Sarcoidosis Mimicking Skull Base Meningioma

Sarcoidose mimetizando meningioma na base do crânio

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Abstract

Sarcoidosis is a systemic disease characterized by granulomatous inflammation. Pulmonary and lymphatic granulomatous involvement are common. We present a rare case report of involvement of the central nervous system affecting the ocular region and mimicking optic nerve sheath meningioma. We report the case of a 79-year-old female patient with progressive visual impairment with an evolution of 4 years. A magnetic resonance imaging scan of the cranium with gadolinium and intense homogeneous contrast enhancement revealed an expansive lesion in the right optic nerve, at the height of the optic canal. The patient was submitted to the neurosurgical approach with lesion biopsy, which showed sarcoidosis of the central nervous system. Due to the rarity of central nervous system involvement, the diagnosis of this pathology may unfortunately be postponed. The present article aims to elucidate this pathology as a differential diagnosis of retro-orbital tumors.

Keywords
- sarcoidosis
- ocular sarcoidosis

Resumo

Sarcoidose é uma doença sistêmica caracterizada por inflamação granulomatosa em que o envolvimento pulmonar e linfático é comum. Apresentamos um relato de caso raro de envolvimento do sistema nervoso central com acometimento ocular mimetizando meningioma da bainha do nervo óptico. Relatamos o caso de uma paciente de 79 anos, do sexo feminino, com diminuição visual progressiva com evolução de 4 anos. A ressonância magnética do crânio com gadolínio, mediante intenso realce homogêneo pelo meio de contraste, evidenciou uma lesão expansiva no nervo óptico direito, na altura do canal óptico. A paciente foi submetida a abordagem neurocirúrgica com biópsia de lesão, que evidenciou sarcoidose do sistema nervoso central. Devido à raridade do acometimento do sistema nervoso central, o diagnóstico dessa patologia pode ser, infelizmente, postergado. O presente artigo tem como objetivo elucidar a patologia como diagnóstico diferencial dos tumores retro-orbitários.

Palavras-Chave
- sarcoidose
- sarcoidose ocular

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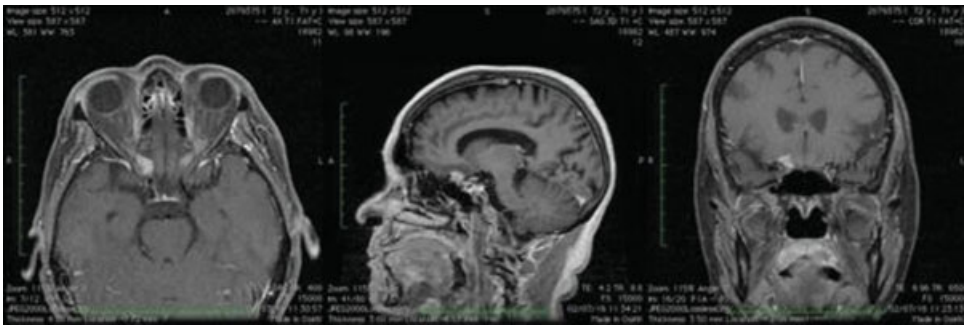

Fig. 1 Thickening with signs and enhancement by contrast of the sheaths of the optic nerve. An expansive lesion with intense homogeneous contrast enhancement in the right optic nerve with apparent implantation base on the sphenoid bone adjacent to the optic canal, measuring 1.3 × 0.9 cm.

Introduction

Sarcoidosis is a chronic multisystemic disease of unknown etiology in which there is an accumulation of non-caseating granulomas in the various tissues of the human body. It is a rare disease, and involvement of the central nervous system occurs in 5% to 10% of the cases. Therefore, the suspicion of involvement of the central nervous system should be considered in all patients diagnosed. However, according to Stern et al, in ∼ 48% of the cases of neurosarcoidosis, the symptomatology started with neurological alterations without previous diagnosis of sarcoidosis. Thus, the mass effect as the cause of the pathology is the only information available, which makes the diagnosis a challenge in the medical practice. Sarcoidosis lesions may resemble brain tumors, especially meningioma, so this pathology should be taken into consideration for the differential diagnosis of brain lesions. We report a case of neurosarcoidosis in a 79-year-old patient whose first symptom was progressive visual loss.

Case Story

A female patient, 79 years old, previously hypertensive, with hypothyroidism and depression, who was also being treated for Parkinson disease, started with a progressive picture of decrease in visual acuity to the right in the previous four years. Upon physical examination, the patient had: a score of 15 on the Glasgow scale; mydriatic pupils; amaurosis on the right, counting fingers to the left; decreased campimetry in the nasal field of the left eye, without other cranial nerve injuries; tremor in the right hand; bradykinesia to the right greater than to the left; bilateral Babinski sign; non-sustained clonus on the right; bilateral dysdiadochokinesia; preserved tactile, painful and vibratory sensibility and absence of meningism.

The patient was submitted to an investigation with computed tomography (CT) of the skull and magnetic resonance imaging (MRI) of the orbits (►**Fig. 1**), which evidenced an expansive lesion in the anterior fossa, obliterating the optic canal to the right. The lesion was initially treated as optic neuritis, and corticotherapy was performed. Lost in the follow-up, the patient presented worsening of the condition within 1 year, and a new investigation was necessary. In this interval, the patient presented complete loss of vision to the right and partial loss to the left, and underwent imaging exams that evidenced a nodular lesion to the right and hypersignal in the sheaths of the optic nerves bilaterally.

