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Case Report

Superficial siderosis and nonobstructive hydrocephalus due to subependymoma in the ventricle: An illustrative case report

Yuta Otomo[1#], Naoki Ikegaya[1], Akito Oshima[1], Shutaro Matsumoto[1], Naoko Udaka[2], Chia-Cheng Chang[3], Kensuke Tateishi[1], Hidetoshi Murata[1#], Tetsuya Yamamoto[1]

Departments of [1]Neurosurgery, [2]Pathology, Graduate School of Medicine, Yokohama City University, Yokohama, [3]Department of Neurosurgery, Iemasa Neurosurgical Clinic, Yokohama, Kanagawa, Japan.

E-mail: Yuta Otomo - o_yuta1018@yahoo.co.jp; Naoki Ikegaya - nikegaya@yokohama-cu.ac.jp; Akito Oshima - t216015c@yokohama-cu.ac.jp; Shutaro Matsumoto - shuu_shuu_1991@yahoo.co.jp; Naoko Udaka - naoudaka@yokohama-cu.ac.jp; Chia-Cheng Chang - changcc@td5.so-net.ne.jp; Kensuke Tateishi - ktate12@yokohama-cu.ac.jp; *Hidetoshi Murata - hmurata@yokohama-cu.ac.jp; Tetsuya Yamamoto - y_neuros@yokohama-cu.ac.jp

[#]Yuta Otomo and Hidetoshi Murata contributed equally to this work.

***Corresponding author:**
Hidetoshi Murata,
Department of Neurosurgery,
Graduate School of Medicine,
Yokohama City University,
Yokohama, Kanagawa, Japan.

hmurata@yokohama-cu.ac.jp

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ABSTRACT

Background: Intraventricular tumors can generally result in obstructive hydrocephalus as they grow. Rarely, however, some intraventricular tumors develop superficial siderosis (SS) and trigger hydrocephalus, even though the tumor has hardly grown. Here, we present an illustrative case of SS and nonocclusive hydrocephalus caused by subependymoma of the lateral ventricles.

Case Description: A 78-year-old man with an intraventricular tumor diagnosed 7 years ago had been suffering from gait disturbance for 2 years. He also developed cognitive impairment. Intraventricular tumors showed little growth on annual magnetic resonance imaging (MRI). MRI T2-star weighted images (T2*WI) captured small intratumoral hemorrhages from the beginning of the follow-up. Three years before, at the same time as the onset of ventricular enlargement, T2*WI revealed low intensity in the whole tumor and cerebral surface. Subsequent follow-up revealed that this hemosiderin deposition had spread to the brain stem and cerebellar surface, and the ventricles had expanded further. Cerebrospinal fluid (CSF) examination revealed xanthochromia. The tumor was completely removed *en bloc*. Histopathological findings were consistent with those of subependymoma. Although CSF findings improved, SS and hydrocephalus did not improve. Therefore, the patient underwent a lumboperitoneal shunt for CSF diversion after tumor resection.

Conclusion: Some intraventricular tumors cause SS and nonobstructive hydrocephalus due to microbleeding, even in the absence of tumor growth. T2*WI and, if necessary, timely CSF examination can allow identification of presymptomatic SS. This follow-up strategy may provide a favorable course by facilitating early intervention in patients with intraventricular lesions, not just subependymomas.

Keywords: Cerebrospinal fluid testing, Nonobstructive hydrocephalus, Subependymoma, Superficial siderosis, T2-star weighted image

INTRODUCTION

Intraventricular tumors can generally result in obstructive hydrocephalus as they grow. Subependymoma, which is a benign ventricular tumor, is generally followed up with imaging

unless they grow or result in hydrocephalus.[5,9,10,17] However, some tumors may develop superficial siderosis (SS)[1,7] and nonobstructive hydrocephalus, even if they do not expand.[8] Early detection of SS is important as SS causes irreversible symptoms.[6,12,13] However, the means for the early detection of SS in intraventricular tumor follow-up have not been established. Here, we present a case of subependymoma of the lateral ventricle. The patient's tumor had barely grown; nevertheless, superficial hemosiderosis and normal pressure hydrocephalus progressed slowly, exacerbating dementia, and gait disturbance. We present this illustrative case and discuss the pathology and follow-up strategies.

