OBSTRUCTIVE HYDROCEPHALUS IN A PATIENT WITH SLE

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Abstract:

Hydrocephalous is a rare manifestation of systemic lupus erythematosus. Cerebral venous thrombosis, immune complex deposition within the arachnoid villi or direct post-inflammatory lesions of the central nervous system are possible causes of developing acute hydrochephalus. We report a case of acute non-communicating hydrocephalus secondary to stenosis of the aqueduct of Sylvius. The condition developed rapidly in a 22-year-old woman with previously diagnosed SLE.

Keywords: *Hydrocephalous; systemic lupus erythematous; stroke; pathogenesis.*

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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] Hydrocephalus in SLE are very rare. We describe a case of non-communicating hydrocephalus in a 22-year-old woman with previously diagnosed SLE without antiphospholipid antibody syndrome or cerebral venous angiographic abnormality.

Case presentation :

Ms. X, 22 years lady, known case of SLE for 1 year was admitted into Square Hospital with complaints of fever & loose motion for 2 days, several episodes of convulsion for 1 day. 1 year ago she was diagnosed as a case of SLE on the basis of hyperpigmentated rash on face & limbs & oral ulcer for 1 month along with positive ANA & dsDNA. She was treated with hydroxycholoroquine. Six months ago suddenly she developed lower limb weakness & after thorough investigation she was diagnosed as a case of non compressive transverse myelitis. She was treated with steroid & immunosuppressive drugs & improved. During admission, her GCS was 8/15. Pupil was dilated with sluggish reaction to light. Pulse - 130/min, blood pressure was nonrecordable. Patient was intubated for protection of airway & inotropes started. Her Hb% was 5.6 gm/dl, WBC – $15.1x10^9$ cells/cmm, PBF show features of hemolysis. Reticulocyte count was increased. CRP was 49.8 mg/dl, procalcitonin 17.1ng/ml. Escherichia coli was found in tracheal aspirates & urine culture. CXR showed features of pneumonia. CSF study revealed protein >300 mg/dl, glucose 29.0 mg/dl, WBC: 159 cells/cmm & PMN 85%. CSF culture revealed no growth. Her troponin I was elevated (2.97ng/ml), ECG – tachycardia, non-specific changes & echocardiography showed features of cardiomyopathy - global hypokinesia of LV, mild to moderate LV systolic dysfunction (EF-40-45%), moderate pericardial effusion. She was treated as a case of septic shock, bacterial meningitis, NSTEMI, pneumonia. MRI brain revealed recent infarcts involving the head of the left caudate nucleus, cranial lobe of both cerebellar hemispheres and both cerebellar vermis with obstructive hydrocephalus. MRV was normal. Gradually her GCS deteriorated. External ventricular drainage was applied on the next day. There was no improvement in the patient's clinical course despite supportive mechanical ventilation and appropriate antibiotic coverage. Unfortunately she died of sepsis.

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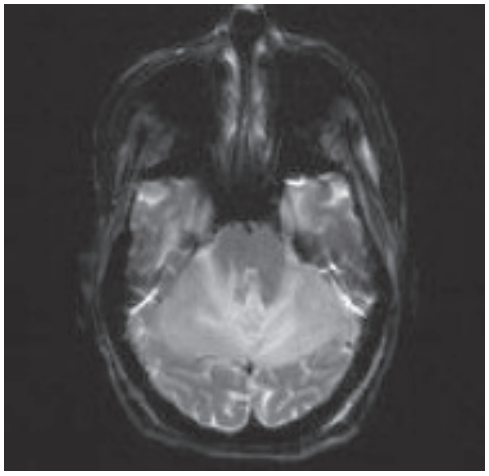

Fig.-1: *T2 image showing bilateral cerebellar infarct with compression of 4th ventricle*

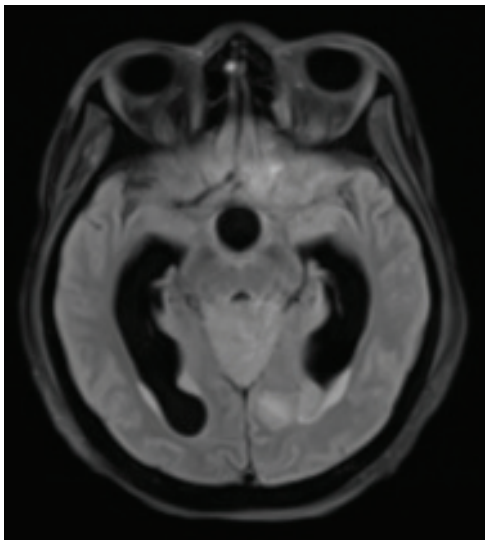

Fig.-2: *dilatation of lateral & 3rd ventricles with obliteration of basal cistern*

