OBSTRUCTIVE HYDROCEPHALUS DUE TO AQUEDUCTAL STENOSIS IN AN ELDERLY MAN WITH SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

Raiza Cansian Tuão^{1,*}, Mariana de Oliveira Macabú¹, Ketty Lysie Libardi Lira Machado¹, Bárbara Ferraço Dalmaso¹, Gustavo Pinto de Oliveira Gomes¹, Débora Marques Veghini¹, Ruben Horst Duque¹, Lídia Balarini da Silva¹, Ana Paula Espíndula Gianordoli¹, Luiza Correa Rodrigues Dalpiero¹, Érica Vieira Serrano¹, Weider Andrade Tomé¹, Luiza Vallory Alochio¹, Paula dos Santos Athayde¹, Isac Ribeiro Moulaz¹, Rodrigo Leite Marinho¹, Marcos Rosa Júnior¹, Valéria Valim¹

1.Universidade Federal do Espírito Santo, Vitória (ES), Brazil.

*Corresponding author: raizatuao@hotmail.com

BACKGROUND

Hydrocephalus, as a neurological manifestation of systemic lupus erythematosus (SLE), is extremely rare, with few cases reported in the literature. This complication may be an initial complaint at presentation or occur years after diagnosis, even in the absence of disease activity. Its pathogenesis has not yet been completely clarified, but in the case of obstructive hydrocephalus due to aqueductal stenosis, it is believed that the chronic inflammatory process of SLE causes the formation of an aqueductal membrane, culminating in stenosis and accumulation of cerebrospinal fluid.

CASE REPORT

A 64-year-old white male patient diagnosed with SLE since 2011, presenting with antinuclear antibody (ANA) 1/640 homogeneous pattern, photosensitivity, polyarthritis, and lupus nephritis. He was taking azathioprine between 2012 and 2014, going into remission. In February 2021, he was taking hydroxychloroquine 400 mg/day when he developed difficulty walking, holocranial headache, cognitive changes, amnesia, and inappropriate speech. On examination, he presented bradykinesia, ataxic gait, decreased muscle strength in the left lower limb (grade IV), and disorientation in time and space. Magnetic resonance imaging (MRI) of the skull showed voluminous supratentorial and aqueductal hydrocephalus, not communicating, exerting an expansive effect on the adjacent cerebral parenchyma (Figure 1), arching of the corpus callosum, and a small linear image located in the inferior part of the aqueduct, suggesting an aqueductal membrane (Figure 2). The findings were compatible with obstructive hydrocephalus due to stenosis of the cerebral aqueduct through the aqueductal membrane. The patient then underwent endoscopic ventriculostomy, progressing with improvement of symptoms, and was discharged with outpatient follow-up and return of azathioprine for immunosuppression.

Figure 1. MRI of the skull showing dilated lateral ventricles.

Figure 2. MRI of the skull suggesting the presence of an aqueductal membrane (red arrow).

CONCLUSION

The authors conclude that even in remission, in light of the chronicity of the disease, the patient with SLE can develop important neurological alterations, such as obstructive hydrocephalus. We stress the importance of not overlooking headaches and any new neurological symptoms, aiming at early diagnosis and medical intervention. We emphasize the relevance of clinical suspicion, detailed anamnesis, and meticulous physical examination for a meaningful evaluation of possible neurological complications of SLE. Early diagnosis of this condition and adequate treatment were essential for the successful outcome of the case.

KEYWORDS

Cerebral aqueduct, Hydrocephalus, Neurologic manifestations, Systemic lupus erythematosus.