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PSEUDOTUMOR CEREBRI AS A DIFFERENTIAL DIAGNOSIS FOR VISION LOSS IN SYSTEMIC LUPUS ERYTHEMATOSUS: CASE REPORT

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BACKGROUND

Pseudotumor cerebri (PTC) is an uncommon cause of vision loss in systemic lupus erythematosus (SLE). Disbalance between cerebral spinocellular fluid (CSF) absorption/production or cerebral veins microthrombosis/stenosis are proposed physiopathological mechanisms for PTC. Headache is the most common symptom, also, patients can present with peripheral bilateral vision loss, bilateral papilloedema, elevated intracranial pressure, normal CSF composition and normal brain imaging exams. Differential diagnosis include optic neuropathy and visual field defects. PTC must be recognized and treated quickly to preserve optic nerve. Most authors report a good response to corticosteroids in SLE patients with PTC. Still, in corticosteroid refractory cases, treatment can be done with acetazolamide, serial lumbar punctures, optic nerve sheath decompression and/or ventricular shunts. We report a PTC case on a patient in her first SLE flare, that did not occur with headache, and did not respond to corticosteroids.

CASE REPORT

An obese previously healthy 20-year-old female was admitted with a progressive four-months history of polyarthralgia, intermittent fever, asthenia, photosensitive malar erythema, non-healing alopecia and oral ulcers, and one month with occasional morning vomit, evolving with seizures and bilateral peripheral vision loss. She denied headache or orbital pain. Physical examination showed isochoric normal reactive pupils. Fundoscopy revealed bilateral papilloedema. Exams showed pancytopenia, elevated c-reactive protein and erythrocyte sedimentation rate, direct antiglobulin test 2+, hypocomplementemia, Anti-DNAds 1:80 and ANA 1:1280 with homogeneous dense fine speckled pattern. Antiphospholipid antibodies were negative. Brain CT and CSF without alterations, except for CSF opening manometry 52cmH2O. MRI of brain and orbits and Angio-MRI were also normal. A cerebral venous angiography was performed and did not find any sign of thrombosis. After therapy with pulse methylprednisolone, patient had significant improvement in all complaints except for peripheral vision loss. Azathioprine, Hydroxychloroquine and 1mg/kg/day Prednisone were initiated for SLE, also Acetazolamide and Topiramate, but patient progressed with gradual vision loss in four weeks. New fundoscopy revealed papilloedema reduction and bilateral atrophy of optic nerve. New CSF opening manometry was 38 cmH2O and increasing doses of Acetazolamide were performed and intensifying weight loss. The CSF opening manometry was maintained

in 20cmH2O with 2g/day Acetazolamide. Patient remains in outpatient follow-up with monthly CSF opening manometry and reports slowly gradual vision improvement.

CONCLUSION

Fundoscopy and CSF opening manometry must be considered in patients with SLE and red flag headache and/or vision loss. Prompt management for PTC can reduce vision sequelae. Unfortunately, this patient did not present a good response to steroid therapy.