical obliteration. If hydrocephaly occurs, a shunt can be installed in the liquor system if necessary. In children, radiosurgery can also be successfully used in the treatment of VGAM. With conservative management, periodic observation (every 6-12 months) using noninvasive methods of research – MRA and X-ray CT angiography – is desirable.[3]

There is little information in the literature about the features of the VGAM clinical picture in newborns and young and older children when combined with other pathological conditions that are characteristic or common in childhood. The relevance of the problem of diagnosis and treatment of VGAM is due to the high mortality rate, disability, and frequent complications.[4]

Case Presentation

Patient Information

A one-day-old newborn boy was admitted to the regional children's clinical hospital from the perinatal center for examination with the diagnosis: "The congenital anomaly of the development of cerebral vessels – the aneurism of the vein of Galen (according to ultrasonography). Cerebral ischemia of the second degree. Vesiculopustulosis."

Mother: A 31-year-old woman, gave birth to her second child with her third pregnancy, had high blood pressure starting from early pregnancy.

Father: A 39-year-old healthy man.

According to the prenatal ultrasound study, the defect of the development of cerebral vessels (aneurysm of the vein of Galen) was discovered for the first time at 39 weeks of gestation. The second delivery, at 39 weeks, by emergency cesarean section.

Clinical Findings

Body weight at birth – 4540g, length – 60 cm, head circumference – 35 cm, chest circumference – 36 cm. The Apgar score – 8-9 points. In the first hour of life, the infant had symptoms of respiratory failure grade 2, according to the Silverman scoring system. At 12 hours of life, a tachypnea was registered up to 60 breaths per minute, lengthening of exhalation, moderate retraction of the intercostal space (Downes scale: Grade 3). At 24 hours of age, showing stable hemodynamics, the newborn infant was transferred from the Children's Division at the Perinatal Center to the Division of Resuscitation and Intensive Care at the Regional Children's Clinical Hospital. There was no need for respiratory therapy. From the third day of life, the examination and treatment of the infant was carried out at the Division of Pathology of Newborn and Premature Babies.

Objective Data

The child's condition, by the nature of the disease, is severe. Consciousness and physique are correct. The skin cover is icteric and clean. No visible edemas; a moderate pastiness of tissues was noted. The ribcage is cylindrical. In the lungs, puerile respiration. The respiratory rate – 40 breaths per minute. Heart sounds are sonorous, rhythmic. The heart rate is 140 bpm. The abdomen is soft; the liver protrudes below the costal margin by 1cm, the spleen is not palpable. Urination is not disturbed. The stool is loose, regular. The child is mixed-fed; it ingests food, with no regurgitation.

Neurological status: The newborn is conscious. The head is rounded. Bregmatic fontanel is 2.0×2.0 cm, not tense. The cranial sutures are closed. Motor activity is sufficient. Reflex responses of innate automatism were evoked but quickly depleted. The cranial innervation is normal. The infant fixes his gaze and follows objects. Muscle tone is moderately reduced. Deep reflexes are lively and symmetrical. Sensitivity is not compromised. Vegetative-trophic functions are not changed.

Diagnostic Assessment

The blood and urine tests, biochemical analysis of blood (total protein, urea, creatinine, glucose, ALAT, ASAT, total bilirubin and its fractions, electrolytes, CRP, serum iron), are within the age normal limits. No antibodies to CMV, mycoplasma, toxoplasma, and chlamydia by the ELISA method are found. Indicators of acid-base state and coagulogram are without deviations.

Chest X-ray: no focal infiltrative changes are detected. The shadow of the mediastinum is expanded due to the lobes of the thymus gland.

Cervical spine X-ray: kyphotic deformity at the level of the C2–C4 segment. Indirect signs of natal injury are noted – dislocation of C1–C2 vertebral bodies.

ECG: sinus rhythm, heart rate of 170 bpm. The position of the electrical axis of the heart is normal. Incomplete block of the right branch of the His bundle.

Echocardiography: open oval window. The heart cavities are not enlarged. The contractility of the left ventricular myocardium is satisfactory. Data for congenital heart disease and pulmonary hypertension are not obtained.

Ultrasound of the organs of the hepatopancreatoduodenal zone and kidneys show no pathology. Neurosonography at 5 days of age: signs of aneurysm of the vein of Galen, hypoxic changes in the substance of the brain, and periventricular edema. MRI of the brain: VGAM (choroidal type).

Examination by an ophthalmologist and an ENT doctor: no pathology.

Neurologist's examination: The congenital anomaly of the development of cerebral vessels – VGAM of choroidal type. Cerebral ischemia of the second degree. Natal injury to the cervical spine.

Treatment

Antimicrobial and dehydration therapy and a course of energy correctors (levocarnitine) were carried out.

Outcome and Follow-up

During the stay in the hospital, the child's condition was satisfactory; no focal neurological symptoms were detected. The dynamics was observed by a neurosurgeon. At 16 days of age, he was discharged for outpatient care with a neurosurgeon's recommendation for surgical treatment at the age of 6-8 months.

Discussion

This clinical case is of practical interest since the vein of Galen aneurysmal malformation is a rare congenital anomaly of cerebral vessels. The frequency of occurrence of this aneurysm in childhood is 30%.[8] In different age periods, if a child has an arteriovenous malformation, it may increase due to involving new vascular formations in the malformation; therefore, the need for expedient early detection and surgical treatment is obvious. After the surgical treatment of arteriovenous malformations in children, focal neurological symptoms, seizures, and developmental delays are often observed. These neurological disorders in children, as a rule, are more easily compensated through rehabilitation measures than in adults.[10,11] The results of arteriovenous malformation treatment in recent years (5-7 years) have improved considerably, but many diagnostic and curative aspects in children require the development of new approaches to addressing this problem.

Competing Interests

The authors declare that they have no competing interests.

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