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Case Report

Hemiplegia and Paresis Unilateral as a Manifestation of Atypical Moyamoya Disease and Antiphospholipid Syndrome: A Case Report

Daniel Martínez-Rodríguez[1], Francisco E. Rivas-Larrauri[2], Armando de Uña-Flores[3], Julio Granados[4], Javier E. Anaya-Ayala[5], Susana Hernández-Doño[4], Angeles Citlali Zavala-Valencia[1], Luz-María Torres-Espíndola[1*]

[1]Pharmacology of Laboratory, National Institute of Pediatrics, Mexico City, Mexico

[2]Immunology Service, National Institute of Pediatrics, Mexico City, Mexico,

[3]Radiology and Imaging Service, National Institute of Pediatrics, Mexico City, Mexico

[4]Division of Immunogenetics. Department of Transplants. National Institute of Medical Sciences and Nutrition Salvador Zubirán, Mexico City, Mexico

[5]Department of Surgery, Section of Vascular Surgery and Endovascular Therapy, Instituto Nacional de Ciencias Médicas y Nutricion Salvador Zubirán, Mexico City, Mexico

***Corresponding author:** Luz-María Torres-Espíndola, Pharmacology of Laboratory, National Institute of Pediatrics, Mexico City, Mexico

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Abstract: Moyamoya Disease (MD) is an occlusive cerebral vasculopathy that is rarely reported in the pediatric population and is associated with Cerebrovascular Accidents (CVA). We herein present a pediatric case with multiple comorbidities associated with MD; imaging studies confirmed cerebral ischemia and arteriovenous malformations. The diagnoses of MD, trisomy 21, and Antiphospholipid Syndrome (APS) were confirmed, and the patient was treated with thrombo-prophylaxis, antithrombotic and immunomodulation treatment; on follow-up, the patient remains asymptomatic. The prompt diagnosis and treatment reduced the risk of new ischemic events in the case presented. The knowledge of the risk factors of MD allows a more in-depth examination of the affected patient.

Keywords: Antiphospholipid antibody syndrome; Cerebrovascular disease; Moyamoya disease; Pediatrics; trisomy 21.

Introduction

Moyamoya disease (MD) is a progressive cerebral occlusive vasculopathy characterized by bilateral lesions in the terminal portions of the internal carotid artery (ICA) with posterior hypertrophy and an abnormal vasculature in the base of the brain due to the proliferation of anterolateral central arteries [1].

There are two types of presentations: idiopathic and Quasi Moyamoya disease, and it includes vascular changes associated with an underlying pathology such as Down syndrome, APS, cardiovascular structural anomalies, PHACE syndrome, and radiotherapy [1-3]. Clinical manifestations include ischemic neurologic symptoms or intracranial hemorrhage [4-5]. The

diagnosis is based on imaging studies, cerebral angiography is the gold standard, [5], and the treatment is pharmacological and surgical. On the other hand, two therapeutic options are available; pharmacological and surgical used as therapeutic in these clinical presentations.

Case Presentation

A five-year-old Mexican female with a history of Trisomy 21, and ventricular septal defect. The patient presented right-side hemiplegia and distal cyanosis. On physical exam, facial asymmetry, hypotonia, and right-hemiparesis. To the suspicion of CVA performed CT scan, which demonstrated hypodense areas in the left frontal lobe suggesting CVA. A nuclear magnetic resonance with contrast demonstrates the decreased caliber of perforating vessels and collateral circulation in the lenticulostriate of the skull base suggesting MD. A T2 FLAIR demonstrated decreased brain volume and zones of hypoperfusion in the frontal and left parietal lobe (Figure1A-1C). A 3DTOF showed the decreased caliber of the ICA and neovasculature in the supraclinoid region (Figure 1 D).

