Neurological picture. Sarcoidosis presenting with hydrocephalus
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Published in:
Journal of Neurology, Neurosurgery and Psychiatry

DOI:
10.1136/jnnp.2008.163725

Citation for published version (APA):
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J Neurol Neurosurg Psychiatry 2009 80: 550-551
doi: 10.1136/jnnp.2008.163725

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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 *Mycobacterium tuberculosis* were all negative. (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 paraganglionic lymphadenopathy and histopathology was consistent with sarcoidosis (fig 1B). 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. 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. 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. Five patients (7%) had hydrocephalus. CSF examinations were performed in 62 patients, and 54 (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.

Neurological picture

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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 ×; 1 cm = 74 μm) and case No 2 (E, Giemsa stain; 270 ×; 1 cm = 37 μm).

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Acknowledgements: We thank Dr E Aronica for her help with the pathology images.

Funding: DvdB is supported by a personal grant from the Netherlands Organisation for Health Research and Development (ZonMw): NWO-Veni grant 2006 (916.76.023).

Patient consent: Obtained.

Competing interests: None.


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