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Case report

A large cavernous malformation of the third ventricle floor: A case report

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ABSTRACT

Suprasellar and third ventricular region cavernous malformations originating from the floor of the third ventricle are extremely rare. We report a case of third ventricular cavernous malformation arising from the ventricle floor in a 24-year-old woman who presented with short-term memory loss and disorientation. Computed tomography revealed a suprasellar mass with calcification in the posterior chiasmatic region. T2-weighted magnetic resonance imaging revealed a mass with heterogeneous intensity and without hydrocephalus. The mass was slightly enhanced subsequent to gadolinium infusion. Using a basal interhemispheric translamina terminalis approach and a neuroendoscope, we confirmed that the tumor was located at the floor of the third ventricle and removed it. Histopathological examination confirmed the diagnosis of cavernous malformation. The postoperative course was uneventful, but the patient's short-term memory loss persisted. Despite its rarity, cavernous malformation should be suspected when a tumor is detected in the vicinity of the third ventricle floor. It is treatable through surgical resection.

1. Introduction

Cavernous malformations (CMs) in the suprasellar region and third ventricle are very rare, and information about them is limited to a small number of case reports [1–4]. In addition, CM originating from the floor of the third ventricle is extremely rare and has only been reported in three cases [4–6]. Here, we describe a rare case of suprasellar CM arising from the floor of the third ventricle.

2. Case report

2.1. History and examination

A 24-year-old woman presented with a 2-week history of short-term memory deficit, disorientation, and reduced ability to engage in activities of daily living. The patient had no significant prior medical history, and examination revealed no apparent cranial nerve palsy, motor paresis, headaches, or vomiting. There were no symptoms of hydrocephalus or signs

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of hypothalamic dysfunction, and her visual field and acuity were intact. With respect to routine blood tests, her prolactin level was high (89.7 ng/ml; normal: 6.12–30.54 ng/ml), and levels of growth hormone, adrenocorticotropic hormone, cortisol, thyroid-stimulating hormone, and free T3 and T4 were normal. Follicle-stimulating and luteinizing hormones were not assessed. Results of insulin and thyrotropin- and gonadotropin-releasing hormone stimulation tests were also normal. Her Mini Mental State Examination score was 30, which was the highest possible score. Her intelligence quotient was 93 on the Wechsler Adult Intelligence Scale III, and her memory quotient was 67 on the Wechsler Memory Scale-Revised; these values were not inferior to the average values for healthy adults.

2.2. *Imaging findings*

Preoperative cranial plain computed tomography (CT) imaging showed a heterogeneous, calcified mass in the suprasellar region, without enlargement of the lateral ventricles (Fig. 1). Magnetic resonance imaging (MRI) showed a 2.2 cm suprasellar mass extending to the floor of the third ventricle (Fig. 2). T1-weighted images showed a heterogeneously iso-to-high intensity mass, and T2-weighted images showed a heterogeneously low-to-high intensity mass surrounded by extremely low signal intensity. The lesion was slightly enhanced following gadolinium administration. The mass appeared to be a suprasellar lesion associated with the pituitary stalk and third ventricle. The tumor reached the foramen of Monro and displaced the third ventricle. Susceptibility-weighted imaging (SWI) and Gradient echo images not studied preoperatively. Cerebral angiography showed no tumor stain or abnormal vessels. Craniopharyngioma and CM were considered as differential diagnoses.

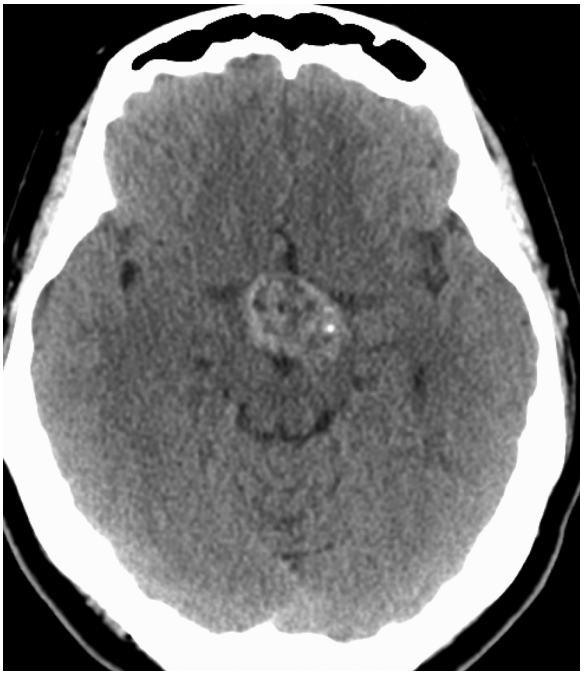

Fig. 1 – Preoperative plain computed tomographic scan showing a heterogeneous and calcified mass in the suprasellar region.

