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UNCOMMON OBSERVATION OF CHIASMATIC SARCOIDOSIS MIMICKING A PSEUDOTUMOR LESION

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ABSTRACT Sarcoidosis is a chronic multisystem disease characterized histologically by the accumulation of noncaseating epithelioid granulomas in the affected tissues. Chiasmatic sarcoidosis is rarely reported and presents a problem of differential diagnosis especially with gliomas and meningiomas. A 27-year-old patient with no previous pathological history presented with painless bilateral visual disturbances predominantly on the left, which rapidly progressed with moderate headaches over two months ago. Humphrey visual field study showed bilateral inferotemporal quadranopsia. Brain MRI showed an inflammatory aspect with thickening of the optic chiasm. Investigations studies revealed an elevated angiotensin-converting enzyme associated with hilar and mediastinal adenopathies. Histological study of the labial biopsy demonstrated sarcoid granulomas. Involvement of the optic chiasm by sarcoid granulomas has been reported in the literature and is most often misdiagnosed as gliomas or meningiomas,

KEYWORDS : sarcoidosis, chiasmatic lesion, sarcoid granuloma.

INTRODUCTION:

Sarcoidosis is a chronic multisystem disease characterized histologically by the accumulation of noncaseating epithelioid granulomas in the affected tissues. Neurologic manifestations are the presenting feature in approximately 50 to 75 % of patients with neurosarcoidosis [1]. It may present a diverse range of clinical features. Chiasmatic sarcoidosis is rarely reported and presents a problem of differential diagnosis especially with gliomas and meningiomas. Here we report a previously healthy patient who presented visual disturbance with headaches related to chiasmatic sarcoidosis associated with systemic involvement of the disease.

CASE REPORT:

A 27-year-old patient with no previous pathological history presented with painless bilateral visual disturbances predominantly on the left, which rapidly progressed with moderate headaches over two months ago. Ophthalmologic examination revealed a decrease in visual acuity (VA) of the left eye (2/10) with bilateral papilledema predominantly on the left. Humphrey visual field study showed bilateral inferotemporal quadranopsia. The neurological examination was unremarkable. Brain MRI showed an inflammatory aspect with thickening of the optic chiasm extending to the optic tracts and partially to the adjacent prechiasmatic segment of the left optic nerve (fig 1).

Biological investigations showed an elevated level of angiotensin-converting enzyme (ACE). Pituitary hormone levels and immunological tests were unremarkable. Lumbar puncture revealed normal protein, glucose, and leukocyte cell counts. Bilateral hilar and mediastinal adenopathies were found on chest CT scan. The labial biopsy demonstrated noncaseating granulomas. The diagnosis of systemic sarcoidosis with involvement of optic chiasm was established and the patient was treated with oral corticosteroids (1mg/kg/day). One month later, he suffered from arthritis involving principally his knees. In view of this joint arthritis and the severity of the chiasmatic lesion, an immunosuppressive treatment based on methotrexate was used in association with corticosteroids. The outcome at 3 months was marked by a clinical improvement of arthritis with significant recovery of the VA as well as a visual field study that became normal for both eyes.

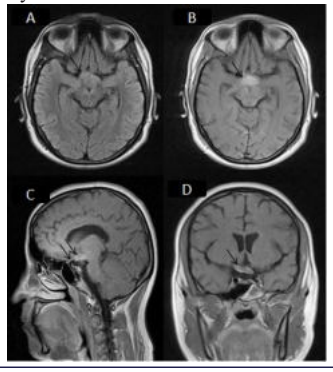

Figure 1: A: Axial fluid-attenuated inversion recovery showing hyperintensity with thickening of the optic chiasm that extends to optic tracts. B: Postcontrast axial T1-weighted image showing homogeneous enhancement of thickened optic chiasm that extends to optic tracts. C: Postcontrast sagital T1-weighted image demonstrating enhancement of thickened optic chiasm and left optic nerve that has normal size. D: Postcontrast coronal T1-weighted image showing enhancement of the enlarged optic chiasm.

DISCUSSION:

Sarcoidosis was first described in 1877 by Sir Jonathan Hutchinson as a disease of the skin. Today, it is recognized as a multisystem granulomatous disorder of unknown etiology [1]. Typical presentations include bilateral hilar adenopathy, pulmonary infiltration, and skin and eye lesions [1]. The diagnosis is determined by the presence of a compatible clinical presentation, supportive laboratory and radiological findings, confirmatory biopsy, and the exclusion of other probable causes of granulomatous inflammation [1]. The most common neurologic complication of sarcoidosis is cranial neuropathy, which occurs in more than half of patients with neurosarcoidosis [2]. In neurosarcoidosis, sarcoid granulomas may develop in the leptomeninges resulting in thickening of the arachnoid layer, particularly around the optic chiasm and basal cisterns [3].

Progressive chiasma damage typically evolves in four stages: inferior bitemporal quadranopsia if the low portion is involved or superior bitemporal quadranopsia if the superior part is affected; bitemporal hemianopia, bitemporal hemianopia with inferior binasal quadranopsia; and finally blindness [4]. Our patient presented a bilateral inferotemporal quadranopsia showing that the chiasmatic involvement was in its initial stage.

The diagnosis is challenging, especially in patients without systemic signs. Involvement of the optic chiasm by sarcoid granulomas has been reported in the literature and is most often misdiagnosed as gliomas or meningiomas, mainly in the cases without systemic signs of sarcoidosis [5]. In a study of 18 patients suspected of having chiasmatic gliomas, 14 of 18 had proven chiasmatic sarcoidosis after biopsy and histopathological study [6].

The radiological aspect of chiasmatic involvement found in our study showed a pseudotumoral appearance and was rarely reported in the literature. Our MRI findings are in agreement with the literature as optic pathway sarcoidosis can show different patterns including neural enhancement of the optic nerve, chiasmal enlargement, and increased T2 signal intensity in the optic tracts or radiations [7].

Histological confirmation is necessary if the diagnosis is uncertain or if there are no other systemic manifestations of sarcoidosis. Fortunately, our patient had an elevated ACE associated with hilar and mediastinal adenopathies with evidence of sarcoid granulomas on the histological study of the labial biopsy. These findings were consistent with

chiasmatic sarcoidosis, and thus biopsy of the chiasmatic lesion was considered not necessary in this situation. In cases where the clinical suspicion for sarcoidosis is sufficiently high without manifestations of other organ involvement, chest CT scan may be useful in identifying mediastinal or pulmonary involvement and may provide an alternative biopsy site if lymphadenopathy is identified [5].

Corticosteroids remain the mainstay of treatment for sarcoid granulomas. Other immunomodulatory agents, including methotrexate, azathioprine, mycophenolate mofetil, cyclophosphamide, and chlorambucil have been used for refractory disease or to prevent unacceptable corticosteroid-related complications [8, 9].

The prognosis is usually good and depends on the degree of severity of the visual damages and the presence or not of serious systemic findings [10]. The worse outcome is associated with CNS involvement, cardiac involvement, lupus pernio, stage III pulmonary disease, hepatosplenomegaly, and nephrocalcinosis [10]. Severe vision loss is uncommon and may occur with optic nerve involvement [10].

CONCLUSION:

Sarcoidosis can mimic a wide range of neurological disorders and may initially present as a chiasmatic mass. Therefore, the search for other systemic manifestations is crucial in establishing the diagnosis of neurosarcoidosis.

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