PRIMARY VASCULITIS OF THE CENTRAL NERVOUS SYSTEM: A CASE REPORT IN A CHILD

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BACKGROUND

Primary vasculitis of the central nervous system is a rare disease that can affect vessels of any size, spinal cord and leptomeninges. In this report, a case of involvement of medium-sized vessels with clinical ischemic stroke is described.

CASE REPORT

A 5-year-old female patient with no previous comorbidities is brought to the emergency room due to sudden right hemiparesis with brachioradial predominance associated with dysarthria and labial commissure deviation to the left. An investigation with cranial tomography was carried out, which showed involvement of the left capsuloganglionic artery compatible with ischemic stroke. Later, cranial magnetic resonance angiography (MRA) was performed and showed stenosis in medium-caliber intracranial arteries, more markedly in the distal communicating segment of the internal carotid artery and proximal branches, suggestive of vasculitis. Infectious screening was performed with lumbar puncture, carotid and vertebral MRA and transthoracic echocardiography without alterations. Still investigated antiphospholipid antibody syndrome, favor V Leiden mutation and prothrombin and all negative. Thus, a diagnosis of primary central nervous system vasculitis was performed. The patient underwent cerebral angiography with findings similar to those described above and underwent angioplasty with a stent. Treatment with six doses of cyclophosphamide 1000 mg/m² with excellent evolution and after maintenance with azathioprine 75 mg/day with good clinical response. At the moment, the patient has not reactivated the disease, maintaining a slight loss of strength in the right upper limb.

CONCLUSION

Primary vasculitis of the central nervous system is a rare disease that can lead to severe, irreversible and recurrent neurological deficits if not diagnosed. Treatment consists of immunosuppression with cyclophosphamide or high-dose corticosteroids with subsequent maintenance to prevent disease reactivation.

KEYWORDS

Vasculitis, Central nervous system, Child.