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Inaugural neurosarcoidosis revealing the systemic disease

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ABSTRACT:

Sarcoidosis is systemic granulomatosis of unknown origin. The neurological involvement is rare. We report the case of a 52-year-old patient whose inaugural neurosarcoidosis is evoked in MRI. The diagnosis of sarcoidosis has been proved through clinical, biological, and histological arguments, imaging aspect, and evolution of signs. MRI is an essential imaging tool that detects inaugural neurosarcoidosis, monitors neurological damage, and controls the efficiency of treatment.

KEYWORDS: Neurosarcoidosis ; Diagnosis ; MRI

INTRODUCTION:

Sarcoidosis is systemic granulomatosis of unknown origin. It usually affects young adults and has essentially thoracic lymphadenopathy and ocular tropism. The neurological involvement of sarcoidosis is rare and can be the first presentation in some cases.

We report the case of a 52-year-old man who presented an inaugural neurosarcoidosis. We underline, through this article, the diagnostic strategy of this disease, and the place of MRI in its management.

OBSERVATION:

We report the case of a 52-year-old man with a well-balanced type 2 diabetes, who was admitted for the heaviness of speech and cerebellar syndrome for 15 days.

The clinical examination finds dysarthria and dyschronometry in a conscious patient (GCS=15), nonfebrile, and without sensory deficits.

A cerebral MRI was performed and objectified hyperintense T2 and T2 FLAIR lesions involving the hypothalamus, mamillary bodies, optic nerves, and the white matter of both cerebellar hemispheres. Post-contrast T1 showed a nodular enhancement on the hypothalamus and mammillary bodies, associated with diffuse micronodular optochiasmatic enhancement. The signal abnormalities of the cerebellar white matter did not show enhancement (Figure 1). MRI aspect evoked a granulomatous origin.

