ISCHEMIC STROKE IN A CHILD WITH COMPLEX CYANOTIC CONGENITAL HEART DISEASE

Aniela Luminita Rugina[1], Violeta Streanga[1], Nicolai Nistor[1], Gabriela Buhusi[2], Ramona Filipescu[2], Irina-Mihaela Ciomaga[1]

[1]"Gr. T. Popa" University of Medicine and Pharmacy, Iasi

[2]"Sf. Maria" Children Emergency Hospital, Iasi

ABSTRACT

Stroke is rare in children compared to adults. Most common causes of stroke in children are heart diseases that cause embolism and cerebral infarction and arteriovenous malformations for the brain hemorrhages, but a number of cases remain idiopathic, without being able to say precise etiology. The authors present the case of a child diagnosed in infancy with complex cyanotic heart malformation, but not operated, admitted with clinical and radiological signs of pneumonia, which evolved favorably with antibiotic and symptomatic therapy. During resolution of the pneumonia, the child becomes suddenly drowsy, presenting right hemiplegia, with absence of reflexes on the same side. Clinical examination correlated with cranio-cerebral CT have established the diagnosis of bilateral Sylvian ischemic stroke. The evolution was favorable under conservative treatment (antiplatelet drugs, physiotherapy), but right hemiplegia persisted.

Keywords: ischemic stroke, congenital heart disease, child, hemiplegia

INTRODUCTION

Ischemic stroke is a rare pediatric emergency, the incidence being estimated to 2-3/100.000 (1). Boys are twice as frequently affected than girls (12). While in adults the most frequent cause (80%) of stroke is atherosclerosis, in children there are many causes, the most common being embolic heart disease. The most frequent clinical manifestations are hemiplegia and convulsions. A lot of imaging techniques help to confirm the diagnosis: transfontanellar ultrasound, in newborn and infant, transcranial Doppler, CT, MRI. The chronology of these tests depends on the children's age and the availability in emergency, but MRI exam is the best (2). In children there is no standard treatment, compared to adults. Special attention should be paid to sequelae which occur in 70% of cases (motor, epileptic, cognitive) (3). However, the great plasticity of the brain at young age explains the unexpected recovery in contrast to the size of the brain infarct (2).

CLINICAL CASE PRESENTATION

SF, male, aged 1 year and 9 months, diagnosed in infancy with complex cyanotic congenital heart disease, is hospitalized for fever, rhinorrhea, nasal obstruction, productive cough with onset for 1 week. At 6 weeks of age the child was diagnosed with abdominal situs inversus, left atrial isomerism, single atrium, atrio-ventricular dual connection through a single atrioventricular valve, single ventricle - left ventricular type with double outlet, complete transposition of the great arteries, valvular and subvalvular pulmonary stenosis. Afterwards the infant was admitted four times to the Institute for Cardiovascular Diseases and Transplantation Târgu Mureş to determine whether surgery is convenient, but the family refused invasive preoperative investigations (transesophageal echocardiography, cardiac catheterization). On admission the child presented malaise, cyanosis, productive cough, pulmonary crepitation bilaterally, SaO_2 = 70% in the atmosphere, improved to 82% with oxy-

Corresponding author:
Violeta Streanga, "Gr. T. Popa" University of Medicine and Pharmacy, 16 Universitatii Street, Iasi
E-mail: streangavioleta@yahoo.com

gen, precordial bulge, apical impulse at the fourth left intercostal space in the midclavicular line, regular heart rate = 90/minute, systolic murmur grade 3 in the left parasternal area, BP = 95/55 mmHg, palpable peripheral pulse, cold extremities, liver located on the left, normal growth chart.

Laboratory findings were noted: polycythemia, inflammatory syndrome; chest X-ray shows opacity in the left upper lung, heart flaunted on the diaphragm (Fig. 1). Electrocardiogram showed superior QRS axis (AQRS = -150˚) and right ventricular hypertrophy; echocardiography reveals stationary evolution of the cardiopathy.

