

[CASE REPORT]

Neurosarcoidosis Presenting with Obstructive Hydrocephalus Successfully Treated with Endoscopic Third Ventriculostomy

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Abstract:

A 58-year-old Japanese woman complained of unstable gait and dizziness lasting for a month. She had been diagnosed histologically with pulmonary and cutaneous sarcoidosis and attended outpatient clinics for routine checkups. Head computed tomography and magnetic resonance imaging (MRI) indicated obstructive hydrocephalus caused by a contrast-enhanced lesion in the cerebral aqueduct. The patient underwent endoscopic third ventriculostomy and a biopsy of the lesion, leading to the diagnosis of neurosarcoidosis. This was a rare case of neurosarcoidosis presenting with obstructive hydrocephalus that was treated with endoscopic third ventriculostomy and diagnosed histologically via an intraoperative biopsy.

Key words: endoscopic third ventriculostomy, neurosarcoidosis, obstructive hydrocephalus, sarcoidosis

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Introduction

Sarcoidosis predominantly involves the lungs, anterior uvea, lymph nodes, and skin. Neurosarcoidosis is seen in 5% of systemic sarcoidosis patients, and a prospective study showed that hydrocephalus occurred in 6-7% of cases (1, 2). The diagnosis of neurosarcoidosis can be challenging because a biopsy of the central nervous system is difficult and highly invasive. Thus, most cases of neurosarcoidosis are diagnosed by imaging findings, such as leptomeningeal involvement, meningeal enhancement, and/or multiple white-matter lesions in patients with known sarcoidosis.

Neurosarcoidosis patients with obstructive hydrocephalus are treated with either ventriculoperitoneal (VP) shunt, steroid therapy, or both (3-5). However, this tends to cause infection by artifacts and steroid use. Obstructive hydrocephalus caused by neurosarcoidosis has rarely been treated with endoscopic third ventriculostomy (ETV).

We herein report a case of neurosarcoidosis with obstructive hydrocephalus that was diagnosed via an intraoperative biopsy and treated with ETV.

Case Report

A 58-year-old Japanese woman born and living in Shizuoka Prefecture presented with a 1-month history of unable gait and dizziness. She had been diagnosed via a biopsy four years earlier with pulmonary and cutaneous sarcoidosis at our hospital and had been given a topical steroid for skin lesion. She was alert, and her vital signs were within normal limits. A neuroexamination revealed gait disturbance due to frontal ataxia. Her routine biochemistry findings were within normal ranges. Although the level of sIL-2R was elevated at 597 U/mL, the angiotensin-converting enzyme (ACE) level was not elevated, and the rest of the laboratory data were unremarkable.

Chest X-ray showed hilar lymphadenopathy. Computed tomography (CT) showed parenchymal nodules in the left lobes and mediastinal lymphadenopathy that were larger than they had been at a previous checkup. Magnetic resonance imaging (MRI) of the head revealed remarkable dilatation of the lateral and third ventricles, and the cerebral aqueduct showed periventricular hyperintensities on fluid-

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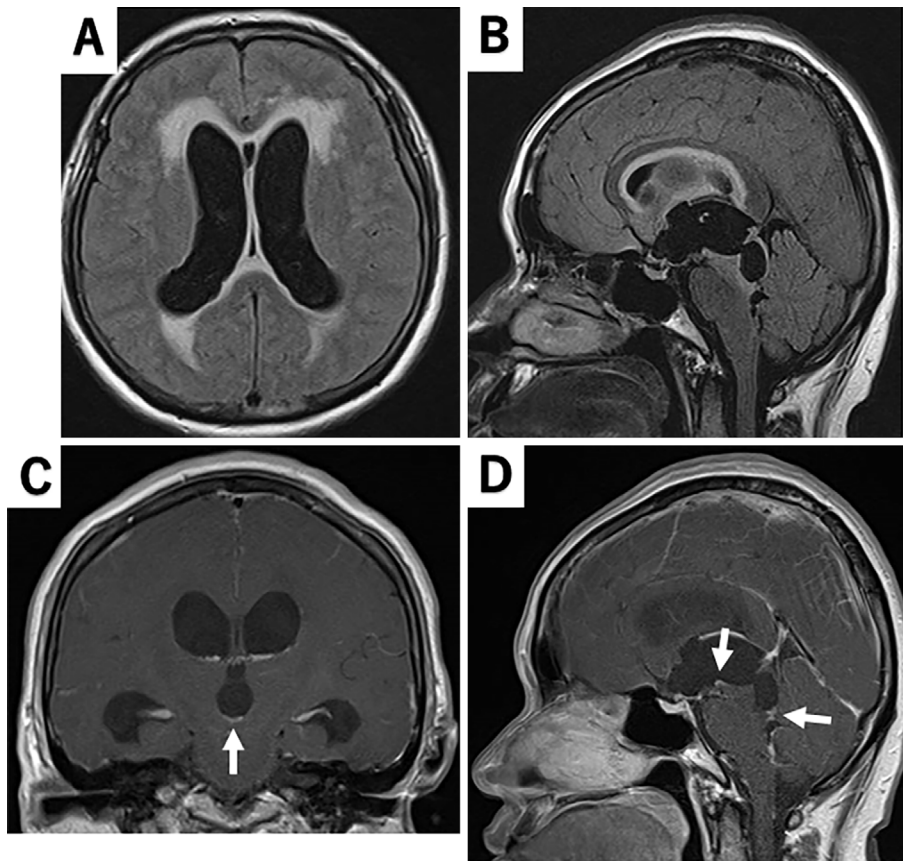