CASE DESCRIPTION

A 78-year-old man with an intraventricular tumor diagnosed 7 years before had been suffering from gait disturbance for 2 years [Figure 1, 1a and 1b]. He also developed cognitive impairment below the Mini-Mental State Examination cutoff value (score <24) 1 year ago. Intraventricular tumors showed annual growth on imaging. The tumor did not lead to occlusion of the foramen of Monro, but ventricular enlargement had been observed 3 years before and had worsened, reaching an Evans index of 0.36 [Figure 1d]. Magnetic resonance imaging (MRI) T2*WI captured small intratumoral hemorrhages from the beginning of the follow-up [Figure 1a, b and c]. Three years before, at the same time as the onset of ventricular enlargement, low intensity of the whole tumor and cerebral surface was observed on T2*WI [Figure 1d]. Subsequent follow-up revealed that this hemosiderin deposition had spread to the brain stem and cerebellar surface, and the ventricles had further expanded [Figure 1e]. A hearing test revealed bilateral sensorineural hearing impairment, which is a permanent symptom of SS.

The timed up and go test before and after the cerebrospinal fluid (CSF) tap test in the presurgical evaluation showed improvement in gait disturbance, suggesting responsiveness to CSF diversion. On the other hand, CSF analysis demonstrated xanthochromia, a white blood cell (WBC) count of 154/μl (normal range 0–15/μL), and protein level of 596 mg/dL (normal range 10–40 mg/dL) [Figure 2 and 2a]. The CSF opening pressure was 7 cmH_2O (normal range 6–15 cmH_2O). This result suggested that persistent bleeding from intraventricular tumors led to the development and exacerbation of SS and nonobstructive NPH. He was then

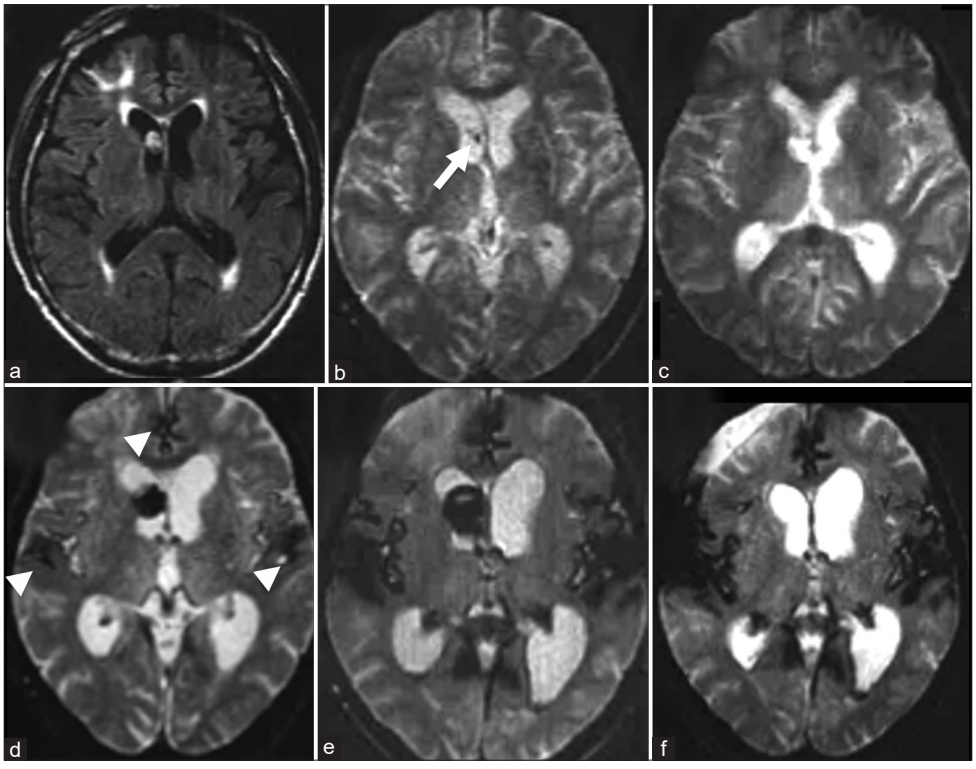

Figure 1: Magnetic resonance imaging (MRI) findings. Initial fluid-attenuated inversion recovery MRI showing an intraventricular tumor (a). Initial T2-star weighted MRI (T2*WI) also revealed a tiny low-intensity spot (arrow), suggesting intratumoral hemorrhage 7 years before surgery (b). Note that 5 years before surgery, this signal change was localized within the tumor and not on the brain surface (c). Low intensity suggesting intratumoral hemorrhage and superficial siderosis on the brain surface (arrowhead) is demonstrated on T2*WI 3 years before surgery and ventricular enlargement progresses (d), and is exacerbated immediately before surgery (e). Hemosiderin deposition and hydrocephalus persisted even after tumor removal (f).

