Pseudotumoral Neurosarcoidosis

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Abstract

Sarcoidosis is a systemic granulomatosis characterized by predominantly mediastinopulmonary involvement. Neurological involvement is reported in 5-16% of cases with predominant involvement of the central nervous system. The pseudotumoral form is only reported in 10% of neurological cases and poses a real diagnostic problem, as is the case with our present observation. Observation: 77-year-old patient hospitalized for treatment of right hemiparesis with aphasia. In his ATCDs, we retain a follow-up for an undetermined psychiatric condition. The onset of the disease dates back 5 months after his hospitalization with the sudden onset of right hemiplegia accompanied by generalized tonicoclonic convulsive seizures, with secondary recovery of the motor deficit and the onset of extra-pyramidal syndrome. Imaging finds the presence of several cerebral lesions processes and at the thoracic level, nodular lesions especially of the 2 upper lobes without mediastinal lymphadenopathy. The laboratory results were normal apart from an inflammatory syndrome and lymphopenia. Stereotaxic biopsy showed chronic inflammatory changes with ischemic necrosis without tumor lesions. Discussion: Pseudotumoral neurosarcoidosis is rare and difficult to diagnose, especially in the absence of characteristic extra neurological involvement. The histological study can find 2 aspects: classic granuloma or rarely lesions of ischemic necrosis. Treatment is based on glucocorticoids and in case of resistance, intolerance or dependence, immunosuppressants (methotrexate, cyclophosphamide).

Keywords: Pseudotumoral Neurosarcoidosis Sarcoidosis granulomatosis.

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INTRODUCTION

Sarcoidosis is a systemic granulomatosis characterized by primarily mediastino-pulmonary involvement. Neurological involvement is reported in 5% to 15% of cases with predominant involvement of the central nervous system [4]. The pseudotumoral form is only reported in 10% of neurological cases and poses a real diagnostic problem, as is the case with our present observation.

OBSERVATION

77-year-old patient hospitalized for treatment of right hemiparesis with aphasia. In his antecedent, we retain a follow-up for a psychiatric condition not determined for some months. There is no cardiovascular risk factor apart from age and sex, in particular there is no high blood pressure, no diabetes, no passive or active smoking, the patient is not obese and there is no dyslipidemia.

The onset of symptoms goes back 5 months after his hospitalization with the sudden onset of right hemiplegia accompanied by generalized tonic-clonic convulsive seizures, progressing in a context of apyrexia. The course is marked by a secondary recovery of the motor deficit and the appearance of an extra-pyramidal syndrome: parkinsonian syndrome. The cerebral morphological explorations (CT and MRI) showed the presence of several lesion processes, including two large and adjacent in the left frontal para sagittal and at least one other lesion in the left subcortical parietal of a smaller size. These lesions do not cause a mass effect, especially on the adjacent scythe of the brain, only discreetly pushing back the frontal horn left and are accompanied by a range of peri-lesional edema, especially in the left semi-oval center.

Chest imaging: chest x-ray and chest CT; observed multiple nodular lesions predominantly in the 2 upper lobes with subpleural nodules and parenchymal condensation of the posterior segment of the right upper lobe, without mediastinal lymphadenopathy.

After carrying out a physiological assessment (search for Bacillus of Koch, intradermoeaction with tuberculin), search for tumor lesions (thoraco-abdomino-pelvic CT) which turned out to be negative; stereotaxic biopsy was performed with a...
A negative bacteriological study. Pathological analysis showed chronic inflammatory changes with ischemic necrosis, without tumor lesion.

The laboratory results were normal apart from an inflammatory syndrome and lymphopenia. Angiotensin converting enzyme and prostate specific antigen levels are normal.

**DISCUSSION**

Sarcoidosis is a granulomatosis affecting several organs of unknown etiology, characterized by a process that alters cellular immunity with an increase in these alterations in affected organs.

Neurological involvement is observed in 5 to 15% of cases. Single or multiple pseudotumor forms are rare and sometimes asymptomatic, so revealed by brain imaging.

Dumas et al. [3] have shown the value of MRI on the therapeutic and evolutionary level. The lesions which take on the contrast correspond to active inflammatory lesions, which can regress on corticosteroids, unlike the lesions appearing in hypersignal on the T2-weighted sequences and which d CNS involvement is secondary to the extension of the granulomatous process involving the leptomeninges [2]. The deep and nodular lesions in cerebral imaging would indicate demyelination, while those under the diffuse and micronodular cortices would be in favor of arteriolar damage.

The diagnosis is easy when the minimum criteria are met: presence of a granuloma with epitheloid cells and gigantocellular cells, non-caseifying; presence of overt clinical manifestations; absence of intervention of pathogens likely to cause granulomatous lesions. Elsewhere, it is a bundle of arguments that will allow us to retain the diagnosis.

The histological study can find 2 aspects: classic granuloma or rarely lesions of ischemic necrosis [6]. Pseudotumoral forms are asymptomatic in 75% of cases [1]. The clinical picture is not stereotypical and depends on the location of the granulomas. Lesions are more frequently supratentorial and may be single or multiple.o not take gadolinium. Treatment with corticosteroids is sometimes reported to be extremely effective [5].
CONCLUSION

Pseudotumoral neurosarcoidosis is rare and difficult to diagnose, especially in the absence of characteristic extra neurological involvement. The treatment is based on glucocorticoids and in case of resistance, intolerance or dependence, immunosuppressants (methotrexate, cyclophosphamide).

REFERENCES