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Transient hemiplegia in a child with Sturge-Weber/Klippel-Trenaunay syndrome

José Maria Pereira de Godoy MD, PhD*,[1], Ana Carolina Pereira de Godoy, MD[2]

[1] *Cardiology and Cardiovascular Surgery Department of the Medicine School in São José do Rio Preto (FAMERP), CNPq (National Council for Research and Development)-Brazil*

[2] *Physician specialized Intensive Therapy Pediatrics and member Research Group of the Clínica Godoy, São Jose do Rio Preto, Brazil*

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ABSTRACT

This study was to describe the neurological evolution of a patient an association of Sturge-Weber and Klippel-Trenaunay syndromes. We report on a girl who was born with port wine-colored lesions over the entire body. The child later suffered two neurological episodes; the second left the patient hemiplegic, without consciousness and unable to speak. She started to recover after the seventh day, however full recovery took more than one month during which time periods of improvement and deterioration of cerebral perfusion were detected. Currently the patient is 23 years old, attended a college and is well without obvious neurological signs.

Key words: Sturge-Weber syndrome–Klippel-Trenaunay syndrome–hemiplegia

1 INTRODUCTION

Cutaneous vascular malformations are rare disorders representing errors in vascular development. It is important to properly diagnose vascular malformations because of their distinct differences in morbidity, prognosis and treatment. Vascular malformations may be associated with underlying disease or systemic anomalies [1].

Sturge-Weber syndrome is characterized by capillary malformations on the face, associated with leptomeningeal and choroidal venous malformations. Klippel-Trenaunay syndrome consists of the triad: capillary malformation of one leg, ipsilateral hypertrophy and varicose veins [2]. Descriptions of dual or overlapping syndromes have been published with many showing that there is no clear distinction between Klippel-Trenaunay syndrome and Sturge-Weber syndrome; there are a clinical and biological overlaps. The complexity of disease phenotypes shows that a classification based on an eponymous category does not enable resolution of the nosological problems [3].

Hemiplegic migraine in the setting of Sturge-Weber syndrome has been previously described [4].One study showed transient changes on brain magnetic resonance imaging in a patient with Sturge-Weber syndrome presenting with hemiparesis [5].

* Email: godoyjmp@gmail.com.

Epilepsy, hemiparesis, mental retardation and ocular problems have been reported as the most frequent and severe features of patients with Sturge-Weber. Cerebral lesions followed a progressive course during early childhood, but not later [6, 7]. The purpose of this study is to report the neurological evolution of a patient with Sturge-Weber syndrome associated with Klippel-Trenaunay syndrome.

2 CASE REPORT

A three-year-old white female patient was consulted in the pediatrics service due to changes in the skin color from birth. A physical examination identified port wine-colored macular lesions on the face, especially in the central region with a slight predominance on the right, on the neck, chest, abdomen and limbs as well as dental abnormalities and thinning of hair on the left.

Electrocardiogram, brain CT scan, scanometry, skull and spine x-ray and ultrasound of the entire abdomen were requested. The left leg was 1.8 cm shorter by scanometry. At seven years old the patient suffered from fainting and so further exams were requested; the brain CT scan and EEG were normal.

At eight years old, a cerebral perfusion study was performed that showed moderate hypoperfusion in the right temporal lobe and in a small area of the right frontal lobe. A whole body scan with labeled red blood cells was normal.

The patient was evaluated by an ophthalmology team who diagnosed glaucoma and started treating with medications. At age 10, she was admitted in the pediatric emergency department complaining of incoordination, mental confusion and difficulty speaking. On physical examination the patient was confused and only responsive to painful stimuli. A brain CT scan without contrast showed no pathological changes. Complete recovery was seen in 24 hours and the patient was discharged. After 10 days of outpatient treatment another cerebral perfusion examination was performed the result of which was better than previous studies. Two months after hospitalization, the patient was again consulted in the emergency room due to reduced consciousness, loss of muscle strength of the right half of the body and inability to talk. A physical examination showed that the patient was sleepy and unresponsive. She was admitted to hospital and evolved with episodes of epileptic seizures and maintained a reduced level of consciousness and aphasia. She began presenting with neurological improvement seven days after admission and complete improvement was seen in about one month. Another EEG showed slow waves in the anterior region of the left cerebral hemisphere where there was blurring of the normal grapho-elements. However, epileptiform disorders were not observed. One month after admission, an updated assessment of the cerebral perfusion was requested which showed hypoperfusion throughout the left cerebral hemisphere and right cerebellar hemisphere. The patient underwent cerebral angiography of 4 vessels (right and left internal carotid arteries and right and left vertebral arteries), which showed agenesis of the A1 and hypoplasia of the left vertebral artery. At 11 years of age, an EEG showed grade 1 diffuse slowing of brain electrical activity in the anterior left region and epileptiform disorders in the left fronto-temporal region. A MRI showed reduced volume in the left cerebral hemisphere with predominance in the choroid plexus which may represent angioma of the choroid plexus. Asymmetry of the cortical branches of middle cerebral arteries was noted with decreased vascularity on the right.

The patient has progressed well in the last six years without obvious neurological signs. Currently the patient is 23 years old, attended a college and is well without obvious neurological signs.

3 DISCUSSION

This study reports on the evolution of the neurological status of a child with an association of Sturge-Weber syndrome and Klippel-Trenaunay syndrome. As the child grew, she suffered impairment of cerebral perfusion neurological disorders. Neurologic involvement in Sturge Weber syndrome has been described in the literature but this has not been reported in association with Klippel-Trenaunay syndrome.

Studies show that there is no clear distinction between Klippel-Trenaunay and Sturge-Weber syndromes; there are clinical and biological overlaps. The complexity of the disease phenotype shows that a classification based on one category of the same name does not allow the resolution of nosological problems [2]. Thus, there is no study evaluating the interaction of these syndromes on the evolution of patients.

In this study the neurological confusion with loss of muscle strength began to improve after the seventh day, but complete improvement only occurred in about one month. The cerebral perfusion examinations suggest that these states are associated to cerebral hypoperfusion. The improvement in the six years of evolution after the last crisis suggests an adjustment and stabilization of cerebral circulation however progressive neurological damage has been previously described [5].

Psychological involvement was not observed in this child who remains active. Skin lesions due to hemangiomas were mitigated with the use of laser and further behavioral problems were not observed during adolescence.

In the first years no neurological changes were detected in examinations including brain imaging and electroencephalography. The first neurological involvement was detected at eight years of age when assessing cerebral perfusion. Therefore, the clinical features of Sturge-Weber syndrome were fully defined after detecting neurological disorders; previous to this only the clinical diagnosis of Klippel-Trenaunay syndrome had been defined. The association of these diseases and the interaction of complications have not been characterized in the literature and so monitoring will be essential to better understand the clinical evolution.

4 CONCLUSION

Currently the patient is 23 years old and is well without obvious neurological signs

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AUTHOR BIOGRAPHY

José Maria Pereira de Godoy MD, PhD Cardiology and Cardiovascular Surgery Department of the Medicine School in São José do Rio Preto (FAMERP), CNPq (National Council for Research and Development)-Brazil

Ana Carolina Pereira de Godoy, MD Physician specialized Intensive Therapy Pediatrics and member Research Group of the Clínica Godoy, São Jose do Rio Preto, Brazil