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CASE REPORT

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Communicating Hydrocephalus in Systemic Lupus Erythematosus

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Abstract: Central nervous system (CNS) involvement has been recognized as the second leading cause of death in patients with systemic lupus erythematosus (SLE), although hydrocephalus is seen very rarely. We report a case of communicating hydrocephalus in a 24-year-old woman having previously diagnosed SLE without antiphospholipid antibody syndrome or cerebral venous angiographic abnormality. We propose direct damage-thrombosis of small sized venous structures or immune complex deposition within arachnoid villi, which impair cerebrospinal fluid flow, as a possible, yet unproven, pathophysiologic mechanism for hydrocephalus in SLE.

Key Words: Systemic lupus erythematosus, hydrocephalus, magnetic resonance imaging

Sistemik Lupus Eritematosumda Komminikan Hidrosefali

Özet: Sistemik lupus eritematosumda ölüm sebebi olarak santral sinir sistemi tutulumu ikinci sıklıkta tanımlanmasına rağmen hidrosefali oldukça nadir görülebilir. Daha önce SLE tanısı konan, antifosfolipid antikor sendromu olmayan ve serebral venöz anjiografisi normal olan 24 yaşındaki bir hastada komminikan hidrosefali vakasını sunduk. Araknoid villi içerisindeki immune complex birikimi yada küçük çaplı venlerin trombozu beyin omurilik sıvısının akımını bozabilir. Bu durum SLE'de izlenen hidrosefalinin patofizyolojik mekanizması açıklayabilir.

Anahtar Sözcükler: Sistemik Lupus Eritematosum, Hidrosefali, Magnetik Resonans Görüntüleme

Introduction

Systemic lupus erythematosus (SLE) is a chronic multisystem, autoimmune disease in which neuropsychiatric involvement occurs in about 50% of patients and carries a poor prognosis (1,2). Involvement of the central nervous system (CNS) is the second most frequent cause of death (1,2). The frequent neurological complications of SLE are aseptic meningitis, cerebrovascular disease, movement disorders, myelopathy and psychiatric symptoms (1,2). Although idiopathic intracranial hypertension is not uncommon in SLE, reports of hydrocephalus in SLE are very rare. We describe a case of communicating hydrocephalus in a 24-year-old woman with previously diagnosed SLE without antiphospholipid antibody syndrome or cerebral venous angiographic abnormality.

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Case Report

A 27-year-old woman with a one-year history of SLE was admitted with headache complaint. She first presented to the Rheumatology Department in August 2001 because of weakness, discoid rash, arthralgias, alopecia, low-grade fever, and photosensitivity. In laboratory examinations, antinuclear antibody was positive, anticardiolipin titers were normal, and Venereal Disease Research Laboratories (VDRL) test was negative. The patient was diagnosed as SLE with the presence of four criteria suggested by the American College of Rheumatology (3).

She complained of headache, blurred vision, dizziness and diplopia for one week. On admission to our hospital, she was afebrile and normotensive; mild alopecia and mild

tibial edema were noted. Neurological examination was entirely normal with normal pupillary reactivity. However, at funduscopic examination both optic discs were minimally swollen. Visual acuity was normal. Initial laboratory data revealed sedimentation rate of 50 mm/h, hemoglobin of 9.8 g/dl with normal indices, white blood count of $4.2x10^3/mm^3$, and platelet count of 350,000/mm^3. Urine sediment was unremarkable with absence of cells, casts and bacteria. Chest radiography and EKG were normal. EEG revealed slowing of all cortical background rhythms. Lumbar puncture produced clear and colorless cerebrospinal fluid (CSF) with protein level of 15 mg/dl; glucose was 4.1 mmol/L. Opening pressure was normal in the CSF (220 mmH_2O, in sitting posture).

Magnetic resonance (MR) examinations demonstrated large dilated ventricles. Both lateral ventricles, and third and fourth ventricles were prominently dilated, and all cisterns of the posterior fossa and suprasellar cistern were also dilated (Figure 1). All these findings suggested that the patient had prominent communicating hydrocephalus. No neural parenchymal lesion or pathologic contrast enhancement was found in this MR exam. Contrast enhanced magnetic resonance venography (MRV) was normal and all dural sinuses and large cortical veins were open and normal (Figure 2). Digital subtraction angiography (DSA) showed that all dural sinuses, and cortical and deep venous structures were normal (Figure 3). Cerebral circulation time, especially the venous phase, was longer than normal and both

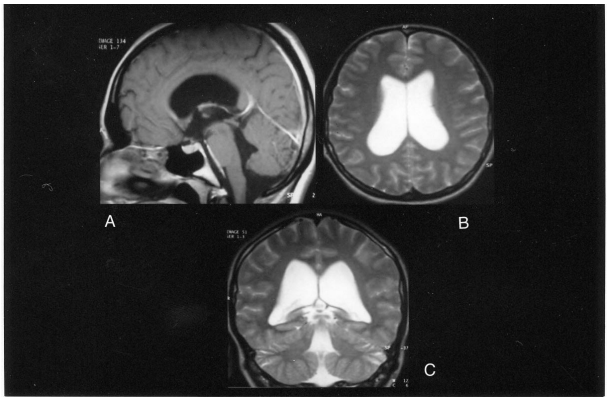

Figure 1. Sagittal T1-weighted with contrast enhancement (Fig. 1A), axial T2-weighted (Fig. 1B) and coronal T2-weighted (Fig. 1C) MR images demonstrate communicating hydrocephalus without neural parenchymal lesion or pathological contrast enhancement. Note wide posterior fossa cisterns, and fourth and third ventricles.

