Neurosarcoidosis which Manifested as Acute Hydrocephalus: Diagnosis and Treatment

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Abstract

Neurosarcoidosis is generally expressed by various symptoms; it is exceptionally revealed by hydrocephalus. We report a 27-year-old African man hospitalized with acute neurological deterioration due to hydrocephalus. The treatment consisted of ventriculostomy and corticosteroid therapy; however a permanent shunt was not required. After six months of follow-up, the patient had fully recovered. Clinical presentation, neuroimaging findings and treatment modalities are discussed.

Key words: hydrocephalus, neurosarcoidosis, diagnosis, MRI, shunt, ventriculostomy

(DOI: 10.2169/internalmedicine.46.0126)

Introduction

Sarcoidosis is a multisystemic disease of unknown origin; it is characterized by an accumulation of non-caseating epithelioid granulomas (1). The lung, skin, eyes and lymph nodes are the most commonly involved organs. Only 5% of sarcoidosis patients show neurological manifestations (2-4), evermore hydrocephalus is uncommonly reported as a presenting manifestation of Neurosarcoidosis. Therefore, the goal of this paper is to report a confusing case of neurosarcoidosis revealed by acute hydrocephalus. The patient was successfully managed by immediate ventriculostomy and long-term corticosteroid therapy.

Case Report

A 27-year-old man of African origin (from Mali) who had no relevant medical history was admitted to the Neurosurgical Emergency Department of the Pitié-Salpêtrière Hospital with acute neurological status. This was expressed in terms of coma, reactive bilateral mydriasis and neurovegetative dysautonomy with associated bradycardia at 54 pulses/min, febricula at 38.5°C and blood pressure of 110/60 cmHg.

CT scan showed acute hydrocephalus with a meningeal contrast-enhancement localized in the posterior cerebral fossa. It was associated with a felted cerebral base. An external ventricular shunt was performed (Fig. 1). Cerebrospi-
Figure 2. Magnetic resonance imaging of the central nervous system, in post-gadolinium axial T1-weighted image (a) showing granulomatous infiltration along the Virchow Robin spaces and in (b) an extensive basal leptomeningeal infiltration, in (c) a post-gadolinium sagittal T1-weighted image showing extra-axial contrast enhancement along the base of the brain, posterior fossa and cervical canal.

Figure 3. Epitheloid follicles composed of macrophages, lymphocytes and fibroblasts; it is well delineated at periphery by a fibrous shell.

Cerebrospinal fluid (CSF) pressure was high and was evaluated at 22 mmHg at the per-surgery stage; upon analysis, an increased total protein rate at 256 mg/dL, hypoglycorrhachia at 27 mg/dL with concomitant glycemia at 98 mg/dL, and predominantly mononuclear pleocytosis at 300 leukocytes/mm$^3$ were found. The bacteriological examination was negative. Complementary contrast-enhanced MRI was performed; this showed leptomeningeal nodular infiltration predominating the brain base, tentorium and cervical canal with infiltration of parenchyma along the Virchow-Robin spaces (Fig. 2).

Intravenous bolus methyl prednisolone was then started at the rate of 20 mg/kg/day for consecutive 3 days; this was followed by high-dose of prednisone (1 mg/kg/day) for a duration of six weeks. Empiric antibacillar therapy was also initiated; it consisted of rifampicin (10 mg/kg/day), isoniazid (5 mg/kg/day), streptomycin (1 mg/kg/day) and pyrazinamide (30 mg/kg/day).
Figure 4. CT scan of control at the fourth month showing a complete regression of the hydrocephalus without vascular complication. The subarachnoid space is clearly visible.

At this stage, the general physical examination revealed bilateral cervical and submandibular lymphadenopathy of 1 cm diameter. Excisional biopsy of the left lateral cervical adenopathy showed non caseating epitheloid granuloma (Fig. 3). Regarding the acid-fast bacilli and fungal infection, the culture was negative. Then, the antibacillar chemotherapy was stopped by the second week after hospitalization and the patient was kept only under corticosteroid therapy. Angiotensin-converting enzyme was significantly elevated (150 U/L for normal value <40 U/L). The chest radiograph was normal.

The clinical course was favorable. A few days after the operative procedure, the patient opened his eyes to tact stimulus. Then, the patient showed progressive improvement of consciousness without papilloedema or focal deficits. After one month of hospitalization in the intensive care unit, the patient was transferred to the rehabilitation hospital where he was kept for four months. Afterwards, he was discharged and returned to his home.

