Systemic lupus erythematosus flare up as acute spinal subarachnoid hemorrhage with bilateral lower limb paralysis

Xiang Yang,1 Seidu A. Richard,1,3 Jiagang Liu,1 Siqing Huang1

1Department of Neurosurgery, West China Hospital, Sichuan University, P.R. China; 2Department of Immunology, Jiangsu University, Jiangsu, P.R. China; 3Department of Surgery, Volta Regional Hospital, Ho, Ghana, West Africa

Abstract

Subarachnoid hemorrhage (SAH) is an uncommon complication of systemic lupus erythematosus (SLE). Solitary association of fatal spinal SAH as a complication of SLE, has not been encountered much in literature although coexisting acute cerebral and spinal SAH have been associated with SLE. We present a 39-year old female with initial diagnosis of SLE eight years ago who suddenly developed a productive cough, acute abdomen and paralysis of the lower limbs. Magnetic resonance imaging of the spine revealed thoracic spinal SAH with varying degrees of thoracic spinal cord compression. The hemorrhage was total evacuated via surgery. She regained normal function of her lower limbs after the operation with no further neurological complications. One of the rare but fatal complications of SLE is solitary spinal SAH without cranial involvement. The best and most appropriate management of this kind of presentation is surgical decompression of the hematoma with total hemostasis. The cause of hemorrhage should be identified intra-operatively and treated appropriately.

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease associated with complex-mediated lesions of blood vessels in multiple body organs, advancing to diverse kinds of vascular complications. Immune complexes that trigger vasculitis in the hematoma with total hemostasis. The cause of hemorrhage should be identified intra-operatively and treated appropriately.

Case Report

We present a 39-year old female with progressive multiple joint swelling associated with pain for eight years. Her symptoms become much severe two years ago and suddenly progressed into a productive cough and acute abdomen with paralysis of the lower limbs days (2) before she arrived at our outfit. She has a past history of suspicious cat scratch disease. Physical examination revealed thinning of hair, dry skin and mucosa membranes with ecchymosis and petechial rashes. Superficial lymph nodes were palpable but not significantly enlarged. There were rashes all over the face with no oral ulcers. Chest examination revealed shallow movements with bronchial breath sounds associated with wet rales. There was marked edema of lower extremities and multiple joint tenderness on palpation. There was sensory loss up to the xiphoid with saddle areas of numbness. The power on left leg was 0/5 while that of the right leg was 0/5. Babinski examination revealed up moving planter reflex.

Magnetic resonance imaging (MRI) revealed thoracic spinal SAH with varying degrees of thoracic spinal cord compression (Figure 1A and B). The nerve root of T3-T7 were compressed by the hemorrhage. CT-scan of the spine also confirmed hematoma (Figure 2A). Chest X-ray show congested lungs with patch opacities. Sputum examination did not yield any pathological organisms as the cause of the lung infection.

Liver function test and renal function test were normal. The hemoglobin was 77g/L indication anemia. Hepatitis B and C, HIV as well as syphils serolog ies was all negative. Full immunization investigation revealed positive antinuclear antibodies (ANA+), immunoglobulin G (IgG): 17.60 g/L, immunoglobulin A (IgA): 40.9g/L, complement C3: 0.69g/L. T-Score: –0.3. Anti-human globulin test: direct Coombs test: positive (+++), and indirect Coombs test: positive (++). Rheumatoid factor (RF) <20.0 U/mL, C-reactive protein (CRP) 3.96 mg/L, anti-streptolysin O (ASO) 28.901 U/mL, anti-cyclic citrullinated peptide antibody (CCP) 45.7 U/mL while anti-keratin antibodies (AKA) was negative (–). Erythrocyte sedimentation rate (ESR) was 60 mm/h. Our working diagnosis based on radiological findings and laboratory investigation above was spina SAH secondary to systemic lupus erythematosus.

The goal of surgery in this patient was to evacuate the blood that was causing the compressive symptoms while preserving the nerve root and also prevent recurrence of the hemorrhage. The patient was put on a prone position after general anesthesia. After marking the location of the hemor-
rhage and draping with povidone iodine, a 10-cm midline incision was made between T5 and T7 and extended to the subcutaneous, intramuscular up to the vertebrae. Osteotomy of the T5-T7 was done with high-speed drill through the lamina of the vertebrae. Further bone and ligament removal was done using the Kerrison rongeur. After securing hemostasis, the blood was seen superficially around the spinal nerve root and spinal cord to the right. The blood was complete evacuated without causing damage to the spinal nerve root during the surgery (Figure 2B). The muscles, subcutaneous tissues and the skin was closed in layers and wound dressing applied. Intra-operatively we notice the blood was extra-dura but compressing on the nerve roots. The patient had a good recovering. Post-operative CT scan done the next day shows no bleeding. The limb function gradually improved. Patient was discharge home two weeks after the operation by which time she could walk with support. A scheduled visit three months later revealed total recovery.