The laboratory tests revealed a normal, but test Anticardiolipin Ig A, Ig G, and Ig M and AntiB2-glycoprotein were positive in two determinations, confirmed APS. This case represented a therapeutic dilemma, due to the risk of thrombosis for APS and bleeding due to vascular anomalies. Was decided reduction of the ischemic risk, with ASA (5mg / Kg / d), acenocoumarin (1mg), and the use of prednisone 0.5 mg/kg / d, dipyridamole (75mg a day), hydroxychloroquine (3.5mg /kg / d), and methotrexate (7.5mg a week) to partially inhibit the immune system, reducing the autoimmune inflammatory state. Presently the patient is responding to the medical therapy without complications at seven years with a favorable prognosis.

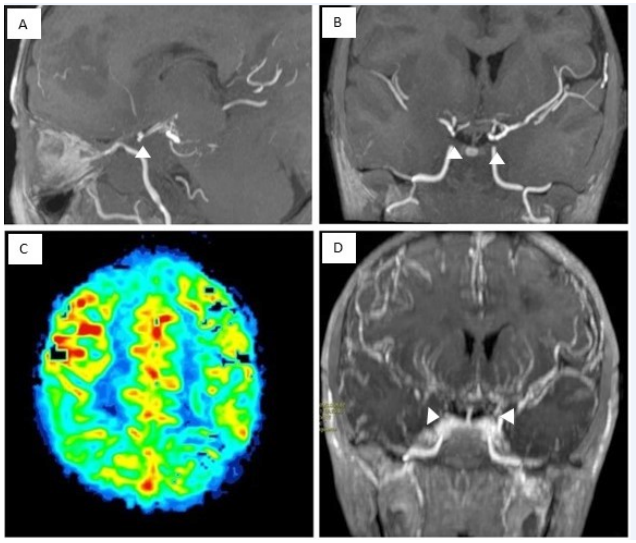

Figure 1 (1A-1D): (A) Magnetic angio-resonance of the skull, (B) Multiplanar parasagittal and coronal reconstruction, sequence T1: decrease in the caliber of perforating thalamus vessels and collateral circulation in the lenticulostriate region of the skull base; (C) MRI perfusion technique: hypo perfused areas in the left frontal and parietal region compared to the right hemisphere; (D) 2D angiographic sequence TOF: decrease in the caliber of internal carotid arteries and neovasculature formation in the supraclinoid region.

Discussion

MD is an uncommon cause of CVA in the pediatric population, it is estimated that 2-3 per 100000 cases of stroke in this patient population, and 6% are secondary to vasculopathy [4,6]. This pathology is unusual in Latin America, it is reported

a peak between the age of 5 and 9 years. The etiology is uncertain and it is associated with underlying pathologies as observed in this patient [7]. Sciascia S, et al. 2015 reported that the presence of any antiphospholipid antibodies seems to confer a fivefold increased risk for CVA when compared with control patients without antiphospholipid antibodies [8]. Korematsu S et al. 2017 [9] reported increased levels of phosphatidyl-choline, phosphatidylethanolamine, and anti-cardiolipin antibodies in pediatric patients with cerebral stroke, therefore the authors suggest that the antiphospholipid antibody syndrome is an important primary disorder leading to cerebral stroke and can cause both artery and vein stroke. The mechanism remains unknown but it has been proposed that the binding of antibodies to the phospholipids of vascular endothelial cells causes activation of the complement, induction of inflammatory cytokine, and coagulation disorder [10].

According to the treatment of patients with MD, the preventive effect of surgical revascularization has been clinically demonstrated; in the case reported, the patient received pharmacological therapy and, at present, the case is evolving favorably without complications or sequelae of the first event. Timely diagnosis and early treatment have reduced new ischemic events. The study of patients at risk could help clinicians to avoid the progression of vasculopathy and provide the best prognosis for this rare disease.

Conclusion

In conclusion, further studies are required to evaluate the role of the immune system in patients with trisomy 21 who develop antiphospholipid syndrome and Moyamoya disease since the use of immunosuppressive drugs can improve the quality of life and avoid surgical procedures.

Conflicts of Interest

The authors declare no conflict of interest.

Informed Consent

The patient gives her written informed consent to participate this study.

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