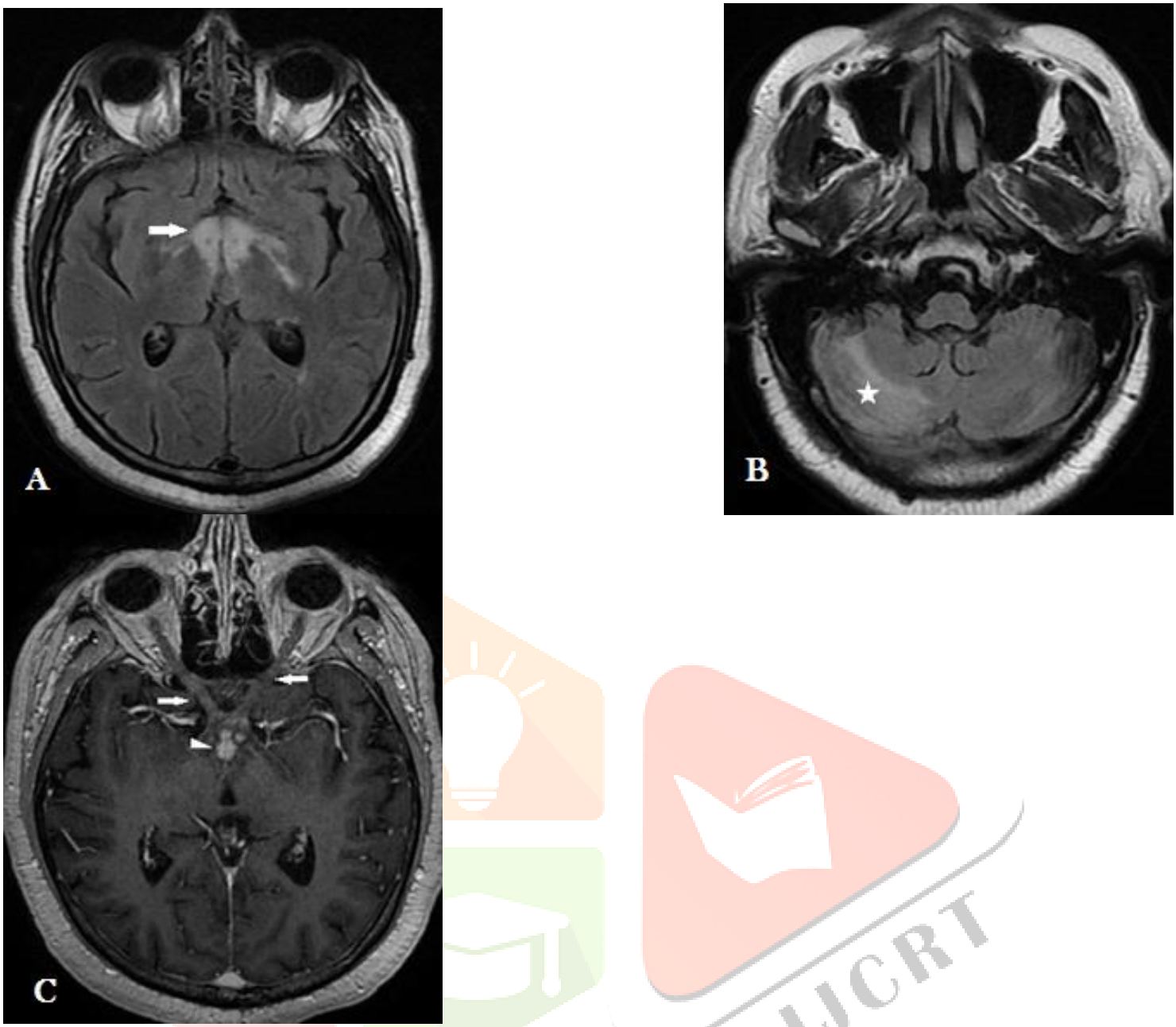

Figure 1: Cerebral MRI in T2 FLAIR sequences (A and B) and post-contrast T1 (C). A: The image shows hypothalamic hyperintense lesions, which extend to optic nerves (arrow).
B: Hyperintensity in the white matter cerebellar (asterisk).
C: Nodular enhancement of the mamillary bodies (arrowhead) and micronodular optochiasmatic enhancement (arrows).

The cerebrospinal fluid test revealed hyperproteinorachia (1.17 g/l), normal glyorachia, and negative cellularity.

A thoracic CT was performed (Figure 2). It showed bilateral hypodense homogeneous non-compressive mediastino-hilar adenopathies, without pulmonary parenchymal lesion.

The blood test showed hypercalcemia of 109 mg/l, hyperproteinorrachia (0.2g/l), and a rise of the angiotensin-converting enzyme (171 U/l). We note a normal rate of glycemia.

Tuberculin intradermal test and Genexpert MTB were negative.

Salivary gland biopsy confirmed the existence of gigantocellular epitheloid granuloma without caseous necrosis. That was compatible with sarcoidosis.

The patient's follow-up showed a favorable clinical evolution, and an apparent regression of lesions under corticoid, demonstrated by the control cerebral MRI performed in the second month of treatment (Figure 3).