Serial CT scan of control was performed and revealed an obvious decrease of the ventricular size well correlated with the clinical improvement (Fig. 4). According to his clinical status, the prednisone dose was progressively decreased after four weeks. Finally, the patient has an independent functional status after one year of clinical follow-up.

Discussion

Neurological manifestations of sarcoidosis are observed in approximately 5% of patients (2-4); these mainly consist of cranial and peripheral nerve lesions, with the most frequent manifestation being seventh nerve palsy (4). Other manifestations include optic neuritis, aseptic meningitis, hearing abnormalities, headache, vertigo, hypothalamic and pituitary dysfunction, mass-lesion effect, myopathy, seizure and psychiatric symptoms (3, 5). Hydrocephalus is a very uncommon finding. It results in fourth ventricular outflow obstruction or reduced absorption of the cerebrospinal fluid (6, 7). To date, there is only one reported case of hydrocephalus occurring in neurosarcoidosis as the first symptom (8).

Classically, most patients have systemic sarcoidosis at the onset of neurological symptoms. In contrast, whenever neurological involvement is the first or only manifestation of sarcoidosis, the diagnosis becomes very difficult; it may be done by clinical exclusion (4). Neurosarcoidosis may mimic other diseases such as tuberculosis, multiple sclerosis, spinal cord abnormalities, Sjögren’s syndrome, neurophilis, neureroborrellosis, Behçet’s disease, Vogt-Koyanagi-Harada disease, lymphoma, anaplastic astrocytoma and peripheral neuropathy (9). However, the granulomatous character of the disease can be confirmed by tissue sampling; brain biopsies are sometimes required (4, 9).

MRI has been reported as the preferred imaging technique to evaluate neurosarcoidosis (10-13). T1-weighted images accurately depict hydrocephalus and spinal cord enlargement while T2-weighted and FLAIR slices exhibit increased signal intensity, especially in a periventricular distribution. Contrast administration can demonstrate leptomeningeal enhancement as well as parenchymal abnormalities and, occasionally cranial nerve lesions (14). Thus, MRI can prove positive in 65% of patients, but no neurodiagnostic tests alone are pathognomonic for neurosarcoidosis (15). Indeed, Lower et al (5) established the following criteria for the diagnosis of neurosarcoidosis: 1) non caseating granulomas identified in pathologic examination, 2) specific MRI findings of the nervous system combined with increased lymphocyte and protein levels in CSF, and 3) when patients with sarcoidosis develop otherwise unexplained diabetes insipidus or facial palsy.

Treatment modalities are based on the clinical course of the patient (8). Surgery is indicated once intracranial pressure is increased, for removing brain tumor or to establish tissue diagnosis. In patients with hydrocephalus, management depends upon the clinical status. Asymptomatic ventricular enlargement probably does not require treatment (9). Nevertheless, ventricular drain is usually lifesaving in symptomatic hydrocephalus (8). This was done in the present case according to the clinical status and CT scan findings. However, it is noteworthy that the derivation system was removed three weeks after surgery. Considering that shunt obstruction is frequent during chronic follow-up (3, 9, 16, 17), we decided to supervise the patient without inserting an immediate definitive ventriculoperitoneal shunt. This decision was also justified by the favorable status of the patient.

Although there has been no controlled trial of medical treatment for neurosarcoidosis, corticosteroid therapy re-
mains the treatment of choice (8). In the case of failure, immunosuppressive agents and radiation therapy can be used (16). The goal for this kind of treatment is to diminish the irreversible fibrosis that can develop as well as the tissue ischemia that might result from perivascular inflammation (9).

In such cases of neurosarcoidosis, the outcome varies according to the neurological presentation (13). Overall, the prognosis for neurosarcoidosis is good (8); 90% of the neurologic manifestations remit or improve with time, nevertheless, hydrocephalus has the worst long-term prognosis with a mortality rate of 75% (8, 13). In the present case, after two years of recession, hydrocephalus did not recidivate and the patient continues his routine life with good functional performance.

**Conclusion**

Sarcoidosis with neurological involvement is a significant cause of morbidity and mortality. Neurosarcoidosis is generally expressed by a wide range of symptoms, but it is exceptionally revealed by hydrocephalus. The diagnosis of this pathology is very difficult; however the management of the disease is straightforward. Successful management of hydrocephalus is possible using ventriculostomy and chronic systemic steroids; it does not systematically require a permanent shunt.

**Acknowledgement**

The authors sincerely thank Dr. M. KUJAS for providing histological images.

**References**


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