

Endoscopic biopsy for the diagnosis of neurosarcoidosis at the fourth ventricle outlet with hydrocephalus

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Abstract

Background: Fourth ventricle mass lesion in neurosarcoidosis is very rare and difficult to be diagnosed pathologically. We report a rare case of progressive hydrocephalus associated with neurosarcoidosis mass lesion located at the fourth ventricle outlet and suprasellar region.

Case Description: A 23-year-old man had a disturbance of consciousness and neck stiffness with fever. Magnetic resonance imaging revealed diffuse leptomeningeal enhancement, and the obstructive mass lesions at the outlet of the fourth ventricle. We performed an endoscopic biopsy of the enhanced lesion at the foramen Magendie, via foramen Monro and the aqueduct of the midbrain. Pathologically, the diagnosis of neurosarcoidosis was confirmed, and we started treatment with prednisolone. His neurological symptoms disappeared after ventriculo-peritoneal shunt and steroid therapy, and he was discharged without deficit 40 days after emergent admission.

Conclusion: Endoscopic procedure is less invasive and more effective for biopsy of neurosarcoidosis with hydrocephalus, even if the lesion is located at the fourth ventricle outlet.

Key Words: Endoscopic surgery, neurosarcoidosis, obstructive hydrocephalus

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INTRODUCTION

Sarcoidosis is a chronic, multisystem, and granulomatous disorder of unknown etiology, which is most commonly recognized in young adults between the ages of 20 and 40 years. It commonly affects the lungs, eyes, lymph nodes, skin, and liver, but the involvement of the central nervous system is not common, ranging from 5% to 20%.^[4,8] In neurosarcoidosis, the most frequent manifestation is the eighth nerve palsy.^[8] On the other hand, hydrocephalus is a rare manifestation, which occurs in 6% of patients with neurosarcoidosis,^[2] and a previous report showed that hydrocephalus associated with neurosarcoidosis has poor long-term prognosis with mortality rate of 75%.^[1] Some reports showed that endoscopic treatment such as ventriculostomy is

effective for the obstructed ventricles associated with neurosarcoidosis.^[5] However, endoscopic surgery for the diagnosis of neurosarcoidosis by biopsy of the lesion at the fourth ventricle has not been reported.

In this case study, we report a rare case of rapid progressive hydrocephalus due to the obstruction of the

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fourth ventricle outlet that was pathologically diagnosed as fourth ventricle sarcoidosis by endoscopic biopsy.

CASE REPORT

A 23-year-old Japanese man was transferred to our hospital because of progressive consciousness disturbances with vomiting. He had no history of any systemic diseases but had a mild fever and headache frequently in 3 weeks prior to admission. He showed abnormal behavior from 14 days before hospitalization and eventually developed disturbance of consciousness. On admission, he showed nystagmus and neck stiffness with a fever of 38.5°C, and his Glasgow Coma Scale (GCS) score was 9 [E (3) V (1) M (5)]. Brain computed tomography (CT) revealed hydrocephalus with all ventricles dilated. We could not find any mass lesions on the CT-scan images. We performed external ventricular drainage (EVD) emergently, and his consciousness gradually improved. After emergent EVD, the magnetic resonance imaging (MRI) revealed diffuse leptomeningeal enhancement, and obvious enhanced mass lesion at the inferior wall of the third ventricle extended to the suprasellar region [Figure 1a and b], and obstructive mass lesions extended to the outlet of the fourth ventricle; bilateral mass located in the lower part of both foramina Luschka and midline mass located just superior from the foramen Magendie. Cerebrospinal fluid (CSF) analysis during EVD surgery showed elevated intracranial pressure (310 mmH₂O) and low glucose (13 mg/dL). Chest radiography disclosed small hilar lesion, which suggested sarcoidosis. Peripheral serum laboratory data showed a small increase in lysozyme (10.9 µg/mL:

Normal value 5.0–10.2 µg/mL) and high C-reactive protein (4.3 µg/mL: Normal value ≤0.20 µg/mL). However, the other laboratory data were almost normal. We speculated that the diagnosis was neurosarcoidosis, but could not confirm this diagnosis. Four days after EVD, we performed an endoscopic opening of the obstructed foramen Magendie, and a biopsy of the enhanced lesion located just superior from the foramen Magendie. Observing the MRI sagittal images, we decided the trajectory of endoscopic approach. After making a burr-hole at 2 cm anterior from the coronal suture and 2 cm lateral from the midline, we passed the endoscope (Olympus flexible endoscopy) into the fourth ventricle via foramen Monro and aqueduct of midbrain. Operative findings showed that smoothly thickened third ventricle floor without protruding mass lesion, and biopsy of the nodular lesions just superior from the foramen Magendie was performed [Figure 2a and c]. Endoscopic fenestration of foramen Magendie was performed to establish the flow of CSF [Figure 2b]. Pathological findings of the nodular lesions showed noncaseating epithelioid cell granuloma with multinucleated giant cells negative to tuberculin reaction [Figure 3]. Therefore, we confirmed the diagnosis of neurosarcoidosis. We started methylprednisolone sodium succinate 1000 mg/day for 3 days, followed by prednisolone at 60 mg/day. Ten days after admission, MRI showed a reduction of leptomeningeal enhancement, and an improvement of hydrocephalus [Figure 1c and d]. We expected the ventricle drainage to be unnecessary, but the volume of daily CSF outflow did not decrease. Therefore, he underwent ventriculo-peritoneal (V-P) shunt 20 days after Magendie foraminoplasty and biopsy for neurosarcoidosis. We could taper the dose of prednisolone. His consciousness was improved to GCS15. Finally, he was

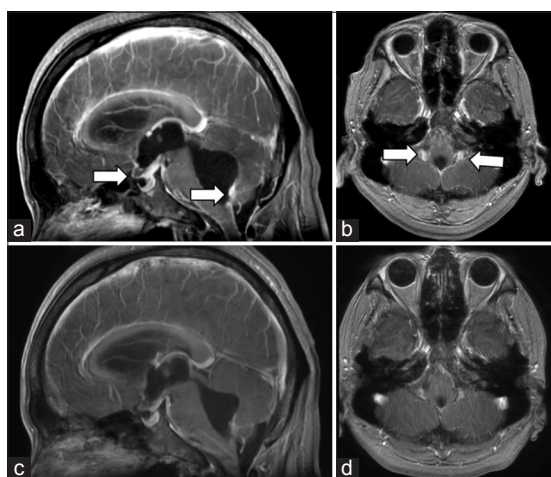


Figure 1: Radiological findings. (a and b) Initial magnetic resonance imaging study revealed leptomeningeal enhancement especially at the inferior portion of the fourth ventricle and suprasellar region (white arrows), ventricles were dilated. (c and d) In posterior fossa, bilateral mass located in the lower part of both foramina Luschka and midline mass located just superior from the foramina Magendie. Follow-up magnetic resonance imaging (1 week after steroid treatment) showed attenuation of the leptomeningeal enhanced lesions and hydrocephalus was improved

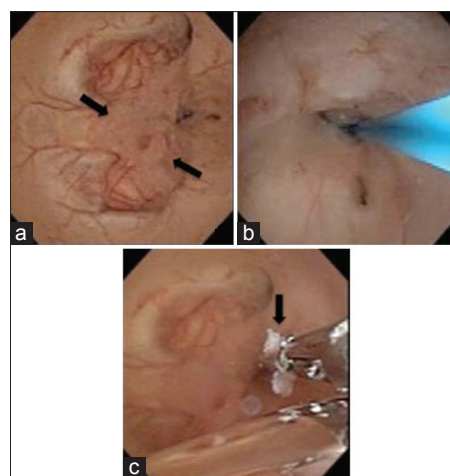


Figure 2: Intraoperative findings of the fourth ventricle floor. (a) the protruding mass lesions located just superior from the foramen Magendie (black arrows) and outlet of the fourth ventricle was obstructed by nodular lesions. (b) Ventriculostomy was performed at the obstruction of the fourth ventricle outlet. (c) Biopsy of the nodular lesions by endoscopy using forceps