An investigation with cerebrospinal fluid (CSF) collection and a serological survey was performed, none of which evidenced alterations. The patient was submitted to the microsurgical approach of the lesion. Intraoperatively, a whitish lesion was observed near the optic nerve, with important adhesion and infiltration of the nerve, and we opted for partial resection. The anatomopathological analysis with classic non-caseating granulomas, consisting of densely arranged epithelioid cells associated with Langhans giant cells and/or foreign-body giant cells, was compatible with neurosarcoidosis (►**Fig. 2**).

Magnetic Resonance of the Skull

Histology

Discussion

Also called Heerfordt syndrome, sarcoidosis is a chronic granulomatous disease of unknown etiology that affects

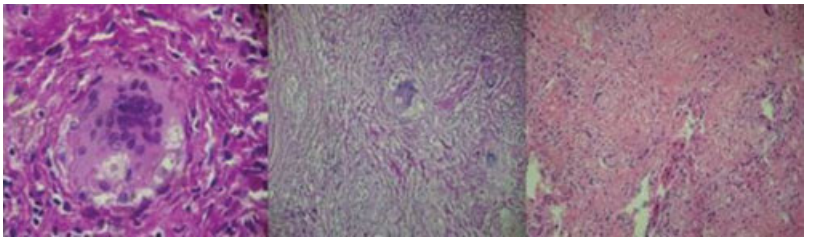

Fig. 2 Classic non-caseating granulomas consisting of densely-arranged epithelioid cells associated with Langhans giant cells and/or foreign-body giant cells.

young adults and reaches the nervous system in the systemic form in ~ 5% of the patients.[1]

The neurological manifestations described are paralysis of the cranial nerves, aseptic meningitis, peripheral neuropathy and myopathy.[2] Histologically, the development of granulomas, the activation of T cells and macrophages occur by a pathway mediated by the classic histocompatibility II complex, with an excessive Th1 response, leading to an overproduction of tumor necrosis factor-α (TNF-α) and interferon-gamma (IFN-γ), as well as interleukin-2 (IL-2) and interleukin-15 (IL-15), and the development of varying degrees of non-caseous necrosis.[3]

Clinically, the patients present diabetes insipidus, hypopituitarism and hyperprolactinemia due to hypothalamic involvement. According to a study by Fritz et al,[4] all cranial nerves may be affected, but the facial nerve and optic nerve present a higher prevalence, and unilateral involvement accounts for up to 65% of the cases, and bilateral involvement accounts for 35%.[4]

In association with these manifestations, vasculitis, convulsive seizures and hearing loss may occur.[5] Granulomas can coalesce, forming isolated intraparenchymal masses that are differential diagnoses of gliomas, Guillain-Barré syndrome, as well as HIV infection, mononucleosis, syphilis, acute porphyria, amyloidosis and multiple sclerosis.[3]

Usually, in neurosarcoidosis there is involvement of basal leptomeninges, causing abnormalities in the cranial nerves or hydrocephalus.[6,7] It is important in these cases to perform the differential diagnosis with carcinomatous meningitis and syphilis.[6,7] Neurosarcoidosis rarely presents itself solely mimicking a brain tumor, such as a meningioma.[8–10] Complementary exams, such as dosing of angiotensin-converting enzyme, serum alkaline phosphatase, and liquor and serum calcium, help in the diagnosis, but they are unspecific. The CSF has a mononuclear pleocytosis pattern, elevated proteins, and the presence of oligoclonal bands.[5] Imaging exams, such as cranial CT and MRI, may reveal hydrocephalus, meningeal enhancement and parenchymal mass, but are unspecific for the diagnosis.[3] Lesions with increased intensity in T2 at the junction of the gray and white substances are highly suggestive of this diagnosis, especially when associated with the enhancement of the meninges and with the hypothalamus lesion.[7,8,10] Only a minority of cases need histological confirmation with non-caseating granulomas in the affected nervous system tissue.[4,9] The diagnosis is of exclusion, but a probable diagnosis is defined as evidence of inflammation of the nervous system on the MRI or CSF with high protein and cellularity, G index of immunoglobin or the presence of oligoclonal bands in combination with evidence of systemic sarcoidosis with histological confirmation.[4,9]

Isolated cranial nerve abnormalities and aseptic meningitis present low risk of progression, with the exception of occasional cases of progressive optic neuropathy, and they respond well to corticosteroids.[7] However, patients with mass lesions, with leptomeningeal involvement with multiple anomalies of the cranial nerves, with spinal cord disease and with hydrocephalus often require high doses and prolonged course of corticoids associated with immunosuppressants.[7] Among the immunosuppressants used are methotrexate (MTX), azathioprine (AZA) and mycophenolate mofetil (MMF), which are equally efficient. Infliximab is a monoclonal antibody, but with caveats for oncologic patients and the possibility of increased risk of tuberculosis reactivation.[7] The diagnosis and management of neurosarcoidosis are still challenging aspects of the disease, as well as its recognition as a differential diagnosis.

It is important to consider the possibility of neurosarcoidosis in the differential diagnosis of brain expansive lesions pre- and intraoperatively. Thus, recognizing this condition leads to the proper clinical treatment with corticosteroids, avoiding an unnecessary extensive surgical treatment.

Conflict of Interests
The authors have none to declare.

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