Figure 1. Magnetic resonance imaging (MRI) revealing remarkable dilatation of the lateral and third ventricles with periventricular hyperintensities on fluid-attenuated inversion recovery (FLAIR) imaging (A, B). No dilatation of the fourth ventricle (B). Gadolinium-enhanced T1-weighted imaging showing high-intensity foci at the floor of the third ventricle and cerebral aqueduct (C, D: arrows).

attenuated inversion recovery (FLAIR) imaging (Fig. 1A and B). The fourth ventricle showed no enlargement (Fig. 1B). Gadolinium-enhanced T1-weighted imaging showed high-intensity foci at the floor of the third ventricle and the cerebral aqueduct (Fig. 1C and D). These findings indicated non-communicating hydrocephalus caused by a contrast-enhanced lesion in the cerebral aqueduct and blockage of the cerebrospinal fluid (CSF) circulating system around the cerebral aqueduct, although they did not reveal the etiology.

To relieve subacute symptoms as well as to obtain a histological diagnosis, the patient underwent ETV, a procedure for obstructive hydrocephalus in which an opening is created in the floor of the third ventricle using an endoscope placed within the ventricular system through a burr hole (Fig. 2). In this operation, we observed brown granular lesions at the cerebral aqueduct (Fig. 2A) and the third ventricle floor (Fig. 2B) and performed a biopsy of the third ventricular lesion.

The cytology of the lesion was negative for neoplastic cells. Pathological findings revealed noncaseating epithelioid granulomas (Fig. 3). On an examination of the CSF collected during this operation, the total protein and glucose levels were normal at 15 and 73 mg/dL, respectively. Predominantly mononuclear cell pleocytosis was found, but

bacteriological examinations were negative. Thus, the patient was diagnosed histologically with neurosarcoidosis presenting with obstructive hydrocephalus.

The clinical course was favorable. A few days after the operation, she was able to walk straight but slowly and was discharged from the hospital 10 days after the operation without any symptoms. She received no treatment, including corticosteroids or immunosuppressive medication for neurosarcoidosis, because her symptoms improved dramatically after the operation. She was followed up at the outpatient clinics for two years without any recurrence.

Discussion

Neurosarcoidosis is suspected in sarcoidosis patients who complain of neurological symptoms. The diagnosis is sometimes based on the clinical history, symptoms, and imaging findings, since histopathological confirmation is often challenging because of the difficulty in performing a biopsy. In the present case, an endoscopic procedure was useful for relieving symptoms and obtaining a diagnosis of neurosarcoidosis presenting with obstructive hydrocephalus.

