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Neurological picture

Sarcoidosis presenting with hydrocephalus

CASE REPORTS

Case No 1

A 33-year-old Afro-American man, born in Surinam, without a previous medical history, presented with a 3 week history of headache, nausea and walking difficulties. Neurological examination showed bradyphrenia and left-sided hemiparesis. Head MRI showed a focal dilated right lateral ventricle caused by a cystic lesion (fig 1A). CSF revealed a leucocyte count of 681/ml (87% lymphocytes) and a protein level of 1.04 g/l. CSF cultures and serological tests for *Taenia solium*, *Treponema pallidum*, *Echinococcus* and *Borrelia burgdorferi* species were negative; cultures and PCR were negative for *Mycobacterium tuberculosis*.

Chest x ray, serum levels of angiotensin converting enzyme (ACE) and lysozyme were normal. Nevertheless, ¹⁸F-fluorodeoxyglucose-positron emission tomography (¹⁸FDG-PET) scan showed lymphadenopathy in the chest, abdomen and neck (fig 1B). Histopathology of needle aspiration of the lymph nodes showed non-caseating granulomas consistent with sarcoidosis (fig 1D). Ventriculocopy assisted fenestration of the lateral ventricle cyst was performed and he made a full recovery without corticosteroids.

Case No 2

A 72-year-old Afro-American woman, also born in Surinam, with a history of cholecystectomy, 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 ACE and lysozyme were normal. ¹⁸FDG-PET showed para-aortal and parainguinal lymphadenopathy and histopathology was consistent with sarcoidosis (fig 1E). She recovered on prednisolone treatment.

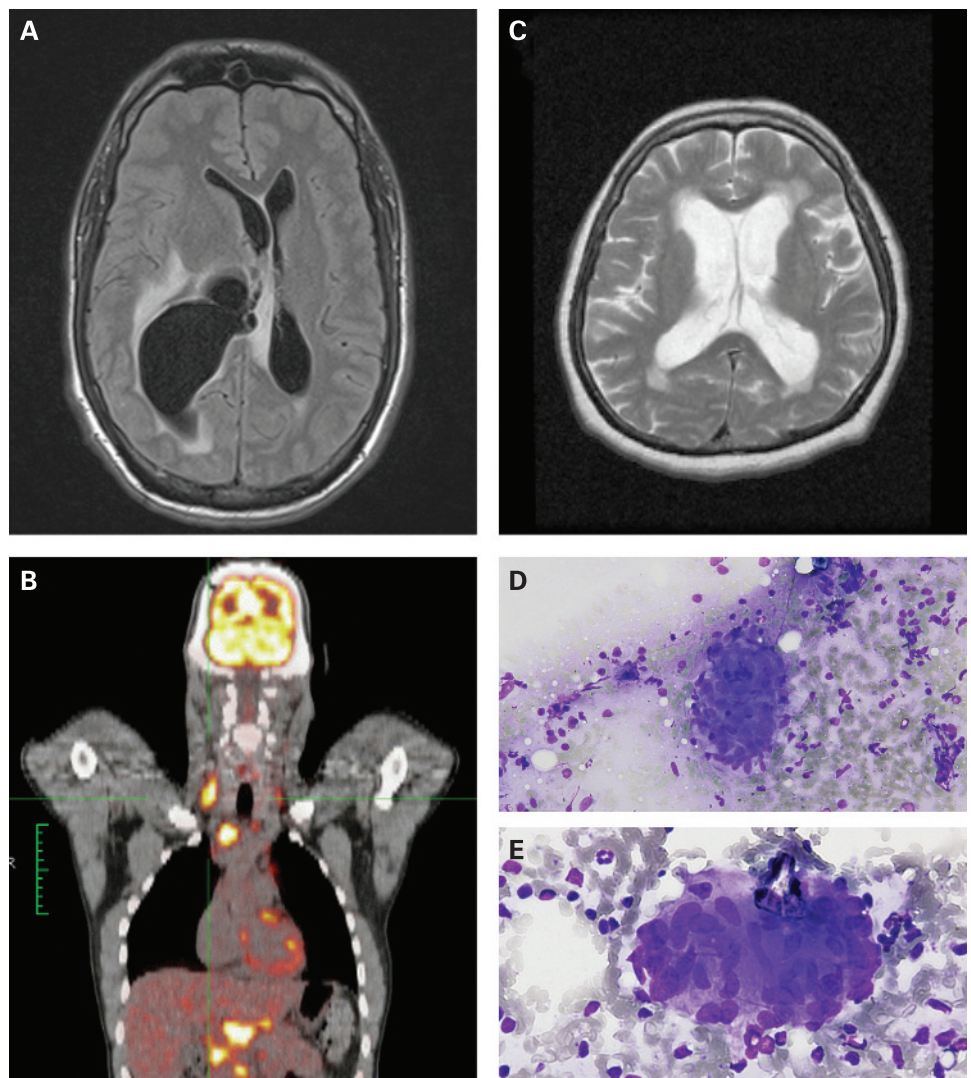
COMMENT

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.^{1–4} All patients with hydrocephalus as the presenting symptom of neurosarcoidosis in the literature had a previous medical history of systemic sarcoidosis.^{1–4}

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 ¹⁸FDG-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 parailiacal. 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 \times ; 1 cm = 74 μ m) and case No 2 (E, Giemsa stain; 270 \times ; 1 cm = 37 μ m).



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