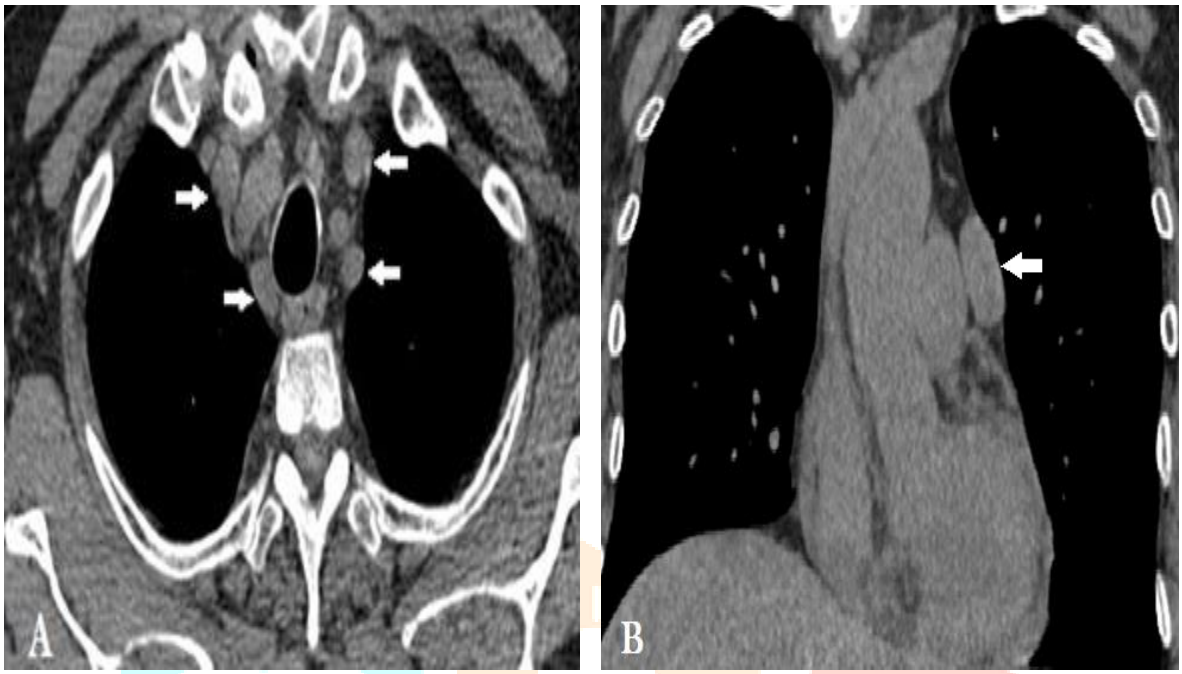

Figure 2: Thoracic CT in the mediastinal window in axial (A) and coronal (B) sections. Images show hypodense homogeneous and non-compressive mediastinal adenopathies.

