Communicating Hydrocephalus in Systemic Lupus Erythematosus

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Background: Central nervous system involvement is common in systemic lupus erythematosus but hydrocephalus, especially in children, is rare. **Case characteristics**: 6-year-old girl with systemic lupus erythematosus with nephritis, on treatment for four months prior to the presentation with features of raised intracranial pressure. **Observation**: Computed tomography revealed communicating hydrocephalus without any evidence of granulomatous lesion, infarction or thrombosis, with no features of lupus flare. Ventriculoperitoneal shunting provided symptomatic relief after failed medical management. **Message**: Hydrocephalus may be seen in systemic lupus erythematosus without tuberculosis or major vessel vasculitis.

Keywords: *Complication, Intracranial pressure, Vasculitis.*

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease in which neuropsychiatric manifestations occur in about 50% cases, and are the second common cause of death [1,2]. The frequent neurological complications are aseptic meningitis, stroke, movement disorders, myelopathy and psychiatric symptoms [1,2], but hydrocephalus is rarely reported, especially in childhood. We report communicating hydrocephalus in a 6-year-old girl with SLE without antiphospholipid antibody syndrome.

CASE REPORT

A 6-year-old female child was diagnosed four months prior to presentation as SLE with nephritis and hypertension with Anti-nuclear antibody and Anti-ds DNA (anti double stranded deoxyribonucleic acid) positive with low complement levels, and was treated with hydroxychloroquine (6 mg/kg), prednisolone (2 mg/kg for 2 months, then reduced and maintained at 0.5 mg/kg till present), mycophenolate mofetil, enalapril and nifedipine. Blood pressure returned to normal range within one month of treatment, and ESR within two months of above treatment. Four months after diagnosis, the child was admitted with headache, decreased vision and vomiting of over one month. On admission, she was afebrile, normotensive, with normal neurological examination and only light-perception. However, fundoscopy revealed bilateral papilledema. Complete blood count was normal with normal urine examination, ESR, C Reactive protein (quantitative), and complement levels. Chest *X*-ray and electrocardiogram were normal. Lumbar puncture revealed normal opening pressure with normal cell count, proteins and sugar. Differential diagnoses considered were optic neuropathy, central nervous system (CNS) infection like tuberculosis or HIV, and primary angiitis of CNS or secondary CNS involvement due to SLE.

Mantoux test, Quantiferon TB gold test and HIV serology were negative. The neuroimaging was delayed due to financial constraints (and was done 5 days later). Methyl prednisolone was given at the dose of 30 mg/kg/d for 5 days followed by oral prednisolone (2mg/kg for 15 days) that was tapered as a treatment for vision loss considering it to be optic neuropathy or CNS vasculitis as Computed Tomography demonstrated communicating hydrocephalus with peri-ventricular ooze. MRI was done, which revealed same findings as CT scan and also showed normal caliber major intracranial vessels, excluding major vessel vasculitis. On the fifth day after neuroimaging, intravenous mannitol was given followed by oral acetazolamide for one month. No significant improvement occurred after steroids and decongestive measures with repeat CT head showing similar features. The patient was referred for ventriculoperitoneal shunting, after which symptoms gradually improved over one month of follow-up.

DISCUSSION

SLE is one of the rheumatic disorder in which CNS involvement occurs in 14-75% of the cases [1]. These large differences in frequency depend on the diagnostic criteria applied, but 50% is thought to represent a reasonable estimate.

Hydrocephalus in SLE has been infrequently reported, and is due to varied etiologies [3-8]. Patients with SLE treated with corticosteroids are at increased risk of opportunistic infections which may cause hydrocephalus [4]. Kitching, *et al*. [5] 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 irregularities of contour of superficial and deep veins. Postmortem examination of one of their patient showed infiltration of leptomeninges and vascular lesions consistent with meningitis. Borenstein and Jacobs [6] reported a 46-year-old woman with non-communicating hydrocephalus. They concluded its cause was aqueductal stenosis due to post inflammatory lesions of CNS lupus. Antiphospholipid antibody syndrome is one of the common association with SLE which can lead to hypercoagulability and cerebral vein thrombosis. Mortifee, *et al*. [7] reported communicating hydrocephalus in SLE with antiphospholipid antibody syndrome in a 24-year-old woman. They concluded that the patient's hypercoagulable state with cerebral vein thrombosis explained the communicating hydrocephalus. The patient reported here had normal prothrombin time, activated partial thromboplastin time and was negative for antiphospholipid antibody.

It is proposed that autoreactive antibodies in SLE could play two roles in CNS involvement: direct injury to neuronal target cells and antibody induced rheological disturbances leading to vascular damage [1]. The pathogenic mechanism 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 CSF 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 this pathological data, we think that the hydrocephalus in this patient probably resulted from direct damage and thrombosis of small-sized venous structures or immune complex deposition within arachnoid vili, which impaired CSF flow. Further studies like specific serological tests, Magnetic resonance angioaraphy and venography, Positron emission tomography, stereotactic brain biopsy etc. are needed to understand and clarify these pathologic mechanisms.

Hydrocephalus, due to a variety of ethiologies, can be a complication of systemic lupus erythematosus. Timely detection of the disease and its cause can help in proper management and better outcome.

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