

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

Fedra Irazoque-Palazuelos, Patricia V. Sosa-Espinosa, and Lilia Andrade-Ortega

Servicio de Reumatología, CMN 20 de Noviembre, ISSSTE, México DF, México

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.

Hematomielia en lupus eritematoso generalizado y síndrome antifosfolipídico secundario: informe de un caso

La hemorragia intraespinal es muy poco frecuente y la hemorragia intramedular, conocida también como hematomyelia, es la forma más rara de hemorragia intraespinal. Existen muy pocos reportes de hematomyelia en la literatura médica. Comunicamos el caso de una mujer de 43 años, con diagnóstico de lupus eritematoso generalizado y síndrome antifosfolipídico secundario en tratamiento anticoagulante que presentó hematomyelia. Describimos los datos clínicos, los hallazgos de imagen y cirugía, así como la información existente en la literatura.

Palabras clave: Hematomyelia. Lupus eritematoso generalizado. Síndrome antifosfolipídico. Tratamiento anticoagulante.

Correspondence: Dra. P.V. Sosa Espinosa.
Andrés Molina Enríquez 1210-101. Colonia Ampliación Sinatel.
09470 México DF. México.
E-mail: psosae@hotmail.com

Manuscript received April 12, 2007; accepted for publication May 31, 2007.

Introduction

Central nervous system hemorrhage is a rare and potentially lethal complication. It occurs with a greater frequency in the intracranial zone and it is mainly intracerebral or subdural. Intraspinal hemorrhage is much less common, can be epidural, subdural, subarachnoid, or intramedullary, and can have devastating consequences. Intramedullary hemorrhage, also known as hematomyelia, is the rarest form of intraspinal hemorrhage.

From the first report of "spontaneous hematomyelia," published by Richardson in 1938, some cases have been reported but its incidence has not been established. The term "spontaneous" makes reference to hemorrhage that is not related to trauma, vascular malformations (VMF), or tumors, and in most cases was associated with a hemorrhagic diathesis or anticoagulant treatment.

We present the case of a patient with systemic lupus erythematosus (SLE) and secondary antiphospholipid syndrome (SAPS) who developed hematomyelia, possibly as a complication of anticoagulant treatment.

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, pentoxifylline, 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, showing improvement, managed afterward with prednisone 10 mg/day, azathioprine 100 mg/day, and acenocumarine. In April 2006 she consulted her rheumatologist because



Figure 1. Magnetic resonance image. Sagittal plane cut. Intramedullary hyperintense image in T1 in addition to medullary widening.

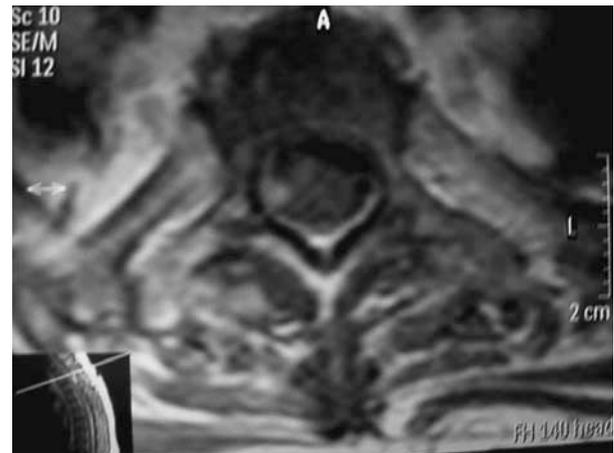


Figure 2. Axial plane cut showing a hyperintense medullary collection with a heterogeneous medullary aspect.

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/ μ L; platelets, 248 000/ μ L; 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/ μ L; lymphocytes, 210/ μ L; platelets, 65 000/ μ L. 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, nitrites (+). 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 medullary widening, a homogeneous intramedullary T1 and T2

homogeneous hyperintensity, corroborated on the axial cuts with heterogeneous images and medullary displacement (Figures 1 and 2) interpreted as a spinal hematoma in an organization phase with signs of medullary 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 medullary syndrome, with high-intensity pain, localized or in a radicular distribution, acute or related with a sensorymotor 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.¹¹

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.^{9,11-14}

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.^{4,11}

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,¹⁵ and the other one was secondary to obstetric trauma.¹⁶ Another 6 cases were related to anticoagulant therapy,⁴ as apparently our patient was and, finally, Matsumura et al¹⁴ 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.⁴

It is important to point out that, among the published cases of hematomyelia associated to anticoagulation, most were in an adequate range of anticoagulation according to the INR.⁴

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.

Intramedullary 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

1. Hylek EM, Singer DE. Risk factors for intracranial hemorrhage in outpatients taking warfarin. *Ann Intern Med.* 1994;120:891-902.
2. Constantini S, Ashkenazi E, Shoshan Y. Thoracic hematomyelia secondary to coumadin anticoagulant therapy: a case report. *Eur Neurol.* 1992;32:109-11.
3. Pisani R, Carta F, Guiducci G. Hematomyelia during anticoagulant therapy. *Surg Neurol.* 1985;24:578-80.
4. Pullakart VA, Kalapura T, Pincus M, Baskharoun R. Intraspinal hemorrhage complicating oral anticoagulant therapy. *Arch Intern Med.* 2000;160:237-40.
5. Koss WT, Bock F. Spontaneous multiple intramedullary hemorrhages. *J Neurosurg.* 1970;32:581-4.
6. Schenk VWD. Hemorrhages in spinal cord with syringomyelia in a patient with hemophilia. *Acta Neuropathol.* 1963;2:306-8.
7. Chen CJ, Hsu WC. Experience in the surgical management of spontaneous spinal epidural hematoma. *J Formos Med Assoc.* 1997;96:283-7.
8. Patel H, Boaz JC, Philips JP. Spontaneous spinal epidural hematoma in children. *Pediatr Neurol.* 1998;19:302-7.
9. Indakoetxea J. Protocolo diagnóstico de la paraplejia y tetraplejia. *Medicine.* 2005;8:5359-61.
10. Lorenzo AMA, Flores DE, Martínez FAI. Paraplejia secundaria a hemorragia medular. *Rehabilitación.* 2003;37:55-8.
11. Vadillo BA, González MA, Rebollo AAM. Patología vascular de la médula espinal. *Medicine.* 2003;8:4977-86.
12. Cakirer S, Basak M, Galip GM. Cervical hematomyelia secondary to oral anticoagulant therapy: case report. *Neuroradiology.* 2001;43:1087-8.
13. Avrahami E, Tadmor R, Ram Z, Feibel M, Itzhak Y. MR demonstration of spontaneous acute epidural hematoma of the thoracic spine. *Neuroradiology.* 1989;31:89-92.
14. Matsumura A, Ayuzawa S, Mikio D, Enomoto T, Takeuchi S, Yoshii Y, et al. Chronic progressive hematomyelia. Case reports and review of the literature. *Surg Neurol.* 1999;51:559-63.
15. Tubbs RS, Smyth MD, Wellons JC, Oakes J. Intramedullary hemorrhage in a neonate after lumbar puncture resulting in paraplegia: A case report. *Pediatrics.* 2004;113:1403-5.
16. Önal C, Yakinci C, Kocak A, Erguvan R, Tekiner A, Kutlu R, et al. Cervical hematomyelia: A rare entity in a neonate with cesarean section and surgical recovery. *Pediatr Neurosurg.* 2002;36:90-5.