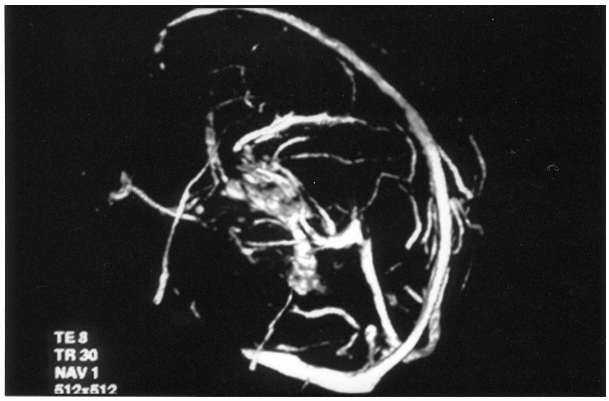

Figure 2. MRV (magnetic resonance venography) shows normal venous structures and dural sinuses.

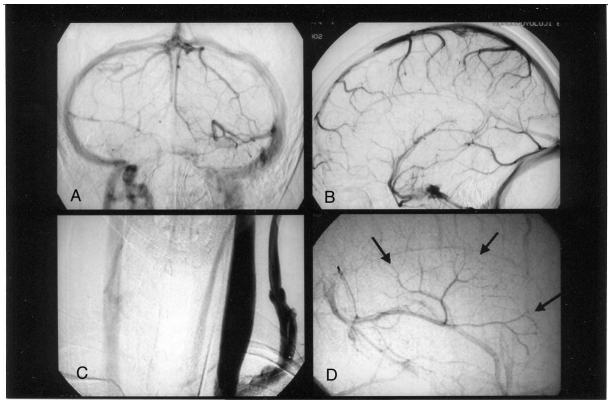

Figure 3. Digital subtraction angiography - AP (Fig. 3A) and LAT (Fig. 3B) images during venous phase show normal supra- and infratentorial venous structures, dural sinuses and bilateral patent jugular veins (Fig. 3C). Note only stretching of subependymal deep venous structures (arrows) due to enlargement of the lateral ventricles (Fig. 3D).

jugular veins were also patent (Figure 3C). Only stretching of subependymal veins was noted due to enlargement of the lateral ventricles from hydrocephalus (Figure 3D). All these findings suggested that the patient's communicating hydrocephalus resulted from direct damage-thrombosis of small sized venous structures (too small to be demonstrated on DSA) or immune complex deposition within arachnoid villi, which impair cerebrospinal fluid flow. Pulse steroid treatment was begun; 1 g/day iv methyl prednisone was given for three days and the dose was decreased to 1 mg/kg/day of oral steroid for two months. Within the second week, there was marked improvement in the patient's headache, blurred vision, dizziness, diplopia and general well-being. After two months, all symptoms had resolved

and the patient was headache-free. Follow-up neurological and eye examinations were normal. Follow-up MR examination revealed that the patient's communicating hydrocephalus was stable (Figure 4).

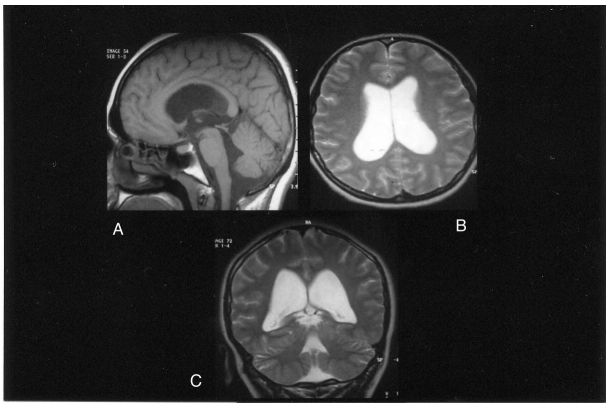

Figure 4. Follow-up MR examination six months after onset of symptoms - sagittal T1-weighted without contrast enhancement (Fig. 4A), axial T2-weighted (Fig. 4B) and coronal T2-weighted (Fig. 4C) MR images demonstrate communicating hydrocephalus is stable with no progression of findings.