2.3. *Operation*

A basal interhemispheric translamina terminalis approach was used. The lamina terminalis was xanthochromatic. Gliosis was discovered when the lamina terminalis was opened, but there was no cerebrospinal fluid (CSF) leakage. The tumor was xanthochromatic and surrounded by a layer of gliosis and contained evidence of old and new hemorrhage. The basilar artery was detected subsequent to complete removal of the tumor. Neuroendoscopic examination also verified the tumor cavity, basilar artery, and mammillary body. However, the choroid plexus and foramen of Monro were not observed, suggesting that the tumor did not grow up in the third ventricle. These findings suggest that the tumor developed within the floor of the third ventricle.

2.4. *Pathological findings*

Histologically, the lesion showed cavernous aggregation of dilated vessels filling peripheral blood and fibrin, lined with bland-looking endothelial cells. No smooth muscle layer was observed in the wall of the vessel. Gliosis with scattered hemosiderin-laden macrophages was observed in the surrounding cerebral tissue. There was no neuronal tissue within the lesions, confirming the diagnosis of CM (Fig. 3).

2.5. *Postoperative course*

The postoperative course was uneventful. Postoperative MRI revealed normal ventricular size and complete removal of the cavernous malformation (Fig. 4a). The floor of the third ventricle was preserved on postoperative sagittal MRI (Fig. 4b, c arrow). The patients transferred to the rehabilitation hospital in three weeks after surgery. Hypopituitarism and diabetes insipidus were controllable with medications of levothyroxine, hydrocortisone and desmopressin acetate. Disorientation was improved, but short-term memory loss persisted. The patients were able to live with family support after 2-month rehabilitation.

3. Discussion

3.1. *Clinical features*

CMs are benign vascular malformations of the central nervous system. Intracranial CMs account for 10–15% of intracranial vascular malformations and have been observed in up to 0.4–0.8% of the population [7]. They are typically located in the subcortical white matter, basal ganglion, brain stem, or cerebellum. Common symptoms include headaches, seizure, and focal neurological deficit [8].

CMs located in the suprasellar region represent 4% of all supratentorial CMs [2]. The major symptoms of CMs that involve the suprachiasmatic region include visual field restriction and endocrine dysfunction. Whereas, in the case classified as the third ventricle CM, the major symptoms were disorientation and short-term memory deficit. Most of that