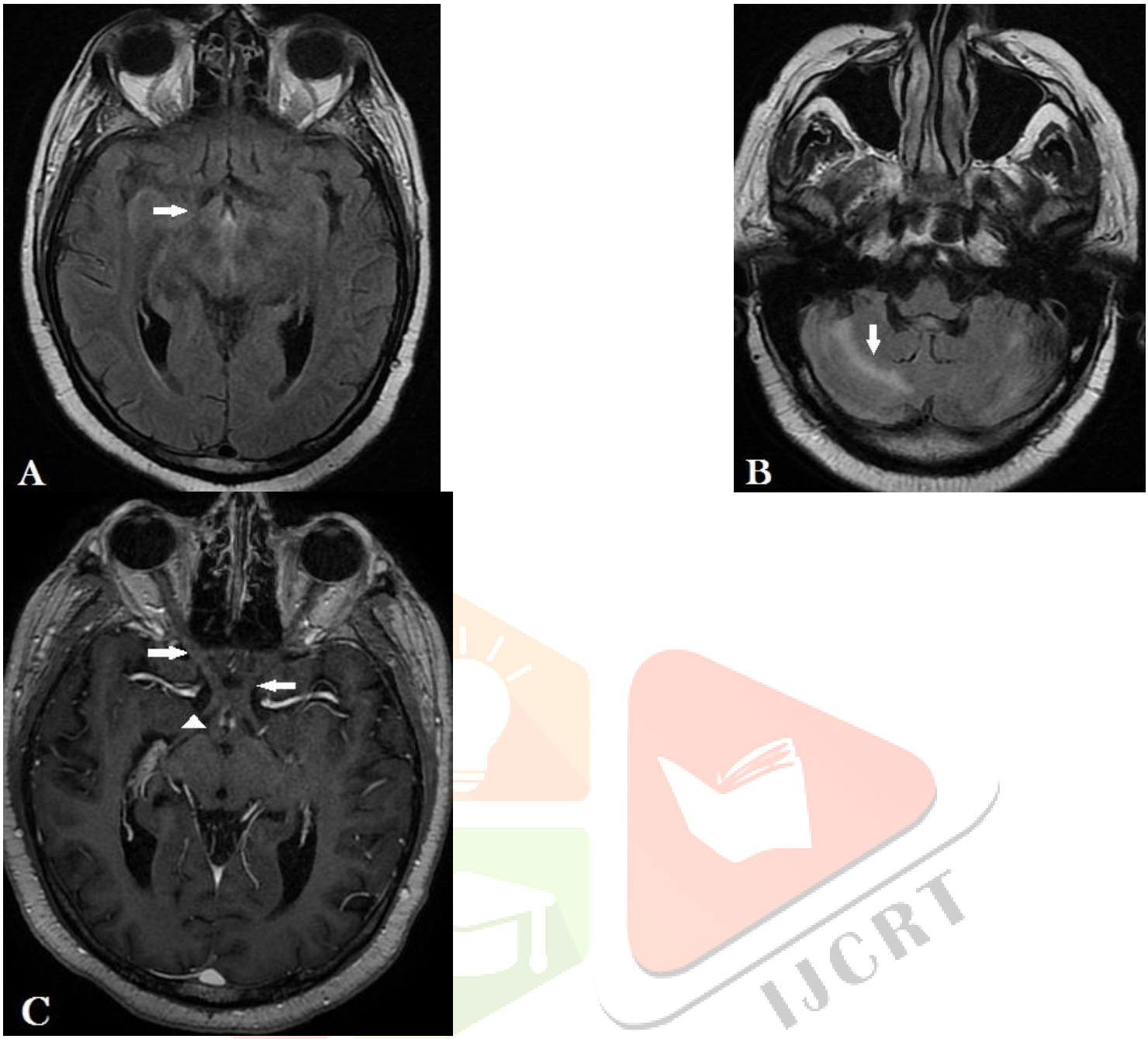

Figure 3: Cerebral MRI after two months of corticosteroid therapy, in T2 Flair (A and B) and post-contrast T1 (C). Images show an apparent regression of signal abnormalities on the hypothalamus (A: arrow), with a stable appearance of cerebellar white matter involvement (B: arrow). There is a decrease in the nodular enhancement of the mamillary bodies (C: arrowhead), and micronodular optochiasmatic enhancement (C: arrows).

DISCUSSION:

Clinical manifestations of neurosarcoidosis are nonspecific; they remain dominated by dysfunction of cranial nerves, especially optic (II) and facial (VII) nerves, and hypothalamic-pituitary dysfunction. Headaches are frequently described as well as seizures, psychiatric disorders, and motor and sensory deficits. Vascular involvement can lead to cerebrovascular accidents that are most often ischemic [1 , 2].

The diagnosis of neurosarcoidosis is easily retained in a patient followed for sarcoidosis, by highlighting the neurological lesions affecting the preferential localization of the disease, on a cerebral and medullary MRI. The neurological involvement can be inaugural and may herald systemic sarcoidosis [3,4]. Magnetic resonance imaging (MRI) is the technic of choice to evoke the diagnosis through the finding of typical lesions. The MRI exploration protocol should include T2-weighed and T2 FLAIR sequences which detect white matter abnormalities, Gadolinium-free, and Gadolinium-injected T1

sequences for the investigation of meningeal and parenchymal lesions, and MRI-angiographic sequences that aid to find signs of vasculitis. However, a normal MRI does not eliminate the diagnosis in case of strong clinical suspicion. Conversely, the lesions described in the MRI do not necessarily correlate with the clinical presentation [5, 6].

Leptomeningeal involvement is the most frequent. It is localized preferentially at the base of the skull, the hypothalamic-pituitary axis, the optic chiasma, and the cranial nerves. We find especially intense micronodular leptomeningeal enhancement after gadolinium injection [5].

Pachymeningeal involvement is characterized by the presence of one or more extra-axial masses, that shows iso-signal on T1-weighted images, hypo-signal on T2-weighted, and significant enhancement after Gadolinium.

