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Bilateral Papilledema In A Child Leading To The Discovery Of Systemic Lupus Erythematosus

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I. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that affects several organ systems, and progresses through relapses and remissions. The prevalence of systemic lupus erythematosus varies in the literature, ranging from 72.8 to 178 cases per 100,000 patients per year (1), and is generally found in patients of African and Asian origin. In the case of SLE, a complex immune system dysfunction affects several body systems. Various autoantibodies, such as anti-double-stranded DNA, anti-Ro, La, Sm, nucleosome, N-methyl-D-aspartate receptor and phospholipid, have been identified as the cause of the disease. The presentation and severity of SLE vary from individual to individual. SLE is rare in children, and optic neuropathy and intracranial hypertension have rarely been reported to coexist with SLE(2). In SLE, the prevalence of optic neuritis is 0.6% to 1% 6 and that of intracranial hypertension 1% to 1.5% 7,8 In this article, we describe a pediatric case of a patient with onset of active SLE revealed by idiopathic intracranial hypertension syndrome and bilateral optic neuropathy.

II. Observation:

A 10-year-old girl with no previous pathological history presented to the emergency department with a 5-day history of headache and jet vomiting. Given the picture of HTIC, the patient was referred to ophthalmology for fundus examination. On examination, the best corrected visual acuity was 10/10 in both

eyes. Both globes were normotonic with no oculomotor involvement. Direct and consensual photomotor reflexes were normal in both eyes, and anterior segment examination was normal. Fundus examination revealed bilateral stage 2 papilledema (Figure 1).

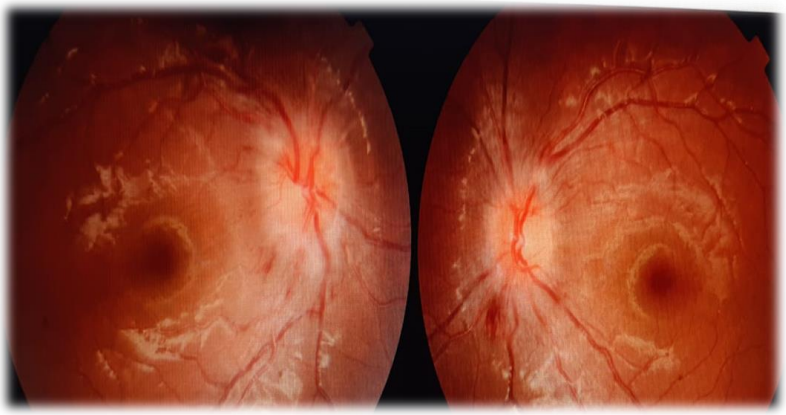

Figure 1: Stage 3 bilateral papilloedema

In view of the clinical HTIC syndrome, a cerebral CT scan was performed, which returned normal. Papillary OCT revealed thickening of the peripapillary retinal nerve fiber layer. Laboratory test results on the second day of admission were as follows: pancytopenia (hemoglobin: 8.1 g/dl, hematocrit: 22 g/dl, white blood cell [WBC] count: 1998/µl, platelet count: 40,000/µl), direct Coombs test: 3+; double-stranded DNA positivity, 1.80; antinuclear antibodies >1.5; homogeneous pattern 120; low complement C3, 0.13 g/L; low complement C4, 0.06 g/L; proteinuria with hematuria; anti-cardiolipin IgG negative; and anti-beta 2 glycoprotein 1 IgG/IgM/IgA positive . According to recent American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) 2019 criteria, 9 the diagnosis was juvenile SLE in this case. Neurological examination was normal. His headaches may have been symptomatic of an underlying neuropsychiatric complication of intracranial hypertension, with increased intracranial pressure leading to bilateral optic disc edema. Magnetic resonance imaging (MRI) revealed no mass-like lesions, but signs indicative of increased intracranial pressure were detected .

The patient was started on acetazolamide 250mg x 2/d with potassium supplementation. The patient progressed well on this treatment, with almost complete disappearance of papilledema after 1 month (Figure 2).