Hydrocephalus is a rare complication in neurosarcoidosis and has a poor prognosis (6). In general, the diagnosis of hydrocephalus caused by neurosarcoidosis is difficult. In

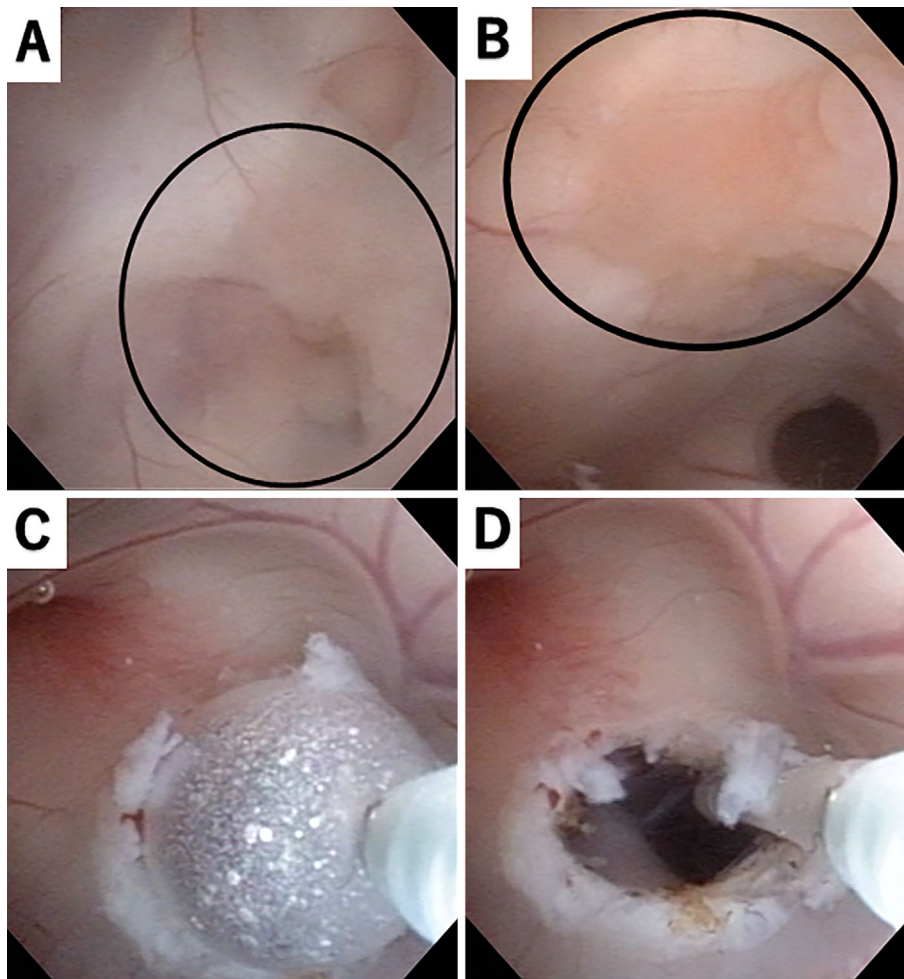


Figure 2. Neuroendoscopic findings showing an occluded portion of the cerebral aqueduct with a brown granular lesion (A, circle) and the biopsied lesion of the same nature at the floor of the third ventricle (B, circle). Endoscopic third ventriculostomy showing enlargement of the fenestrated area by a Fogarty catheter (C, during and D, after procedure).

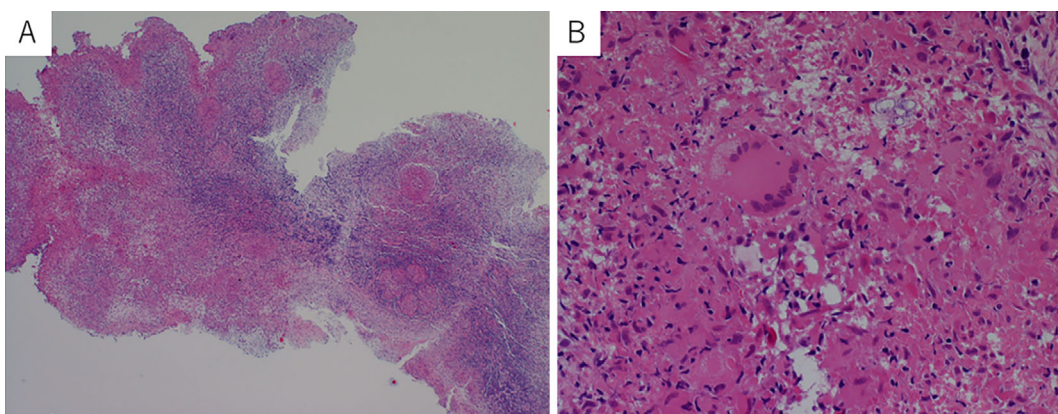


Figure 3. Histopathological findings of the intraoperative biopsied specimen from the third ventricle showing several noncaseating granulomas composed of epithelioid cells and multinucleated giant cells (Hematoxylin and Eosin staining: A, 10× original magnification; B, 100× original magnification).

such cases, ETV may be an alternative to VP shunt. ETV has attracted substantial attention recently and is now regarded as an established treatment for non-communicating

hydrocephalus (7, 8). In ETV, the third ventricular floor is fenestrated to communicate with the ventricular system. This procedure requires only a small hole be drilled in the skull.