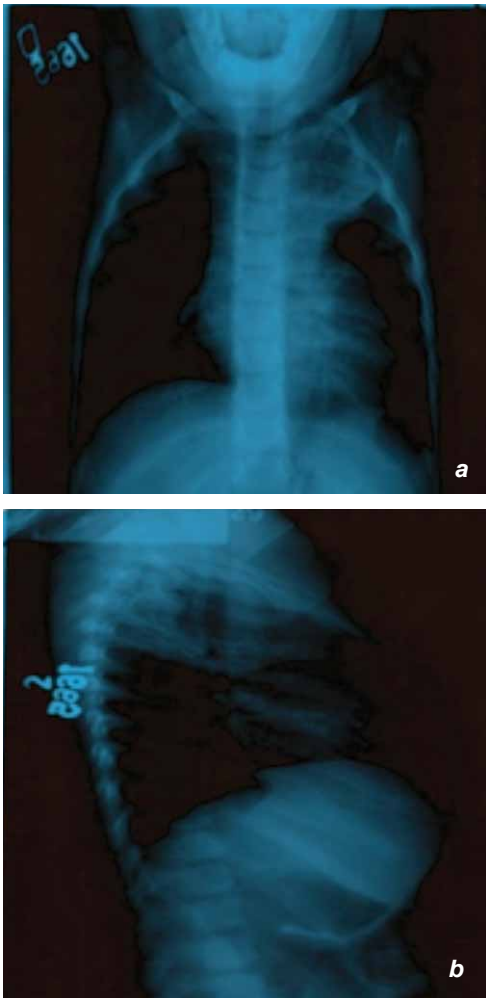

FIGURE 1. *Chest X-ray, a – postero-anterior, b – lateral*

The outcome of the respiratory disease was favorable after one week of treatment with antibiotics, anti-inflammatory and expectorants; chest x-ray showed the resolutive aspect of the pulmonary opacity, maintaining interstitial infiltration (Fig. 2). The child suddenly became drowsy, right hemiplegia occured; we ascertained the absence of reflexes on the same side and therefore we suspected cerebral infarction due to thrombo-embolism (Fig. 3). Cranial CT-scan revealed cerebral ischemic stroke in the territory supplied by both middle Sylvian artery, more importantly on the left side (Fig. 4).

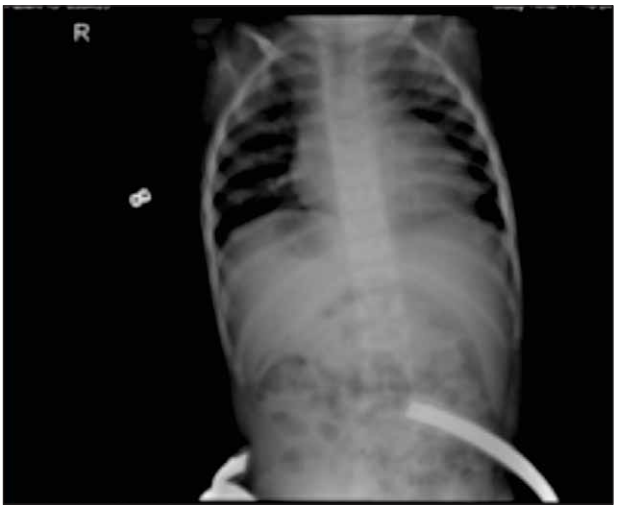

FIGURE 2. *Chest X-ray check-up*

FIGURE 3. *S.F., 1 year and 9 months. Right hemiplegia*

FIGURE 4. *Cranio-cerebral CT: left Sylvian ischemic stroke*

The neurosurgeon opted for medical treatment, as neurosurgical treatment was not mandatory at that time. The child received antiplatelet therapy and physiotherapy. He returns for annual reexamination and the persistence of right hemiplegia is aknowledged.

DISCUSSION

The World Health Organization (WHO) defines stroke as a clinical syndrome characterized by a rapidly occurring focal or global disturbance of cerebral function, lasting more than 24 hours or leading to death, with no obvious nonvascular cause (4). This definition does not take into account, however, neuroimaging, considered now essential to determine the neurovascular origin of symptoms. Therefore, it is considered that the modern definition of stroke is a clinical syndrome characterized by:

- a neurological deficit related to the perfusion territory of a cerebral artery;
- neuroimaging evidence of an ischemic injury (5).

This case falls within the age group most commonly affected by ischemic injury in childhood. Data shows that pediatric stroke is most frequently encountered between 1-5 years of age and rarely at extreme ages of childhood (under 1 year and over 15 years) (6). Up to 25% of ischemic strokes in children are due to heart disease, most cases being previously diagnosed with cardiac malformation. Stroke is more common in children with unoperated cardiac malformations. Thrombus may originate in the atria (eg. atrial septal defect + pulmonary hypertension), ventricle (eg. ventricular septal defect + pulmonary hypertension) or arterial (pulmonary arteriovenous fistula) (7).

Both in adult and in child, cerebral ischemia results from decreased cerebral blood flow (CBF), mostly related to the occlusion of a cerebral artery by embolic material. The consequences of this tissue hypoperfusion depend on its duration and intensity (8). The cerebral ischemia area can be divided schematically into three parts, from the periphery to the center:

- a moderate ischemia area, where the reduction of cerebral perfusion has no clinical expression;
- an area of "shadows" where CBF is still sufficient to ensure an energy supply for cell survival, but not enough to enable their operation; this area is responsible for the neurological deficit;
- an area of necrosis, caused by an impaired cellular defense system to hypoxia and cell death; this area is responsible for constituted neurological deficit, which persists even if normal CBF is early restored.

Ischemia is responsible for anaerobic glycolysis, intracellular acidosis and intracellular calcium inflation due to disruption of calcium channel receptors (2).