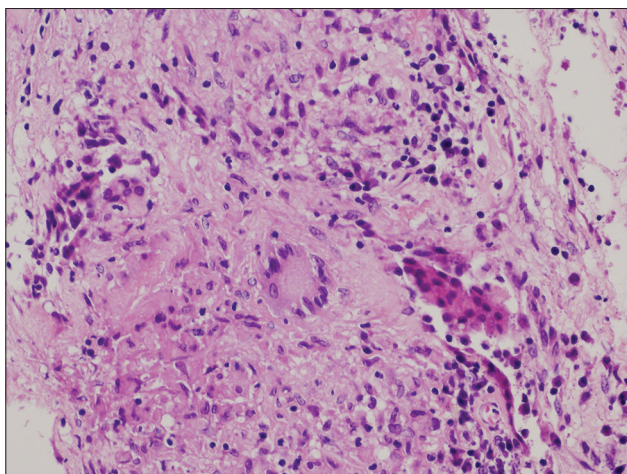


Figure 3: Pathological findings of nodular lesions of the fourth ventricle outlet. Photograph was taken at $\times 400$ magnification. H and E staining revealed granuloma containing giant cells

discharged with prednisolone 50 mg without recurrence and complications.

DISCUSSION

Sarcoidosis is a multisystem, noncaseous granulomatous disease of unknown etiology. Neurological manifestation of sarcoidosis are observed in approximately 5–20% of patients with sarcoidosis.^[4,8] Of the neurosarcoidosis, the most frequent manifestation is palsy of the eighth nerve. Other manifestations include optic neuritis, meningitis, vertigo, hearing abnormalities, hypothalamic and pituitary dysfunction, myopathy, seizure, and psychiatric symptoms.^[4,8,10] Hydrocephalus was involved in 6% of patients with neurosarcoidosis in a prospective study.^[2] Both obstructive and communicating hydrocephalus could occur with neurosarcoidosis, and the main cause of hydrocephalus was usually meningitis.^[5] Obstructive nodular lesion at the outlets of the fourth ventricle is extremely rare, and there is only one case reported with a detailed description in the CT/MRI era.^[6,9] Recently, endoscopic surgery for hydrocephalus caused by neurosarcoidosis was reported.^[3,5,9,11] Simple CSF shunt placement is not effective for hydrocephalus with multiple septations and compartments, whereas a combination therapy of endoscopic procedures of septostomy and V-P shunt is effective for multiple septum formations.^[5,7] In this case, we did not perform the third ventriculostomy to avoid surgical complications, because the nodular lesion at the third ventricle was too thick. Therefore, we performed endoscopic fourth ventriculostomy at the obstruction of foramen of Magendie, and a biopsy of the granulation tissue. To our knowledge, this is the second reported case of endoscopic-assisted biopsy of intracranial lesion for the diagnosis of neurosarcoidosis^[7] and the first report of endoscopic biopsy of the mass

lesion at the fourth ventricle outlet via foramen Monro and aqueduct. Although this technique is possible in very limited cases with enlarged aqueduct, we considered this simple and useful method. We assumed that the shunt tube could be removed after the endoscopic procedure, but the patient finally needed the V-P shunt treatment. We investigated the two reasons for hydrocephalus after treatment. In this case, we continued the EVD over 20 days after the endoscopic fourth ventriculostomy, due to which, the CSF outlet from the fourth ventricle might be re-occluded. We considered that the EVD had to be removed sooner after ventriculostomy. Another possibility is the dysfunction of CSF absorption because of meningeal sarcoidosis. In the case of Miscusi *et al.*,^[9] who received the third ventriculostomy, ventricle dilatation remained even after lesion-disappearance.

Here, we present a rare case of hydrocephalus with neurosarcoidosis. Our data suggests that endoscopic biopsy can enable “definite” diagnosis of neurosarcoidosis. We suggest that endoscopic surgery for hydrocephalus associated with neurosarcoidosis is not only therapeutic but also a biopsy tool for diagnosis. Endoscopic procedures are less invasive and effective for hydrocephalus associated with neurosarcoidosis, even when located at the floor of the foramen of fourth ventricle, and can be considered regularly in clinical practice.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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