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EXTENSIVE NEUROSARCOIDOSIS AND OPTIC NERVE COMPLICATIONS

J. El Mouedden, M.C. Duray, D. Hernalsteen, G. Cosnard[1]

We present the case of a 35-year-old patient suffering from nasal obstruction and headache for 3 years. The patient was hospitalized for a recent and progressive decline of vision of the right eye associated with afferent pupillary deficit and inferior altitudinal hemianopsia. He was diagnosed with systemic sarcoidosis involving the central nervous system as illustrated by magnetic resonance imaging (MRI) scans showing different type diffuse lesions of meningo-encephalitis. Our case is characterized by severe cerebral pachyleptomeningeal lesions complicated by optic nerve compression and cervical spinal cord damage. MRI value of diagnosis for systemic neurosarcoidosis was supported by histological examination of a biopsy of the sphenoid sinus lesions that showed epithelioid granulomas presence without caseous necrosis. Thus, MRI of the brain and spinal cord is a powerful tool method in monitoring and diagnosing aymptomatic and symptomatic neurosarcoïodosis. MRI is also a powerful tool in monitoring the neurosarcoidosis during therapeutic treatments.

Key-word: Sarcoidosis.

Sarcoidosis is a granulomatous inflammatory disease that can affects almost any organ and many parts of the body. This disease is termed neurosarcoidosis when the nervous system is involved. Although sarcoidosis is well described in literature, however, the cause of the disease is still not understood.

Neurosarcoidosis may be asymptomatic or present debilitating chronic condition such as headache, cranial nerve palsy, seizures, paresis and paresthesias. The prevalence of neurosarcoidosis is estimated to be one-fourth of the systemic sarcoidosis patients that have histological evidence of central nervous system involvement.

Herein, we report the MR findings of a 35-year-old patient with neurosarcoidosis. The lesions were characterized by pachy- and leptomeningeal lesions complicated by optic nerve compression and cervical spinal cord damage.

Case report

A 35-year-old patient, who had complaints of nasal obstruction and headache for 3 years was hospitalized for a recent decline and progressive loss vision of the right eye. Clinical examination showed a grasping and behavioral disorder of frontal lobe type, afferent pupillary deficit and inferior altitudinal hemianopsia right. Lumbar puncture showed slight increase of protein and glucose concentration, and oligoclonal bands identical to those of serum. Biology showed high levels of converting enzyme angiotensin.

In the year 2006, he underwent resection of cervical lymphadenopathy with the diagnosis of sarcoidosis. MR images showed thick, nodular pachymeningeal lesion and leptomeningeal extension in frontal (Fig. 1, 2) and right temporal area (Fig. 3) associated to meningeal enhancement in the hypothalamo-pituitary area (Fig. 2).

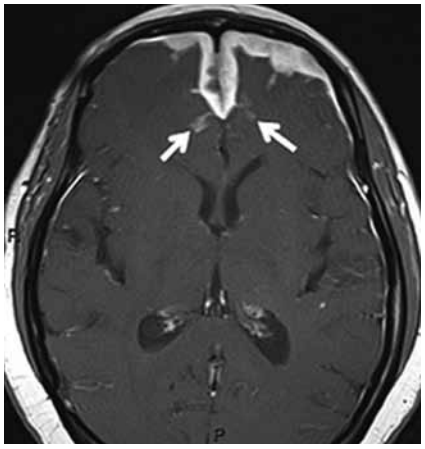

Fig. 1. — Axial T1-weighted gadolinium contrast-enhanced MR image shows a thick nodular pachymeningeal lesion and leptomeningeal extension (arrows).

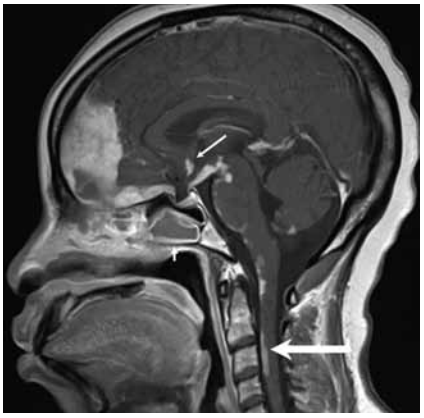

Fig. 2. — Sagittal T1-weighted contrast-enhanced image shows basal meningeal enhancement extended to pituitary area (thin arrow), brainstem and sphenoid sinus (arraowhead). Cervical spinal cord myelo-meningitis is also observed (thick arrow).

Right optic nerve was compressed and stretched by pachymeningitis. He also had multifocal leptomeningeal enhancement of the spinal cord and cervico-dorsal myelitis. Biopsy of the sphenoid sinus lesions showed epithelioid granulomas without caseous necrosis.

Discussion

Neurosarcoidosis is a serious clinical affection (1.2) which manifests in 5% of cases of sarcoidosis and is observed in 14 to 27% of autopsy series (1-5). The clinical manifestations are highly polymorphic, depending on the location, size of lesions and their evolution (1, 3, 6).

Histologically, there is an accumulation of epithelioid granulomas without caseous necrosis affecting preferentially the leptomeninges of the skull base (2) (Fig. 1 and 2) but also pachy meninges around the brain parenchyma and cranial nerves (1, 3-7). MRI can detect neurological lesions in 10% of patients with multisystem sarcoidosis (4) and is the imaging modality of choice in the diagnosis and follow-up of nervous involvement of sarcoidosis, even if the lesions are highly polymorphic (1, 3, 4, 6-7).