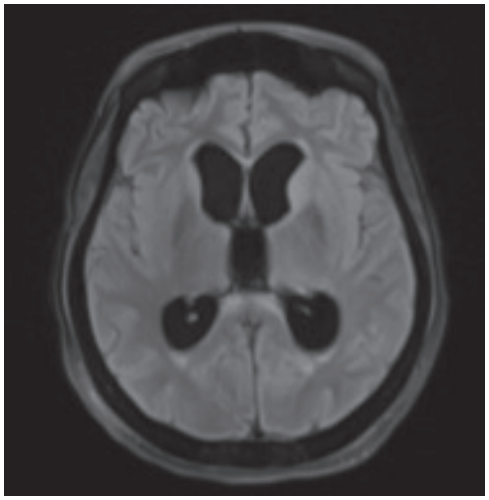

Fig.-3: *dilatation of 3rd & 4th ventricles*

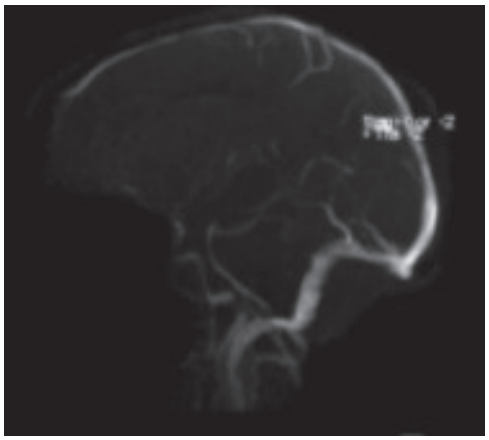

Fig.-4 : *normal MRV*

Discussion

Patients with SLE may experience a variety of neurological and psychiatric manifestations, collectively named NPSLE, that account for significant morbidity and mortality.[3] The prevalence of NPSLE varies widely, from 21% to 95% in various cohorts[4,5], in part due to the heterogeneity of manifestations and definitions used.[6] The neuropsychiatric manifestations of SLE classified as primary NPSLE & secondary NPSLE.

In primary NPSLE, direct neuronal injury due to autoantibodies against N-methyl-D-aspartate glutamate receptor (anti-NR2), accelerated atherosclerosis and thrombotic diathesis caused by the presence of anti-phospholipid are considered potential pathogenic mechanisms.[6] Secondary NPSLE may be caused by complications of the disease or its therapy, or may be unrelated to SLE and be due to infections, metabolic abnormalities and adverse drug reactions.

The pathogenesis and pathophysiologic mechanism of hydrocephalous associated with SLE are yet unproven although several alternative mechanisms have been proposed in the literature. [7] One hypothesis is that corticosteroids and other immune suppressive agents used in the treatment of SLE can lead to increased risk of opportunistic CNS infection, and these infections can lead to impairment of CSF drainage and subsequent hydrocephalus [8]. Normal pressure hydrocephalus in a 77-year-old patient with SLE has been reported, but no cause was found [9]. On the other hand, Krauss and associates showed a highly significant association between idiopathic NPH and arterial hypertension [10].

Verrees hypothesizes that the development of hypertension beyond the limits of cerebral autoregulation leads to breakdown of the blood brain

barrier in the cerebellum and development of posterior fossa edema secondary to focal transudation of protein and fluid. All of these studies show hypertension as an important contributing cause of NPH and obstructive hydrocephalus due to vascular encephalopathy [11]. In some lupus patients, hyperviscosity disrupt blood flow and might be involved in hydrocephalus[12]. Immune complex deposition can affect the brain parenchyma directly, or within the cerebrovascular system that can impair CSF flow into the arachnoid villi.

Thromboembolic formation that blocks the small arteries, choroid plexus or cerebral venous system can be conceived as another pathophysiologic mechanism to explain the development of intracranial hypertension.[10] Kitching et al. described two cases of communicating hydrocephalus in SLE patients, with cerebral phlebitis involving both deep and cortical veins demonstrated through angiography.[13] In postmortem examination of one of the patients, periphlebitis and periarteritis were noted in the brain and leptomeninges, and thrombosis and recanalization were seen in veins and arteries. Secondary antiphospholipid antibody syndrome [Hughes syndrome] is another cause of communicating-type hydrocephalus.[13,14,15] The hypercoagulable state caused by antiphospholipid antibodies increases the risk of developing generalized thrombosis in both arteries and veins. In rare cases, catastrophic antiphospholipid syndrome, associated with a high risk of death, may cause rapid organ failure usually painless, sudden onset of paralysis, loss of speech and intracranial hypertension syndrome.[16] Borenstein and Jacobs [13] reported the case of a 46-year-old woman with SLE and non-communicating hydrocephalus. They concluded that the cause of the noncommunicating hydrocephalus was aqueduct stenosis caused by post-inflammatory lesions of CNS lupus.

Our patient presented with convulsion with low GCS for 1 day. MRI revealed recent infarcts involving the head of the left caudate nucleus, cranial lobe of both cerebellar hemispheres and both cerebellar vermis resultant mass effect is causing compression over the 4th ventricle. Both lateral & 3rd ventricles are moderately dilated. CSF study showed high protein >300 mg/dl, glucose 29.0 mg/dl, WBC: 159 cells/cmm & PMN 85%. Though CSF culture revealed no growth, high protein, low sugar & neutrophilic leukocytosis – all are suggestive of bacterial meningitis. Bacterial infections can lead to impairment of CSF drainage & causing hydrocephalus. Another possibility may be due to CNS lupus. CNS lupus occurs during active stage of SLE, which may lead to bilateral cerebellar infarct with secondary occlusion of 4th ventricle leading to hydrocephalus. As patient has history of transverse myelitis 6 months back, this time involvement of brain may be most reliable explanation. May be both were present so rapid deterioration of her condition has occurred. MRV was normal & antiphospholipid antibody was negative , so cerebral venous sinus thrombosis was excluded. It should be mentioned that cerebral angiography, PET or MRS has greater sensitivity for detecting cerebritis & CNS vasculitis. As bacterial culture was negative, CSF for PCR of bacterial pathogens should be done. Unfortunately we could not obtain such investigation to strengthen our study.

Conclusion :

We can conclude that multiple pathogeneses are responsible for development of both communicating and non-communicating hydrocephalus. In the case reported here, CNS vasculitis & bacterial meningitis led to bilateral cerebellar ischemic infarct and brain edema. This resulted in secondary aqueduct stenosis and then to non-communicating hydrocephalus within 1 day. Since hydrocephalus is associated with significant morbidity and mortality, prevention is vital. Early diagnosis of hydrocephalus may play a key role in the choice of treatment strategy, may improve patient prognosis.

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