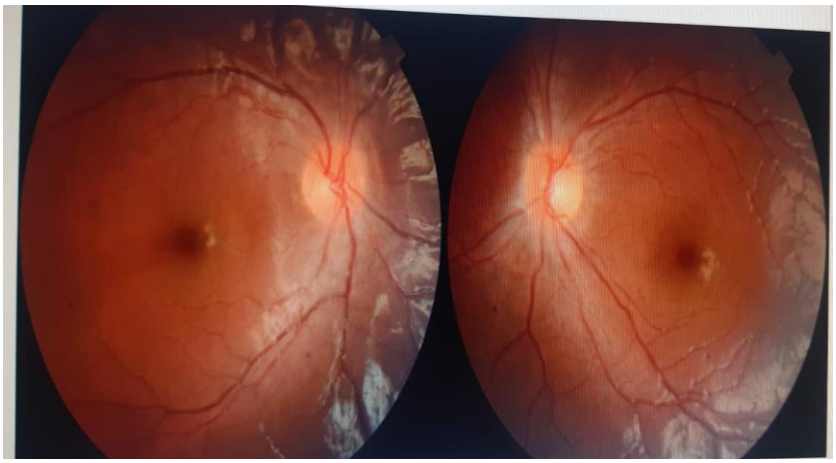

Figure 2: Retinophotography after 1 month of treatment

III. Discussion:

The severity of SLE depends on the organs involved in the immune processes. SLE can be difficult to diagnose, as its early signs and symptoms are non-specific and may resemble those of other diseases. Almost a third of SLE patients present with ophthalmic involvement (3); the most frequent association is keratoconjunctivitis sicca or dry eye syndrome, while the most serious ocular complication is retinal or optic nerve damage(4). Idiopathic intracranial hypertension, a cause of headache, and optic neuritis, a presentation of demyelinating disorders, are associated with LESP with central involvement.Headache is common, affecting around 38.3% of juvenile SLE patients and 72.5% of NPSLE patients. Intracranial hypertension is an

uncommon cause of intractable headache, presenting as the first symptom of active SLE. Numerous studies have reported intracranial hypertension as the first symptom of juvenile-onset SLE (5). Idiopathic intracranial hypertension was first reported by Bettman et al in 1968. Since then, several cases of SLE have been reported to be associated with intracranial hypertension. To date, however, the mechanism of increased intracranial pressure remains unknown, but is assumed to be associated with the vasculitis process, immune complex deposition or direct antibody damage to arachnoid villi, which blocks CSF absorption and ultimately causes elevated CSF pressure(6). In our case, the patient's headache may have been a symptom of an underlying neuropsychiatric complication of intracranial hypertension, with increased intracranial pressure leading to bilateral optic disc edema. As there are no specific criteria for the diagnosis of intracranial hypertension in patients with SLE, we adopted the modified Dandy criteria (7). The following criteria were met in our case: symptoms and signs of increased intracranial pressure, absence of neurological signs, CSF pressure >25 cmH2O, normal CSF profile and normal neuroimaging. The most serious complication is loss of vision due to severe papilledema, known as fulminant intracranial hypertension. . As the pathophysiology of intracranial hypertension involves excessive production of inflammatory cytokines and obstruction of the arachnoid villi by inflammatory cells, the basic treatment is high-dose corticosteroids, which can radically resolve symptoms. It can be difficult to confirm that SLE is the cause of intracranial hypertension, as there are currently no biological markers for this condition. Moreover, we had no pathological evidence. The clinical clue that supports the hypothesis that intracranial hypertension is due to SLE is that clinical onset was concomitant with the patient's active SLE state and rapid clinical response to treatment with intravenous corticosteroids.The course of SLE complicated by increased intracranial pressure and optic neuritis can be good with early steroid treatment as in our case. The complication of delayed or no treatment in such cases is deterioration in visual function.

Moreover, this is the first case of its kind; the long-term outcome after successful treatment can only be described after further follow-up.

IV. Conclusion

The association of optic neuropathy and intracranial hypertension in SLE is very rare; after a review of the literature, it was concluded that both conditions can occur simultaneously, as in our patient. On the other hand, either of these conditions may occur alone in cases of early- or late-onset SLE. In complicated cases, a problem-based approach enables underlying etiologies to be assessed, in-depth investigations to be carried out and the condition to be managed appropriately. There are several arguments in favor of early treatment for better results. Future studies may bring new findings to determine the exact pathophysiology and specific treatment.

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