The finding on the imaging examinations are however nonspecific. The differential diagnosis with other diseases such as tuberculosis, Wegener's granulomatosis, fungal meningitis, lymphoma, and meningeal carcinomatosis can be difficult. In neurosarcoidosis, the clinical presentation is often of low expression or even asymptomatic, in discrepancy with the major radiolog-

From: 1. Department of Medical Imaging, UCL Saint Luc, Catholic University of Louvain, Brussels, Belgium.
Address for correspondence: Dr J. El Mouedden, M.D., Department of Medical Imaging, UCL Saint Luc, Catholic University of Louvain, Avenue Hippocrate 10, B-1200 Brussels, Belgium.

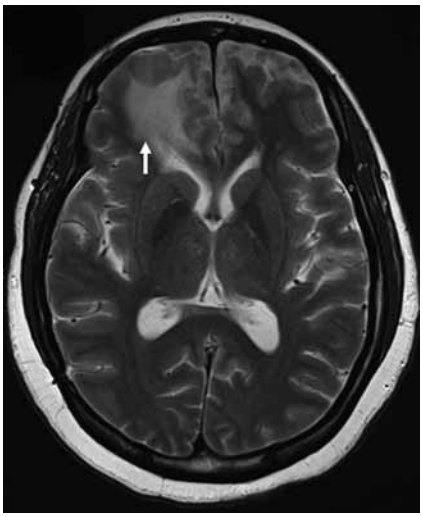

Fig. 3. — Axial T2-weighted image shows meningeal lesion associated with high signal intensity on T2 of the subcortical right frontal white matter (arrow).

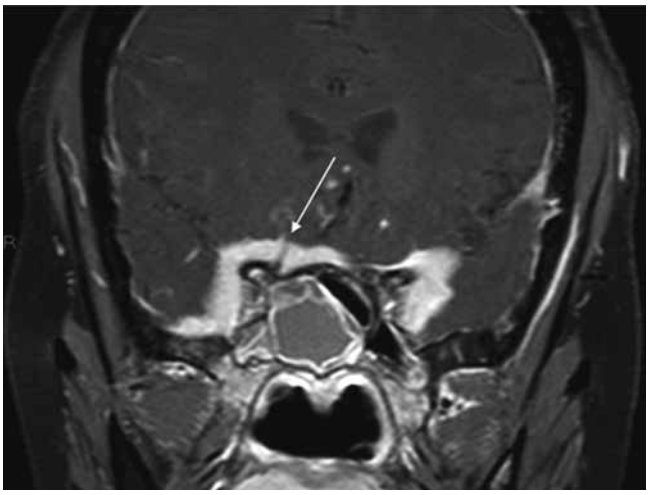

Fig. 3. — Axial T2-weighted image shows meningeal lesion associated with high signal intensity on T2 of the subcortical right frontal white matter (arrow).

ical extension lesions. The association of leptomeningeal enhancing lesions and hypothalamic-pituitary after injection of gadolinium is most suggestive of diagnosis (1) (Fig. 1 and 2).

In one third of cases, there is cranial nerve damages (4.8) often symptomatic, sometimes with a wide predilection for the facial and the optic nerve (3-4, 8-9). In our case there was an excellent correlation between radiological observation and clinical symptoms. Oculomotor nerve damage is very rare and is mainly secondary to intracranial hypertension (3, 6). These lesions were observed frequently in women (4). Optic nerve damage may be primary (unilateral or bilateral) or secondary to chiasmatic lesions (6, 8). Nerve damages are of 2 types: either by perivascular lymphocytic infiltration (3) or more often, as observed in our case, compression by granulomatous pachymeningitis (Fig. 4).

Involvement of the brain parenchyma himself is rarely observed. If so, lesions are caused by the extension of sarcoid granulomas from the meninges into the spaces of Virchow-Robin and present as nodular or pseudo-tumor lesions enhancing after intravenous gadolinium (1, 3-6).

These tumor-like lesions, however, are the most clinically symptomatic (4). It may also present as white matter lesions, of high signal intensity on T2, and non enhancing after gadolinium injection (Fig. 3). They are either gliotic scars of deep inflammatory lesions or lesions secondary to microangiopathy sarcoidosis (10). Moreover, acute ischemic injury in a young person without known etiology must evocate the diagnosis of neurosarcoidosis (11). Lesions of cerebral white matter are considered irreversible (2), have no clinical correlation, and do not always regress under immunosuppressive therapy (4, 9).

A spine cord myelo-meningtis is classic but not systematic in a neurosarcoïodosis. It has the same radiological and histopathological features as brain damage and affects most of the entire cervical spine (3.4).

A normal MRI does not exclude the diagnosis of neurosarcoidosis, especially in patients with only cranial neuropathy (3, 6) or in patients previously treated with corticosteroids (1).

The imaging can assess the response to immunosuppressive therapy (3, 4). The regression of lesions on MRI is often delayed compared with clinical improvement (3, 6).

Conclusion

Our patient is diagnosed with neurosarcoidosis, presenting as lesions of the pachy- and leptomeninges of brain and cervical spine. Our report confirms other similar observations already described in literature by Authors using MRI. Thus, our observations support the usefulness of MRI as a powerful tool for diagnosing and monitoring symptomatic neurosarcoidosis.

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