Discussion

Neurologic problems are common in cases of SLE, affecting up to 75% of patients. The symptoms include seizures, cranial nerve disorders, paralysis, peripheral neuropathy, and mental changes. These symptoms may wax and wane during the course of the disease. SAH either cranial or spinal is a potentially lethal complication in patients with SLE. Our case adds up to the few unusual presentations of SLE flaring up as acute spinal SAH with bilateral limb paralysis without cranial involvement. The first case was reported by Fody et al. in 1980. Cerebral vascular pathological changes as a result of autoimmunity in SLE patients includes cerebral thrombosis, cerebral infarction and cerebral hemorrhage which are serious threat to patients with SLE. We believe the cause of this spontaneous hemorrhage in our patient is due to thrombosis of bridging vessels as result of vasculitis.

Hemorrhage affecting the spinal cord in the general population is rare, and delayed diagnosis is often associated with poor outcome. Common causes of spinal SAH reported in literature includes arteriovenous fistula, syphilis, polyanergic nodosa, and aortic coarctation. Other causes of spontaneous subarachnoid hemorrhage of the cord include angioami and telangiectasis, solitary aneurysm, coarctation of the aorta, mycotic aneurysm of a spinal artery, various tumors (ependymoma, schwannoma, neurofibro-

Figure 1. A) T1 and B) T2 magnetic resonance imaging respectively showing subarachnoid hemorrhage with spinal cord and nerve root compression.

Figure 2. A) Computed tomographic scan image confirming subarachnoid hemorrhage; B) intraoperative image showing total evacuation of the hemorrhage.
that digital subtraction angiography (DSA) showed diffuse irregular narrowing and some occlusion of the medium and small arteries of the intracranial and spinal vasculature, suggesting vasculitis. Edward et al. with a similar case noted at autopsy that the spinal cord contained a large adherent blood clot dorsally, covering much of the thoracic region and extending to the cervical and lumbar levels, but not including the site of lumbar puncture. Lesions were not noted macroscopically. The hemorrhage ended abruptly at the foramen magnum.

Conclusions

One of the rare but fatal complications of SLE is solitary spinal SAH without cranial involvement. The best treatment option in our case after careful evaluation was spinal decompressing of the acute hemorrhage; identification of the cause of the hemorrhagic and tacking it appropriately was the very crucial part of the surgery. We managed the patients so as to avoid recurrence of the subarachnoid hemorrhage. After surgery, we recommend that patient be referred to rheumatologist to continue the management.

References

1. Torné R, Rodríguez-Hernández A, Bernard T, et al. Subarachnoid hemorrhage in systemic lupus erythematosus: Systematic review and report of three cases. Clin Neurol Neurosurg 2015;128:17-24.
2. Tang S, Lee C, Lee C, Jeng J. Systemic lupus erythematosus flare up manifestation as cerebral and spinal subarachnoid hemorrhage. Lupus 2011;20:1211-3.
3. Fody EP, Netsky MG, Mrak RE. Subarachnond spinal hemorrhage in a case of systemic lupus erythematous. Archiv Neurrol 1980;37:173-4.
4. Ramos-Casals M, Nardi N, Lagrutta M, et al. Vasculitis in systemic lupus erythematosus: prevalence and clinical characteristics in 670 patients. Medicine 2006;85:95-104.
5. Futrell N, Millikan C. Frequency, etiology, and prevention of stroke in patients with systemic lupus erythematosus. Stroke 1989;20:583-91.
6. Bernatsky S, Clarke A, Gladman D, et al. Mortality related to cerebrovascular disease in systemic lupus erythematosus. Lupus 2006;15:835-9.
7. Owada T, Takahashi K, Kita Y. Subarachnoid hemorrhage in systemic lupus erythematous in Japan: two case reports and a review of the literature. Modern Rheumatol 2009;19:573.
8. Baizabal Carvallo J, Cantú Brito C, Estañoñ B, García Ramos G. Subarachnoid hemorrhage as a complication of systemic lupus erythematous. Cerebrovasc Dis 2007;24:301-4.
9. Zhong W, Chen H, You C, et al. Spontaneous spinal epidural hematoma. J Clin Neurosci. 2011;72:218-21.
10. Penar P, Fischer D, Goodrich I, et al. Spontaneous spinal epidural hematoma. Int Surg 1986;72:218-21.
11. Baek BS, Hur JW, Kwon KY, Lee HK. Spontaneous spinal epidural hematoma. J Korean Neurosurg Soc 2008;44:40-2.
12. Matsumae M, Shimoda M, Shibuya N, et al. Spontaneous cervical epidural hematoma. Surg Neurol 1987;28:381-4.