SOCIEDADE BRASILEIRA DE REUMATOLOGIA

REVISTA BRASILEIRA DE REUMATOLOGIA

www.reumatologia.com.br

Case report

Intracerebral hemorrhage with a favorable outcome in a patient with childhood primary angiitis of the central nervous system

Marco Felipe Silva[a], *Ana Raquel Feitosa*[a], *José A. Paz*[b], *Nádia Emi Aikawa*[a,c], *Clovis A. Silva*[a,c,*]

[a] *Pediatric Rheumatology Unit, Faculdade de Medicina, Universidade de São Paulo, São Paulo, SP, Brazil*
[b] *Pediatric Neurology Unit, Faculdade de Medicina, Universidade de São Paulo, São Paulo, SP, Brazil*
[c] *Division of Rheumatology, Faculdade de Medicina, Universidade de São Paulo, São Paulo, SP, Brazil*

ARTICLE INFO

Article history:
Received 20 December 2013
Accepted 21 May 2014
Available online 30 November 2015

Keywords:
Childhood primary angiitis
Central nervous system
Vasculitis
Intracerebral hemorrhage
Stroke

ABSTRACT

Childhood primary angiitis of the central nervous system (cPACNS) is a rare inflammatory brain disease of unknown etiology. Of note, brain hemorrhage has been rarely reported in cPACNS patients, generally associated with a delayed clinical diagnosis, or with a diagnosis only at necropsy. We present the case of a boy with cPACNS that previously suffered an ischemic stroke. At the age of 7 years and 10 months, he presented a sudden and severe headache, vomiting and reduction in consciousness level (Glasgow coma scale 7), requiring prompt tracheal intubation. Brain computed tomography demonstrated intraparenchymal hematoma in the right parieto-occipital lobe and a small focus of bleeding in the right frontal lobe, vasogenic edema, herniation of the uncus and a 10 mm deviation to the left from the midline. C-reactive protein (9.2 mg/dL) and von Willebrand factor (vWF) antigen (202%) were elevated. Decompressive craniotomy was performed and methylprednisolone and cyclophosphamide were administered. One week later, the patient had left hemiparesis without other sequelae. Importantly, motor deficits have been improving progressively. Our case reinforces the inclusion of this vasculitis as a differential diagnosis in children and adolescents with CNS hemorrhage.

Hemorragia intracerebral com evolução favorável em paciente com angiíte primária do sistema nervoso central juvenil

Palavras-chave:
Angiíte primária na infância
Sistema nervoso central

RESUMO

Angiíte primária do sistema nervoso central juvenil (APSNCJ) é uma doença inflamatória cerebral rara e de etiologia desconhecida. Hemorragia cerebral tem sido raramente reportada em pacientes com APSNCJ, geralmente associada com atraso diagnóstico, ou com um

* Corresponding author.
E-mail: clovisaasilva@gmail.com (C.A. Silva).
http://dx.doi.org/10.1016/j.rbre.2014.05.007

Vasculite
Hemorragia intracerebral
Acidente vascular encefálico

diagnóstico somente por necrópsia. Relata-se um caso de um paciente do gênero masculino com APSNCJ e que previamente sofreu um acidente vascular cerebral isquêmico. Aos 7 anos e 10 meses de idade, o menino apresentou subitamente cefaleia intensa, vômitos e redução do nível de consciência (escala de coma de Glasgow 7), requerendo imediata intubação traqueal. Uma tomografia computadorizada cerebral demonstrou hematoma intraparenquimatoso no lobo parieto-occipital direito e um pequeno foco de sangramento no lobo frontal direito, edema vasogênico, herniação do úncus e um desvio de 10 mm da linha média para a esquerda. A proteína C-reativa (9.2 mg/dL) e o fator antígeno de von Willebrand (202%) estavam elevados. Foi realizada uma craniotomia descompressiva, seguida pela administração de metilprednisolona e ciclofosfamida. Transcorrida uma semana, o paciente apresentava hemiparesia esquerda, sem outras sequelas. É digno de nota que o *déficit* motor tem melhorado progressivamente. Nosso caso reforça a inclusão dessa vasculite como diagnóstico diferencial em crianças e adolescentes com hemorragia do sistema nervoso central.

