Hematomyelia in Systemic Lupus Erythematosus and Secondary Antiphospholipid Syndrome: Case Report

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Intraspinal hemorrhage is very rare and intramedullary hemorrhage, also called “hematomyelia,” is the rarest form of intraspinal hemorrhage. There are few reports in medical literature. We report the case of a woman of 43 years with diagnosis of systemic lupus erythematosus and secondary antiphospholipid syndrome under anticoagulant therapy that presented hematomyelia. We describe the clinical course, the findings in the image studies and surgery, and the available information in the literature.

Key words: Hematomyelia. Systemic lupus erythematosus. Antiphospholipid syndrome. Anticoagulant therapy.

Clinical Case

A 43 year-old woman, whose mother had SLE and 2 cousins had rheumatoid arthritis, presented a case of deep venous thrombosis of the left leg at 37 years of age and 1 month later, deep venous thrombosis of the right leg. Simultaneously, the patient presented symmetric polyarthritis, oral ulcers, hair loss, positive antinuclear antibodies, anti-dsDNA, anti-Sm, and IgG anticardiolipin antibodies, as well as hypocomplementemia, lymphopenia, and renal affection with an OMS stage IIb glomerulonephritis, with which a diagnosis of SLE and SAPS was reached. She received treatment with steroids at a low-to-medium dose, chloroquine 150 mg/day, azathioprine up to 100 mg/day, pentoxifilline, and anticoagulation with acenocumarine at variable doses, achieving an adequate control. Three years later she presented renal reactivation and was treated with 6 intravenous boluses of cyclophosphamide, shewing improvement, managed afterward with prednisone 10 mg/day, azathioprine 100 mg/day, and acenocumarine. In April 2006 she consulted her rheumatologist because...
of headache and paresthesias on the face, sudden pain on the neck and back and, after some hours, a loss of muscle force on the legs, oppressive chest pain anesthesia from the T4 level downward, acute urinary retention, sphincter incontinence, abdominal distension, and, finally, paraplegia. Upon hospitalization she presented lymphopenia, 160/µL; platelets, 248 000/µL; INR, 1.16. A transverse myelitis was suspected for which she was treated with 6 g of intravenous methylprednisolone and 1 g of intravenous cyclophosphamide, and sent to our hospital. When examined we found generalized, burning pain, without sphincter control, with conserved superior mental functions, with hair loss, unaffected cranial nerves, a sensitive level at T4, with right upper extremity muscle strength of 2/5 proximal and 4/5 distal; on the upper left extremity, 4/5 proximal as well as distal; the rest was paraplegic. She had no Achillean reflex; she presented a left Babinski sign but it was indifferent on the left side.

The blood count showed: hemoglobin, 9.4 g/dL; hematocrit, 28%; leucocytes, 5050/µL; lymphocytes, 210/µL; platelets, 65 000/µL. Blood chemistry was normal. A 24 hour creatinine clearance of 41 mL/min was documented; triglycerides, 325 mg/dL; liver function tests were normal; C3, 67.8; C4, 6.31; acute phase reactants with a erythrocyte sedimentation rate of 56 mm/h; and a C-reactive protein of 47.9 mg/L was found. Coagulation tests showed TP, 20.8; TPT, 23; INR, 2.1. Urine testing showed a pH of 6, leucocytes 0-5 per field, erythrocytes 10-15 per field, abundant bacteria, nitrates (+). An spinal tap found thick, port-red wine colored liquid without an increase in pressure, erythrocytes, 531 200; crenocytes, of 10%; leucocytes, 310; proteins, 2756 mg/dL; LDH, 2616; glucose, 8 mg/dL; coagulability (–), hemoglobin (++++); MEX-SLEDAI, 7.

A cranial computerized tomography (CT) was normal. On the magnetic resonance (MRI) there was a medullar widening, a homogeneous intramedullar T1 and T2 homogeneous hyperintensity, corroborated on the axial cuts with heterogeneous images and medullar displacement (Figures 1 and 2) interpreted as a spinal hematoma in an organization phase with signs of medullar degeneration. Anticoagulation was suspended and a bilateral hemilaminectomy at the T2-T3 level was performed, showing a thoracic hematoma at the T2-T3 level, and a subdural hematoma at the C4. The anatomopathology report showed an old hematoma, which was partially organized and had neuronal degeneration without evidence of neoplasia. The evolution of the patient was poor, developing spinal degeneration without recovery of the muscle strength or the sensitive level and without SLE activity.

**Discussion**

Intraspinal hemorrhage is infrequent and intramedulla hemorrhage or hematomyelia is the rarest form of intraspinal hemorrhage.1-4 There are very few reports of hematomyelia in the medical literature, most in relation to anticoagulant treatment. The male:female ratio has been established at 1.5:1, and is described more frequently between the sixth and seventh decades of life, this attributed to a larger use of anticoagulant in this age group.4 Among the predisposing factors, anticoagulation, trauma, arteriovenous malformations, tumors, and hemorrhagic diathesis can be included.4-6 The most frequent localization in children is C5-T1, different from adults, more frequently found at a low cervical and thoracolumbar level.7,8 Hematomyelia clinically presents as an abrupt medullar syndrome, with high-intensity pain, localized or in a radicular distribution, acute or related with a sensormotor deficit, which is associated to sphincter atony. It is rapid in its onset and evolution, making a prompt diagnosis and opportune treatment of the utmost importance.4-9-11
Diagnostic suspicion of the disease is fundamentally based on clinical findings and must be considered in all patients presenting a case with medullar topographical localization installed in an acute or hyperacute manner, and in which a traumatic cause has been ruled out. Sudden pain accompanied by a medullar neurological deficit of rapid installation and a history of therapeutic risk factors firmly contributes to the diagnosis. Among the complementary tests that must be performed, MRI is superior to CT. Typical findings consist of hyperintense images both on T1 as on T2, medullar widening, and even an image showing medullar compression. Treatment of the intraspinal hemorrhage, apart from treating the cause, must consider complications left by the myelopathy and the attempt to surgically excise the lesion as soon as possible. Prognosis depends on the rapidness of surgical decompression and, overall, on the neurological state before the surgery, though the eventual possibility of complications must be considered, such as hemorrhage and irreversible lesions in patients in which a rapid evacuation is not carried out.

There are no reports in the literature of patients with SLE or SAPS and hematomyelia. A total of 8 cases of acute, spontaneous hematomyelia, 2 of them in newborns, 1 after a spinal tap, can be found, and the other one was secondary to obstetric trauma. Another 6 cases were related to anticoagulant therapy, as apparently our patient was and, finally, Matsumura et al described 2 cases of chronic, progressive, and idiopathic, surgically treated and with a good outcome, indicating that acute and chronic presentations have a different prognosis because the evolution of the patients with spontaneous hematomyelia associated to anticoagulation was generally poor as happened to our patient. Anticoagulant treatment was immediately suspended in our patient, though excessive anticoagulation was never evidenced. She underwent a hemilaminectomy which showed a thoracic hematoma with spinal degeneration and a bad subsequent course, not regaining muscle strength or sensibility.

Intramedullar spinal hemorrhage is extremely rare and anticoagulant treatment is an important risk factor for this complication. Clinical suspicion, an opportune diagnosis using MRI, and a timely intervention are essential in attaining a better neurological prognosis.

References