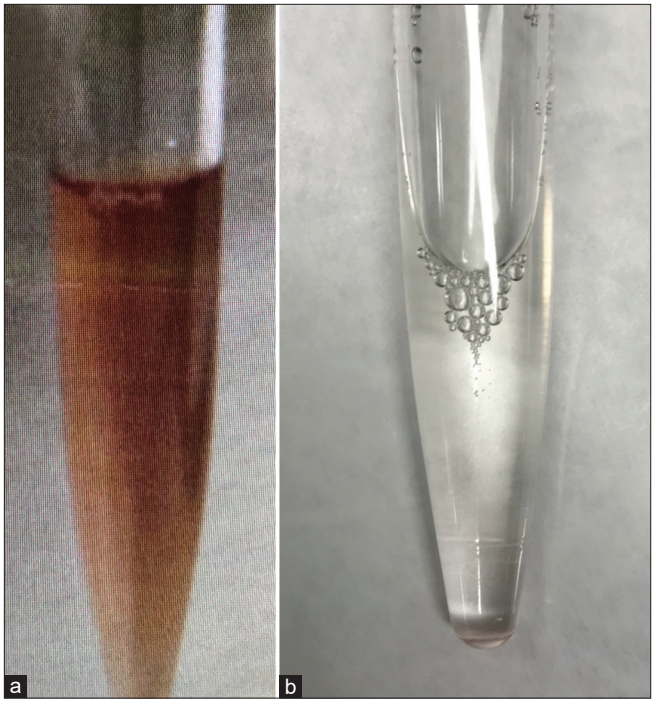

Figure 2: Cerebrospinal fluid (CSF) testing before and after surgery. (a) CSF before surgery showing xanthochromia. (b) CSF after surgery showing clear crystals.

advised to receive treatment for the tumor and was referred to our hospital after 7 years of follow-up of the tumor. The patient wanted to have the tumor surgically removed.

Tumor resection was performed through a right frontal craniotomy. Visual inspection revealed that the ventricular wall turned yellowish-brown, suggesting deposition of hemosiderin on the ependyma [Figure 3a]. The tumor surface was a mixture of yellow and red, suggesting the presence of hemorrhage at different times [Figure 3b]. The tumor was completely removed *en bloc* by dissection from the septum pellucidum.

Histopathological findings were consistent with those of subependymomas. Although the tumor surface color suggested that multiple hemorrhages in different phases and slight microbleeding changes were observed in the tumor, Elastica van Gieson staining did not reveal any significant disruption of the tumor blood vessels [Figure 4].

During the postoperative course, MRI revealed total tumor removal and no postoperative complications [Figure 1f]. Postoperative CSF was drastically improved, being crystal clear, WBC count of 1/μl, and protein level of 88 mg/dL [Figure 2b]. However, ventricular enlargement and NPH-related symptoms did not improve. Therefore, an LP shunt was performed for NPH 4 weeks after tumor removal. Unfortunately, he developed shunt malfunction and infections 3 weeks after surgery without any improvement.

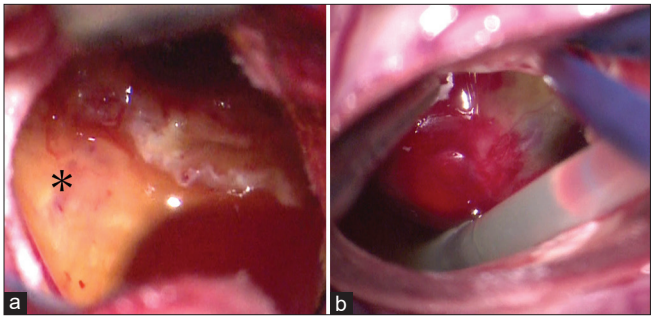

Figure 3: Intraoperative photographs of the left lateral ventricle. (a) Black asterisk showing the ventricular wall is colored yellowish-brown, suggesting deposition of hemosiderin on the ependyma. (b) The tumor surface turns a mixture of yellow and red, suggesting the presence of hemorrhage at different times.

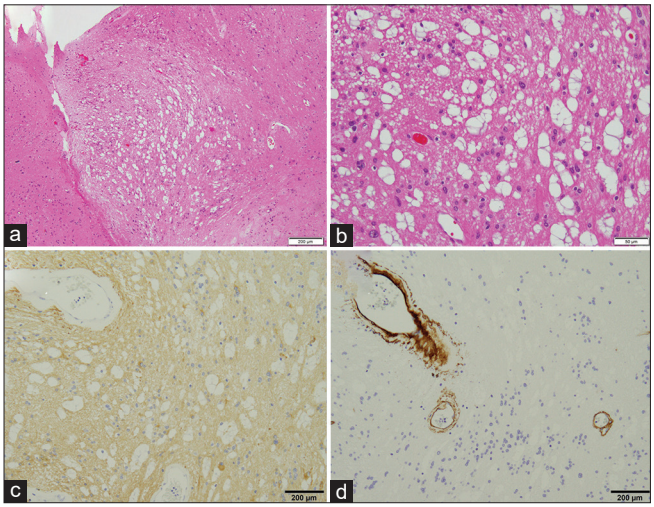

Figure 4: (a) Low magnification and (b) high magnification: hematoxylin and eosin staining. Clusters of oval-shaped cells and isomorphic nuclei are embedded in a fine fibrillary back ground. Many microcystic formations and microbleeding changes were also seen in the tumor. (c) Immunohistochemical staining revealing a positive reaction for glial fibrillary acidic protein. The pathological findings of a-c are consistent with those of subependymoma. (d) Elastica van Gieson staining revealed a positive reaction for collagen type IV on the vessel walls, which suggests that the vessel walls in the tumor are normal.