Clinical symptoms vary depending on the age of the child. Thus, the onset is brutal under the age of 10 years, mostly by occurrence of hemiplegia (as in the case presented) and/or seizures, often generalized; after this age, clinical symptoms are close to those seen in adults (9). Diagnosis is confirmed by neuroimaging, which reveals vessel obstruction, as well as the extent and location of ischemic lesions. In the presented case, ischemic lesions were large, computed tomography showing hypodense areas in the superficial and deep territory supplied by the left middle cerebral artery (Sylvian) (Fig. 4) but also in the superficial territory supplied by the right Sylvian artery. We proceeded to: upright positioning the patient in order to improve venous return and to reduce the risk of inhalation pneumonia; monitoring vital signs and the level of consciousness; platelet administration.

For patients with acute stroke, anticoagulants (unfractionated heparin or heparinoids) are not indicated; only antiplatelet agents can be used. Secondary prevention with anticoagulation therapy may be initiated in the event of transient ischemic attack or minor cerebral infarction in patients presenting embolic heart with high risk of relapsing. Therapy should be initiated immediately or within days, if there are no contraindications. Intravenous thrombolysis in child is practiced only sporadically in some well-equipped centers, this technique not being validated in patients under 18 years of age. For treatment of intracranial hypertension due to cerebral infarction or cerebral hemorrhage, mannitol, glycerol, furosemide, hypertonic solutions and corticosteroids are not recommended (3).

The patient we presented had survived, but with sequelae (hemiplegia). The literature shows that infant mortality of stroke is about 16%; it can increase to 40% if previous severe disease exist (10). Moderate or severe hemiplegia is found in 42% of cases. Other waste disorders include: dysphasia, in the case of right brain impairment, and ataxia, in the event of damage to the rear. Regarding cognitive development, it seems that patients who have suffered an ischemic stroke have a normal IQ value (85-95), but may have impaired attention and memory, visual-spatial function disorders and reduced quality of life in adolescence (11,12).

CONCLUSIONS

Ischemic stroke, although rare in children, should be considered in patients with known con-

genital heart malformations, especially cyanogens, if hemiplegia or seizures occur suddenly without another cause. Neuroimaging examination reveals the location and the extent of ischemic lesions, which determine the therapeutic decision upon the acute phase and the recovery of possible sequelae.

REFERENCES

1. **Janjua N., Nasar A., Lynch J.K., Qureschi A.I.** Thrombolysis for ischemic stroke in children: data from the nationwide inpatient sample. *Stroke*. 2007; 38:1850-1854.
2. **Béjot Y., Hervieu M., Osseby G.V. et al.** Les accidents vasculaires cérébraux chez l'enfant: pourquoi y penser? 52e Congrès National d'Anésthesie et de Réanimation Médecin. Urgences vitales, *SFAR* 2010.
3. **Bollaert P.E., Vinatier I., Orlikowski D., Meyer P.** Prise en charge de l'accident vasculaire cérébral chez l'adulte et l'enfant par le réanimateur (nouveau-né exclu), (hémorragie méningée exclue). *Réanimation* 2010; 19:471-478.
4. **Aho K., Harmsen P., Hatano S.** Cerebrovascular disease in the community: results of a WHO collaborative study, *Bulletin of the World Health Organization* 1980; 58(1): 113–130.
5. Group TPSW, Ed., Stroke in Childhood. Clinical Guidelines for Diagnosis, Management and Rehabilitation. *The Lavenham Press*, Suffolk, UK, 2004.
6. **Lee Y.Y., Lin K.L., Wang H.S. et al.** Risk factors and outcomes of childhood ischemic stroke in Taiwan. *Brain Dev* 2008; 30 (1):14-19.
7. **Kumar K.** Neurological complications of congenital heart disease. *Indian J Pediatr* 2000; 67:287–291.
8. **Leys D., Pruvo J.P.** Strategie pratique en urgence face a un AVC. EMC - Neurologie 2002; 17-046 A50:1-10.
9. **Lopez-Vicente M. , Ortega-Gutierrez S., Amlie-Lefond C., Torbey M.T.** Diagnosis and management of pediatric arterial ischemic stroke. *J Stroke Cerebrovasc Dis* 2010; 19(3):175–183.
10. **Adams R.J., Brambilla D.** Discontinuing prophylactic transfusions used to prevent stroke in sickle cell disease. *N Engl J Med* 2005; 353: 2769–78.
11. **Pavlovic J., Kaufmann F., Boltshauser E.** Neuropsychological problems after paediatric stroke: two year follow-up of Swiss children. *Neuropediatrics* 2006, 37(1): 13–19.
12. **Steilin M., Wwhrli E.** L'accident vasculaire ischémique en pédiatrie. Quand y penser- quoi faire! *Paediatrica* 2009; 20 (2) : 22-26.