All patients who have an obstruction between the third ventricle and the cortical subarachnoid spaces, such as those with aqueductal stenosis, are potential candidates for this procedure (9). The complications related to this procedure include intraoperative hemorrhaging, diabetes insipidus, weight gain, and precocious puberty. Bouras et al. reported that the overall complication rate was 8.5% in young infants (10), although permanent complications were few. It is true that VP shunt is a treatment option for hydrocephalus, but ETV is useful in terms of making the histological diagnosis as well as treating obstructive hydrocephalus, leading to a shunt-free life.

While there have been some cases of hydrocephalus caused by neurosarcoidosis diagnosed by neuroendoscopy and treated by VP shunt (11), there have been no cases of obstructive hydrocephalus caused by neurosarcoidosis and diagnosed by ETV. Thus, the prognosis of such cases is unknown. The present patient received no treatment, including corticosteroids or immunosuppressive medication for neurosarcoidosis after the operation. In their study of neurosarcoidosis patients with hydrocephalus, Yoshinoya et al. reported that most patients used steroids before or after VP shunt operation because two-thirds of the patients who did not use steroids developed recurrence or exacerbation. They also reported that patients with symptomatic intracranial disease and/or hydrocephalus were at an increased risk of developing progression (11). There is a possibility that the sarcoid lesion may still remain in cerebral ventricles, such as the floor of the third ventricle in this case, and ideally, the patient should have been given steroids. Follow-up MRI should be performed regularly.

The authors state that they have no Conflict of Interest (COI).

References

1. Allen RK, Sellars RE, Sandstrom PA. A prospective study of 32 patients with neurosarcoidosis. *Sarcoidosis Vasc Diffuse Lung Dis* **20**: 118-125, 2003.
2. Scott TF, Yandora K, Valeri A, Chieffe C, Schramke C. Aggressive therapy for neurosarcoidosis: long-term follow-up 48 treated patients. *Arch Neurol* **64**: 691-696, 2007.
3. Labarca G, Ramirez R, Monsalve X, Mira-Avendano I. Dementia, gait disturbance, and urinary incontinence in a patient with pulmonary sarcoidosis. *Respirol Case Rep* **4**: e00182, 2016.
4. Tabuchi S, Uno T. Hydrocephalus with paraventricular enlargement as the primary manifestation of neurosarcoidosis: a case report. *J Med Case Rep* **7**: 20, 2013.
5. Zajicek JP, Scolding NJ, Foster O, et al. Central nervous system sarcoidosis- diagnosis and management. *QJM* **92**: 103-117, 1999.
6. Brouwer MC, de Gans J, Willemse RB, van de Beek D. Neurological picture. Sarcoidosis presenting with hydrocephalus. *J Neurosurg Psychiatry* **80**: 550-551, 2009.
7. Yadav YR, Parihar V, Pande S, Namdev H, Agarwal M. Endoscopic third ventriculostomy. *J Neurosci Rural Pract* **3**: 163-173, 2012.
8. Shim KW, Park EK, Kim DS, Choi JU. Neuroendoscopy: current and future perspectives. *J Korean Neurosurg Soc* **60**: 322-326, 2017.
9. ReKate HL. Selecting patients for endoscopic third ventriculostomy. *Neurosurg Clin N Am* **15**: 39-49, 2004.
10. Bouras T, Koch D. Mechanism of failure of endoscopic third ventriculostomy in young infants. *J Neurosurg Pediatr* **7**: 643-649, 2011.
11. Yoshinoya K, Itsukage M, Kasamatsu Y, Kadoya M, Kasamatsu Y, Yamada K. A case of neurosarcoidosis with hydrocephalus. *The Japanese Journal of Sarcoidosis and Other Granulomatous Disorder* **34**: 51-54, 2014 (in Japanese).

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