He also developed severe pneumonia and died a month after LP shunt placement.

DISCUSSION

We describe a case of SS and hydrocephalus caused by subependymoma of the lateral ventricles. It is very important to recognize that some intraventricular tumors can cause SS and nonobstructive hydrocephalus without tumor growth due to microbleeding. The clinical course of the patient has two important clinical issues. First, subependymoma of the

lateral ventricles can be a source of bleeding, leading to SS and nonobstructive hydrocephalus. Second, MRI T2*WI and a final CSF examination might allow the identification of presymptomatic SS and prevent irreversible SS through intraventricular tumor resection.

This case macroscopically showed bleeding on both the surface and the inside of the tumor, but histopathological examination failed to reveal evidence of significant disruption of the tumor vessel walls. This finding may indicate that intratumoral bleeding was limited to very minor bleeding. In fact, MRI showed SS but no apparent intraventricular hemorrhage. Vascular degeneration and coexisting cavernous-like malformations may contribute to subependymoma hemorrhage, but the cause of bleeding is not always clear, as in our case.[2,19] Although hemorrhagic presentation of subependymoma is rare, observed in <10% of cases on imaging and pathological examination,[3,16] it would be meaningful, on follow-up, to recognize that subependymoma hemorrhage can occur in the CSF space, leading to SS. In this case, hemorrhagic changes in T2*WI, SS, and hydrocephalus appeared simultaneously, suggesting a causal relationship between nonobstructive NPH and SS. Chronic bleeding into the CSF space can cause SS and hydrocephalus, both of which harm the brain surface and arachnoid granulations.[11] The mechanism of NPH may be explained by chronic bleeding into the CSF resulting in SS as well as dysfunction of Pacchionian granulations with malabsorption of CSF.[1,7] Therefore, tumor resection may also prevent nonobstructive NPH by precluding SS. However, NPH could also be caused by other causes. In such case, tumor resection is more significant to prevent progression of SS than to prevent progression of hydrocephalus.

Second, MRI T2*WI can identify hemorrhagic changes with high sensitivity and is useful for the detection of bleeding that remains in the lesion, as in this case.[4] Of course, persistent bloody CSF does not always occur, even if intratumoral hemorrhage is observed. However, MRI screening is easy to repeat and is sufficient to issue a warning. Detection of changes on MRI T2*WI allows us to determine if the follow-up strategy needs to be intensified. Hemorrhage into the CSF space is a prerequisite for the pathogenesis of SS and should have existed before the establishment of SS. Therefore, bloody CSF could provide crucial information regarding the risk of SS in patients with intraventricular lesions. In a previous report, bloody CSF was associated with an SS at a high rate of 91%, with a reliable diagnostic value.[12]Although bleeding may not be detected, it can still occur within the lesion. Therefore, we believe that the early detection of SS with high sensitivity and specificity would be possible by screening for bleeding with T2*WI change and, if necessary, confirmation with timely CSF examination. The present case showed that resection of the tumor, which was the bleeding source, was effective in improving bloody CSF, and early intervention may have prevented the progression to established SS.

Our follow-up strategy with MRI T2*WI and timely CSF examination can also be applied to other lesions in the CSF space. Ependymoma is the major tumor of the central nervous system that causes SS, and is occasionally associated with intratumoral hemorrhage in imaging studies.[8,14] This indicates that our strategy may be useful for ependymomas. Furthermore, other lesions such as meningioma, paraganglioma, pilocytic astrocytoma, and craniopharyngioma can cause SS, suggesting the potential utility of this follow-up method.[8,15,18]

CONCLUSION

Some intraventricular tumors can cause SS and nonobstructive hydrocephalus, even if the tumor has not grown. We describe a case of SS and hydrocephalus caused by subependymoma of the lateral ventricles. T2*WI-based radiographic screening and, if necessary, CSF examination might allow the identification of presymptomatic SS and prevent irreversible SS through intraventricular tumor resection. Early detection of SS would be helpful in timely early intervention, contributing to a favorable course in patients with intraventricular lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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