A CASE REPORT

SPONTANEOUS INTRACRANIAL HYPOTENSION OR NEUROSARCOIDOSIS: THAT IS THE QUESTION!

AUTHORS: I.A. Di Vico[1], A. Novelli[1], A. Picchioni[1], G. Carlucci[1], L. Massacesi[1]
[1]MD Università degli Studi di Firenze, Dipartimento NEUROFARBA Azienda Ospedaliera Universitaria Careggi.

Background

MRI evidence of pachymeningeal enhancement is a relatively common finding and represents a diagnostic challenge for the clinician. It may arise from various processes, including iatrogenic or spontaneous intracranial hypotension (SIH), autoimmune disorders, malignancies and infections.

Case Presentation

A 70-years-old man was admitted to our Neurology Department with a 7 year history of tension fronto-occipital headache associated with tinnitus and impaired hearing. The symptoms worsened in the last few months before admission and showed only mild response to over-the-counter analgesics. The headache was described as continuous and not orthostatic. Neurological examination was normal.

A brain MRI showed bilateral, thick, linear, pachymeningeal enhancement with normal ventricular spaces, interpreted in first instance as a possible intracranial hypotension (**Figure 1**). Medical history was significant for prostatic cancer treated with radical surgery, and mild chronic cough. A thoracic CT showed some enlarged mediastinal lymph-nodes. FDG-PET showed mediastinal and hilar lymphadenopathy increased FDG uptake (**Figure 2**).

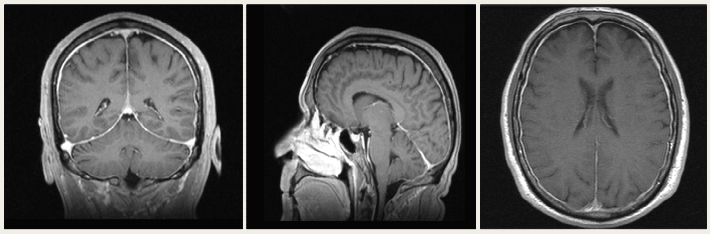

Figure 1: MR T1 sequences evidence diffuse pachymeningeal enhancement after contrast injection.

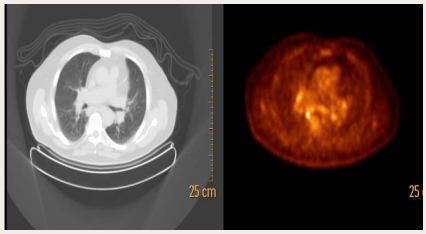

Figure 2: PET-FDG evidences hilar and mediastinal lymphadenopathy increased FDG uptake.

Routine bloodwork, including PSA, was not significant except for mild increase of erythrocyte sedimentation rate (ESR) and C-reactive protein. A lumbar puncture was performed with traumatic tap: it showed elevated protein (1.01 g/L), mild pleyocitosis (24 WBC/microL), negative serological tests for infections and a normal opening pressure (120 mmH_2O). Further investigations were significant for ANA 1:320 (pattern NuMA), Angiotensin-converting enzyme in the upper normal limit and significantly elevated chitotriosidase levels (436 nmol/mL/h).

The patient was treated with intravenous fluids and 7 days of iv corticosteroids (Methylprednisolone 20 mg/die). At the discharge he reported a significant clinical improvement.

Discussion

The initial hypothesis of SIH was not clearly supported by the headache features and by the normal CSF pressure. Significantly elevated chitotriosidase levels, even though not included in the diagnostic criteria, has been reported to have an high sensitivity and specificity (88.6% and 92.8%, respectively) as a diagnostic biomarker of sarcoidosis. Pachymeningeal involvement is a common finding in neurosarcoidosis. Overall, the evidence of high levels of chitotriosidase in association with CT finding of enlarged mediastinal lymph-nodes and the optimal clinical response to steroid therapy are highly suggestive of sarcoidosis with neurological involvement, though we still need the histological confirmation to make a definite diagnosis.

References
Bargagli E. Human chitotriosidase: a sensitive biomarker of sarcoidosis. J Clin Immunol. 2013.
Antony J. Pachymeningeal enhancement-a comprehensive review of literature. Neurosurg Rev. 2015.
Brouwer MC. Clinical features, treatment and outcome in neurosarcoidosis: systematic review and meta-analysis. BMC Neurol. 2016.

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