The first differential diagnosis of an extra-axial mass is the meningioma, which is usually T2 iso or hyperintense. Fibroblastic or calcified meningioma appears in hypo-signal on T2-weighted images like the sarcoïdosic masses. However, the meningeal thickening that is enhanced by Gadolinium in contact with the tumor is typically found in meningioma [2, 6].

In the case of diffuse meningeal enhancement, other causes of meningitis will be evoked, dominated by tuberculous meningitis, which is the principal differential diagnosis [2].

The meningeal involvement in our patient has typical localization and appearance. It is compatible with the leptomeningeal neurosarcoidosis and shows a favourable evolution under corticoid (Figure 3).

The parenchymal involvement is the consequence of leptomeningeal neurosarcoidosis extension, via the perivascular spaces of Virchow-Robin. Lesions may appear as multiple disseminated nodules or as circumscribed masses, with distinct contrast enhancement and low peri-lesional oedema [2].

Intra-axial granulomas can also simulate a brain tumour or lymphoma. We can refer to the systemic attacks of sarcoidosis and the regression of the mass under corticoid. However, neurosarcoidosis may not regress under corticoid, and lesions such as lymphoma and histiocytosis may regress under this treatment [7, 8].

White matter abnormalities are not specific for the disease, bat often described during neurosarcoidosis (30% of cases), as the cerebellar white matter signal abnormalities found in our patient. Lesions reported in the literature are frequently periventricular. They are hyperintense on T2-weighed images, and without enhancement after gadolinium injection. The white matter lesions do not respond to treatment and are irreversible [2, 7].

Spinal cord sarcoidosis generates a smooth or nodular leptomeningeal enhancement after Gadolinium, with irregular peripheral cord thickening. Fusiform cord enlargement is usually found, and it is replaced in later stages by the atrophy of the spinal cord [9, 10].

The most important differential diagnosis for cerebral white matter involvement and marrow involvement is multiple sclerosis, according to MRI appearance, topography, and variable clinical manifestations [2]. We note the absence of abnormality of the spinal cord in our patient.

Differential diagnosis	specifications
Meningioma	Meningeal thickening that is enhanced by gadolinium in contact with the tumor.
Tuberculous meningitis	Clinical signs of tuberculosis infection, Genexpert MTB is positive.
Brain tumor	Prominent peri-lesional oedema and absence of inflammatory involvement of other organs specific to sarcoidosis.
Multiple sclerosis	Absence of systemic sarcoidosis signs.

Table 1: Principal differential diagnoses of neurosarcoidosis.

In almost 50% of cases, the neurologic symptoms represent the first manifestation of sarcoidosis [3]. So the diagnosis process consists in knowing if other organs are subclinically involved, if its involvement is spontaneously resolved, or it is an isolated neurosarcoidosis. At the same time, it is keeping in mind that the vital principle is the exclusion of infection and malignant causes before settling on the diagnosis of neurosarcoidosis [3, 11].

Thus, tissue biopsy of accessible non-neural sites is useful to establish a definite diagnosis of the disease.

Treatment of sarcoidosis is based on corticoid, and in the second-line on immunosuppressive medication. It is required appropriately to affected organs and the severity of clinical manifestations. MRI is used to monitor the effectiveness of treatment and to differentiate between stable and reversible lesions. Contrary to irreversible white matter abnormalities, meningeal and cranial nerve lesions have usually a favourable response to treatment, we note that radiological regression is often delayed compared to clinical improvement [4, 8].

CONCLUSION:

Multiple differential diagnoses of neurosarcoidosis are often discussed mainly in the case of inaugural involvement. Sarcoidosis must be proved based on the clinical and biological presentation, imaging and histology finding, and response to treatment.

MRI has a central place in the diagnostic strategies of neurosarcoidosis; it strengthens diagnostic arguments, detects subclinical lesions, and control treatment efficiency.

CONFLICTS OF INTEREST: None.

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