Neurological picture. Sarcoidosis presenting with hydrocephalus

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Neurological picture

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CASE REPORTS

Case No 1

A 72-year-old Afro-American woman, also born in Surinam, without a previous medical history, presented with a 1 year history of progressive gait disorder, urinary incontinence and short term memory loss. Neurological examination showed bradyphrenia and gait ataxia. Head MRI showed communicating hydrocephalus (fig 1C). Lumbar puncture showed an opening pressure of 170 mm H₂O. 215 CSF leucocytes/ml (90% lymphocytes) and CSF protein of 0.92 g/l. Again, serological testing and culturing were all negative.

Chest x ray, serum levels of angiotensin converting enzyme (ACE) and lysozyme were normal. ¹³FDG-PET showed para-oral and paranguiual lymphadenopathy and histopathology was consistent with sarcoidosis (fig 1E). She recovered on prednisolone treatment.

COMMENT

Our cases show that sarcoidosis should be actively sought for in patients presenting with hydrocephalus and pleiocytosis, and also in those without a known systemic sarcoidosis and normal chest x ray and serum levels of ACE and lysozyme. Hydrocephalus has been described in 5–7% of patients with neurosarcoidosis, but rarely as the presenting symptom.¹⁴ All patients with hydrocephalus as the presenting symptom of neurosarcoidosis in the literature had a previous medical history of systemic sarcoidosis.¹⁴

Diagnosing neurosarcoidosis can be challenging, and diagnostic criteria have been proposed.¹ Using these criteria, the diagnosis of probable neurosarcoidosis can be made with a clinical presentation compatible with neurosarcoidosis, exclusion of other possible causes and positive histology. Positive nervous system histology is required to diagnose definitive neurosarcoidosis. Our patients met the criteria for probable diagnosis of neurosarcoidosis with a clinical presentation compatible with neurosarcoidosis, exclusion of other possible causes and positive histology.⁴

A retrospective cohort study described 68 patients with definite or probable neurosarcoidosis.¹ Five patients (7%) had hydrocephalus. CSF examinations were performed in 62 patients, and 34 (55%) had raised CSF white cell counts, ranging from 200 to 700/ml. In this case series, chest x ray was abnormal in 21 of 68 patients (21%) and serum ACE levels were abnormal in 12 of 51 patients (24%). The diagnostic accuracy of ¹³FDG-PET scanning in the diagnosis is unknown but in our experience it can be helpful in the detection of lymphadenopathy and so facilitate diagnostic biopsy.
Figure 1 Case No 1. Axial (A) fluid attenuated inversion recovery weighed MRI shows focal cystic dilation of the occipital horn of the right lateral ventricle and transependymal CSF effusion. \(^{18F}\)-fluorodeoxyglucose positron emission tomography (B) shows enhancing lymph nodes supraclavicular, mediastinal, in the liver hilus, para-aortal and paraaortic.

Case No 2. Axial (C) T2 weighted MRI showing hydrocephalus with transependymal CSF effusion. Granulomatous lymphadenitis from fine needle aspirate in case No 1 (D, Giemsa stain; 135 ×; 1 cm = 74 \(\mu\)m) and case No 2 (E, Giemsa stain; 270 ×; 1 cm = 37 \(\mu\)m).

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