Discussion

SLE is one of a family of rheumatic disorders, which include rheumatoid arthritis, scleroderma, polymyositis, dermatomyositis and Sjögren's syndrome. CNS involvement is reported to occur in 14-75% of SLE patients (1). These large differences in frequency depend on the diagnostic criteria applied, but 50% is thought to represent a reasonable figure (1,2). Neurological presentation, however, is uncommon and occurs in approximately 3% (1,2). The spectrum of neurological involvement is broad and includes psychosis, seizures, visual disturbances, organic brain syndromes, cerebrovascular disease, migraine, cranial neuropathies, myelopathy and focal neurological findings (1,2).

Of the more frequently encountered CNS complications, headaches, are especially common and usually benign in SLE. Common causes of headache in SLE include chronic tension-type headaches, migraine, dural sinus thrombosis and idiopathic intracranial hypertension. In idiopathic intracranial hypertension, symptoms and signs are those of elevated intracranial pressure alone without alteration of consciousness, with ventricles of normal size (normal CT scan), and CSF of normal composition (1,2,4,5).

Although idiopathic intracranial hypertension is not uncommon in SLE, reports of hydrocephalus in SLE are very rare (5-10). Patients with SLE treated with corticosteroids and other agents are at increased risk of opportunistic CNS infection and these infections may cause hydrocephalus; this group of patients is not included in our discussion (6). Hydrocephalus directly related to SLE is reported rarely in the literature. Kitching et al. (7) described two cases of communicating hydrocephalus and SLE with angiographically demonstrated cerebral phlebitis involving both deep and cortical veins. Cerebral angiography of both cases demonstrated marked irregularities of contour of superficial and deep cerebral veins. Postmortem examination of one of their patients showed infiltration of the leptomeninges and vascular lesions, consistent with meningitis. Periphlebitis and periarteritis were noted in the brain and leptomeninges. Thrombosis and recanalization were seen in veins and arteries. Borenstein and Jacobs (8) reported a 46-year-old woman with SLE and non-communicating hydrocephalus. They concluded that the cause of the non-communicating hydrocephalus was aqueductal stenosis caused by post inflammatory lesions of CNS lupus. In addition, normal pressure hydrocephalus in a 77-year-old patient with SLE was reported (9); no causal relationship was demonstrated. Finally, Mortifee et al. (10) reported communicating hydrocephalus in SLE with antiphospolipid antibody syndrome in a 24-year-old woman. They were unable to demonstrate thrombosis of superior sagittal or other major sinuses by either CT or MR imaging in their patient, and an angiogram was not obtained due to impaired renal function. They concluded without objective imaging findings that their patient's hypercoagulable state with cerebral venous thrombosis explained their patient's communicating hydrocephalus. We are aware of only four reported cases of hydrocephalus as a complication of SLE - three communicating hydrocephalus (2 with abnormal venous angiography and 1 SLE with antiphospholipid antibody syndrome) and one non-communicating hydrocephalus.

It is clear that our SLE patient had communicating hydrocephalus confirmed by MR imaging with negative antiphospholipid antibody syndrome. Angiography demonstrated moderate enlargement of lateral ventricles with stretching of the subependymal veins. However, there was no abnormality of cortical or deep cerebral veins, dural sinuses or jugular veins.

Several alternative mechanisms have been proposed to explain why and how patients with SLE produce circulating autoantibodies. The assumption is that the presence of these antibodies is indeed of pathological as well as diagnostic significance. It is proposed that autoreactive antibodies could play two roles in CNS involvement: direct injury to neuronal target cells and antibody-induced rheologic disturbances leading to vascular damage (1). The clear and consistently described pathological microthrombotic vascular changes perhaps add more direct support to the latter, although some interpretations of lupus CNS pathology offer at least indirect support for the former. The pathogenic mechanisms of idiopathic intracranial hypertension implicated include the general increase in coagulability (even in the absence of lupus anticoagulant), thrombosis of cerebral venous systems, and immune complex deposition within arachnoid villi, which impair cerebrospinal fluid flow. It is conceivable that similar pathophysiological mechanisms that explain the development of intracranial hypertension could also be evoked as explanations for some cases of communicating hydrocephalus. In the light of these pathological data, we think that our case's communicating hydrocephalus etiology probably resulted from direct damage-thrombosis of small sized venous structures or immune complex deposition within arachnoid villi, which impair cerebrospinal fluid flow. Since anticardiolipin titers were normal and VDRL was negative in our patient, we excluded the diagnosis of antiphospholipid antibody syndrome. Further studies are needed to understand and clarify these pathological mechanisms.

CNS involvement has been recognized as the second leading cause of death in patients with SLE; however, hydrocephalus in SLE may be seen only rarely. Radiological modalities such as CT, MR or DSA can be helpful to demonstrate hydrocephalus, to detect the type of hydrocephalus, to differentiate from other neurological involvement patterns and to understand the main cause of hydrocephalus (such as dural sinus thrombosis, post inflammatory lesion of CNS lupus, cortical-deep venous structure damage, etc.).

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