ISOLATED NEUROSARCOIDOSIS: A CASE REPORT

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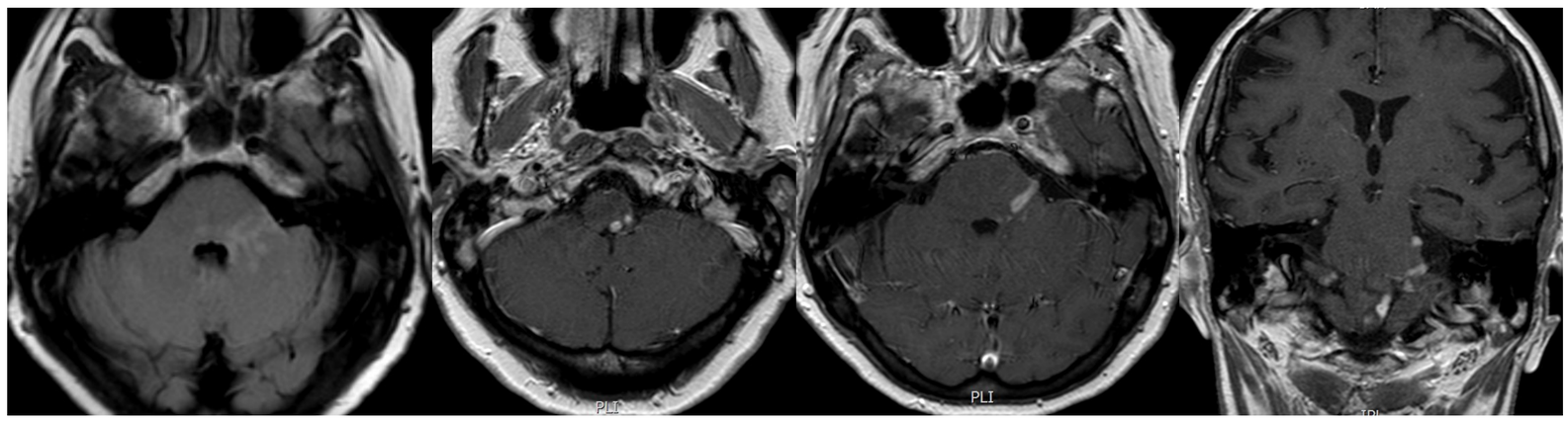
Introduction

Sarcoidosis is a multisystemic granulomatous disorder of unknown cause that leads to neurologic complications in 5% to 10% of cases. In more than half of these patients neurological symptoms are the primary presentation. The diagnosis is difficult due to the heterogeneous clinical presentation and low sensitivity of ancillary investigations.

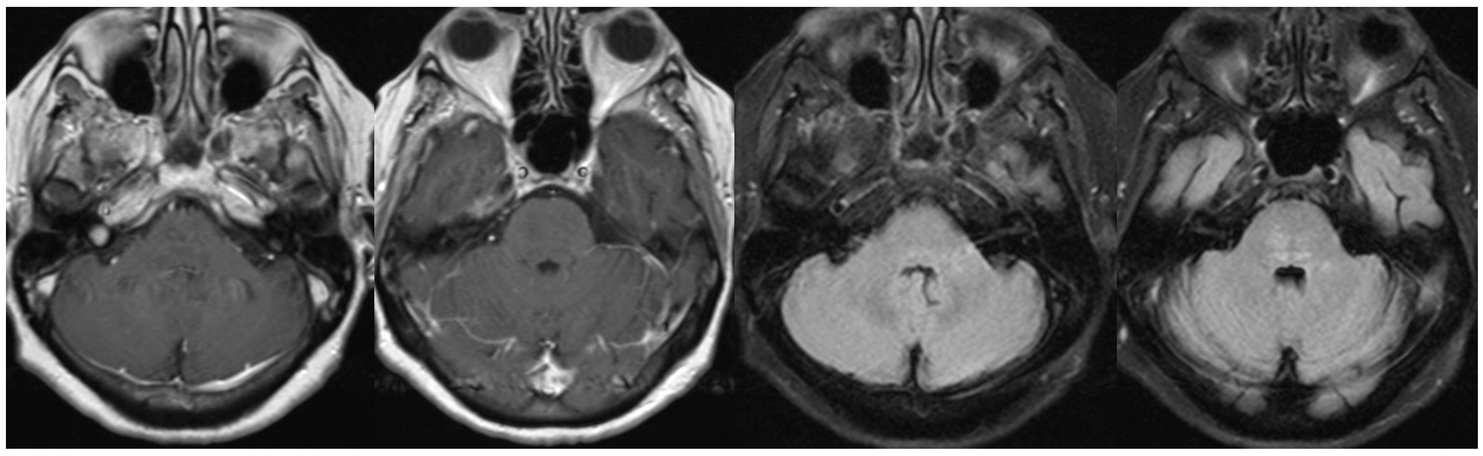
Case report

A 70 years old woman presented to the emergency room for left earache and left perioral paresthesias, developed in the morning. She had a 10-days history of dizziness and vomit accompanied by postural instability. Neurological examination showed nystagmus in all positions of gaze and left lateropulsion of the body. No other clinical signs were observed.



MRI of the brain showed hyperintensity on T2 weighted and FLAIR sequences in the left part of medulla and in the left middle cerebellar peduncle. This lesions showed gadolinium enhancement involving also the first part of V, VII and VIII left cranial nerves.

The CSF exam showed a mild augmentation of proteins and cells (for the most part monocytes). Laboratory tests and imaging studies excluded infectious, autoimmune or tumoral diseases. No signs of systemic disease were found. Serum level of ACE was normal. No changes were observed after a ten days therapy with Ampicillin and Acyclovir. A dramatic improvement of clinical examination and MRI imaging was achieved after corticosteroid therapy.



MRI of the brain showed a reduction of hyperintensity on T2 weighted and FLAIR sequences in the left part of medulla and in the left middle cerebellar peduncle associated to reduction of the gadolinium enahncement.

After ten months of steroid therapy our patient is paucisymptomatic and there aren't any parenchymal abnormalities on brain MRI .

Conclusions

Clinical presentation, exclusion of other diseases and response to corticosteroids, suggest the diagnosis of possible isolated neurosarcoidosis.
Following the diagnostic criteria, a definite diagnosis would require a biopsy of the nervous system, a procedure not always applicable, as in our case, given the deep localization of the lesions. Early recognition of neurosarcoidosis and aggressive treatment play a key role in halting the progression of the disease and, in some cases, they help to restore neurological function.

Box 1
Diagnostic criteria for neurosarcoidosis

In patients with a clinical presentation suggestive of neurosarcoidosis and exclusion of other diagnoses, classification is as follows:

Definite

- Histologic confirmation of affected neural tissue

Probable

- Evidence of CNS inflammation on MRI or CSF (elevated protein, cells, IgG index, or presence of oligoclonal bands) compatible with neurosarcoidosis AND
- Evidence of systemic sarcoidosis with histologic confirmation and/or at least two of the following indirect indicators: fluorodeoxyglucose PET, gallium scan, chest imaging, serum angiotensin-converting enzyme

Possible

- Above criteria not met

Adapted from Zajicek JP, Scolding NJ, Foster O, et al. Central nervous system sarcoidosis-diagnosis and management. QJM 1999;92:104.