Possible reversible cerebral vasoconstriction syndrome (RCVS) in a patient with undifferentiated connective tissue disease

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Objectives: Reversible cerebral vasoconstriction syndrome (RCVS) and undifferentiated connective tissue disease are two rare conditions. The possible involvement of cerebral arteries in connective tissues diseases has not been extensively investigated.

Materials and Methods: A 48-year-old woman experienced three episodes of recurrent, sudden-onset, severe, frontal, pulsating headache associated with nausea, vomiting, visual blurring and postural instability. The acute pain improved within two hours after analgesic therapy but mild headache persisted between exacerbations. She came to the emergency department on day four after the first episode of "thunderclap headache". Brain computed tomography was unremarkable and neurological examination was normal. Basic laboratory tests, including complete blood count, electrolytes, coagulation screen, renal and liver function tests, immunoglobulin electrophoresis, erythrocyte sedimentation rate, C-reactive protein and 24-hour urinary metanephrines were within normal range. CSF analysis was also negative. The patient had, since three years, a history of recurrent and diffuse polyarthralgia, malar rash, photosentivity and xerophthalmia with a high titre speckled anti-nuclear antibody (ANA) pattern and anti-extractable nuclear antigens. Recent history did not reveal exposure to vasoactive substances, toxins or cocaine. She had no recent pregnancies.

Results: Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) revealed multi-focal narrowing of cerebral arteries, without parenchymal abnormalities (Figure 1). A diagnostic possibility of RCVS was considered and the patient was started on oral calcium-channel antagonist.

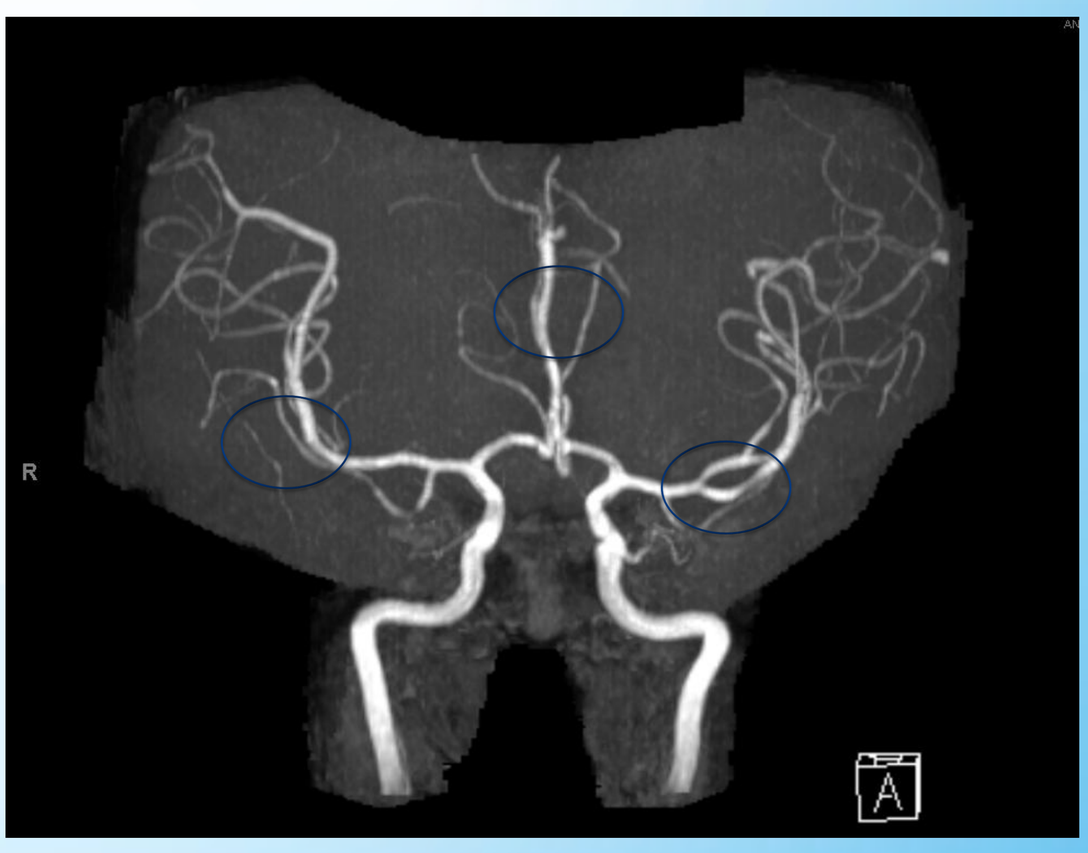
ANA	Positive speckled, 1:640
ENA	Anti-Sm-RNP +
Rheumatoid factor	-
ANCA	-
Ab anti- cardiolipin	-
Ab anti-phospholipids	-
Ab anti ds-DNA	-

. Digital subtraction angiography, performed eight days after the initial symptoms, showed an improvement of the segmental vasoconstriction of cerebral arteries while serial transcranial doppler ultrasonography documented elevated mean flow velocities in the left middle cerebral artery for two weeks after headache onset. Blood investigations during hospitalization confirmed positive ANA (titre 1:640) and positive anti-Smith/ribonucleoprotein antibodies. Rheumatoid factor, anti-neutrophil cytoplasmic, anti cardiolipin, anti-phospholipids, anti ds-DNA antibodies were negative. A retrospective diagnosis of undifferentiated connective tissue disease was made and hydroxychloroquine was introduced.

Discussion: In the following weeks the patient did not experience further episodes of headache and nimodipine was progressively discontinued. Brain MRA, performed three months later, demonstrated complete regression of vasoconstriction. Clinical, laboratory and radiological findings excluded a diagnosis of primary angiitis of central nervous system or secondary vasculitis. The hypothesis of RCVS had been taken into account.

Conclusion: RCVS could be considered, to some extent, a Raynaud's phenomenon of cerebral arteries in patients with systemic autoimmune disorders associated to signs and symptoms not sufficiently evolved to fulfil any of the accepted classification criteria for the defined connective tissue diseases.

Figure 1.



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