Introduction

Vasculitides are characterized by inflammation and necrosis of endothelium.[1] They can affect blood vessels of any organ and system, including the central nervous system (CNS). CNS vasculitis most often is secondary to an underlying condition, such as infectious, neoplastic, vascular, metabolic or inflammatory disorders, but it also can be idiopathic.[2,3]

Of note, childhood primary angiitis of the CNS (cPACNS) is a rare inflammatory brain disease of unknown etiology that occurs in previously healthy children. It has protean clinical manifestations, such as seizures, cognitive dysfunction, behavior changes, headaches, neurologic deficits and strokes.[3]

Ischemic stroke seems to occur more often than hemorrhagic events, which are seldom described in the literature. As far as we are concerned, only seven cases were reported regarding hemorrhagic stroke and cPACNS in the literature, most of which had delayed clinical or necropsy diagnosis.[1,4–8]

We describe a patient with cPACNS and intracerebral hemorrhage submitted to prompt drainage and immunosuppressive therapy with good prognosis.

Case report

At the age of 7 years and 2 months, a boy presented intense headache during about 5 minutes with spontaneous improvement. In the next twenty-four hours, he had labial commissure deviation to the right side and left hemiparesis, being hospitalized in another hospital. Laboratory exams were: hemoglobin 12.7 g/L, hematocrit 36%, white blood cell count 10,300/mm^3 (neutrophils 39%, lymphocytes 52%, monocytes 5% and eosinophils 4%), platelets 300,000/mm^3, urea 30 mg/dL (normal range 10-50), creatinine 0.53 mg/dL (normal range 0.32-0.60), C-reactive protein (CRP) 1.1 mg/dL (normal < 5), erythrocyte sedimentation rate (ESR) 7 mm/1st hour (normal range 0-20) and activated partial thromboplastin time 27.2 s, International Normalized Ratio (INR) 1.1. Homocysteine levels were 5.5 micromol/L (normal range 5-15), total cholesterol 176 mg/dL (normal < 200), low-density lipoprotein cholesterol 112 mg/dL (normal < 100), high-density lipoprotein cholesterol 49 mg/dL (normal > 40) and triglycerides 79 mg/dL (normal < 150). Brain magnetic resonance imaging (MRI) showed ischemic areas in the right middle cerebral artery territory, characterized by subcortical foci of restricted diffusion in the nucleocapsular region, whereas carotid and vertebral magnetic resonance angiography, conventional angiography and carotid Doppler ultrasound were unremarkable, as well as cerebrospinal fluid analysis. Immunological tests were positive for antinuclear antibodies (ANA) 1:80 (fine dense speckled pattern) and negative for other serum antibodies: anti-double stranded DNA (anti-dsDNA), anti-Sm, anti-RNP, anti-Ro, anti-La, anticardiolipin IgM, anticardiolipin IgG, lupus anticoagulant, anti-β2-glycoprotein-1 and antineutrophil cytoplasmic antibodies. Urinalysis was normal, and blood culture was negative. Protein C (115%), protein S (126%) and factor VIII (87%) activities were within the normal range. Factor V Leiden mutation and prothrombin gene polymorphism were both absent. Aspirin (5.0 mg/kg/day) was introduced, and after 4 months of rehabilitation physiotherapy, complete recovery of the motor deficits was achieved and aspirin was withdrawn. At the age of 7 years and 8 months, he returned asymptomatic in an outpatient visit, bringing a new brain MRI that revealed encephalomalacia in the right nucleocapsular region and leptomeningeal enhancement in the left precentral sulcus, left central sulcus, right occipital sulcus, sphenoid portion of the right Sylvian fissure and inferior aspect of the cerebellar hemispheres, compatible with CNS angiitis (Fig. 1). Prophylactic aspirin was reintroduced, and he was referred to our University Hospital. At that moment, the patient did not have complaints. He had normal weight and height development. Peripheral artery pulses were palpable, there was no claudication of extremities, and blood pressure was 99 × 65 mmHg, without differences in the limbs. No right-left shunt was observed during echocardiography with microbubbles, CRP was 0.9 mg/dL, and ESR was 11 mm/1st hour. At the age of 7 years and 10 months, he had a sudden and severe headache, vomiting and reduction in consciousness level (Glasgow coma scale 7), requiring prompt tracheal intubation. Urgent brain computed tomography demonstrated intraparenchymal hematoma in