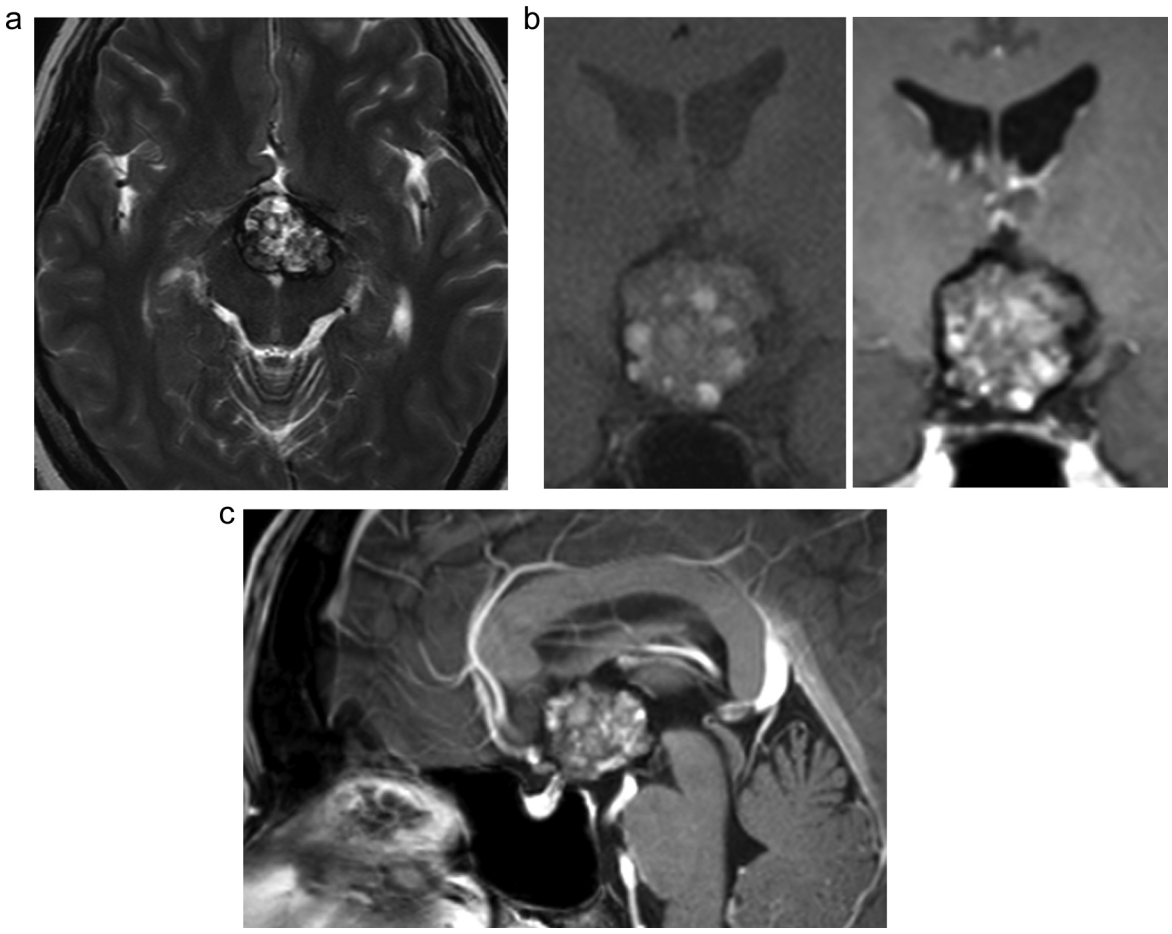

Fig. 2 – Preoperative magnetic resonance images. (a) A T2-weighted coronal image showing a heterogeneous low-to-high signal with a surrounding low-intensity rim. (b) Pre- (left) and postcontrast (right) T1-weighted axial images showing a slightly enhanced tumor. (c) Postcontrast T1-weighted sagittal images showing a homogeneously enhanced tumor in the posterior and superior chiasmatic lesion.

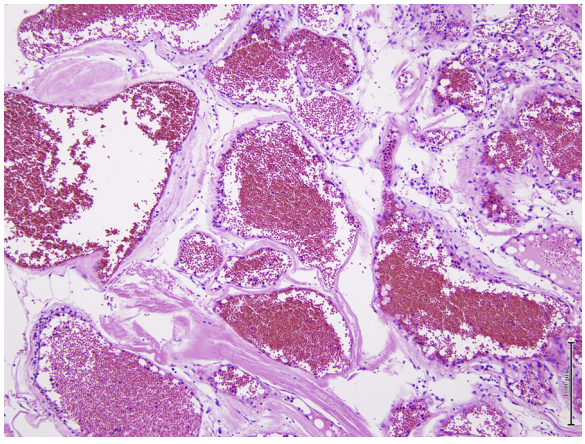

Fig. 3 – Photomicrograph of the tumor (hematoxylin and eosin stain, original magnification 100×) showing gaping vascular spaces with simple endothelial linings and scant stroma.

case were progressed into the ventricle and were often reported as intraventricular CM [4–6]. Tumors involving the foramen of Monro may include hydrocephalus, leading to headaches and vomiting. In CMs originating in the lateral wall and floor, the most common initial symptoms are short-term memory loss, disorientation, and depression [5].

Several groups have reported CMs of the parasellar region. The most common origins were dural-based or anterior optic pathways [3]. With respect to third ventricular CMs originating from the floor of the ventricle, only two cases have been reported [4–6]. The first case described a CM originating from one of the mammillary bodies and involved memory deficit [4]. The second case was reported as a third-ventricle CM, and the patient presented with short-term memory loss and disorientation [5]. In the third case, the patient presented with gait disturbance due to a CM located in the left thalamus and hypothalamus [6]. In these two of three reports, the CM was adherent to the floor of the third ventricle and extended into the ventricle. In the case reported herein, the short-term memory deficit and disorientation observed were similar to

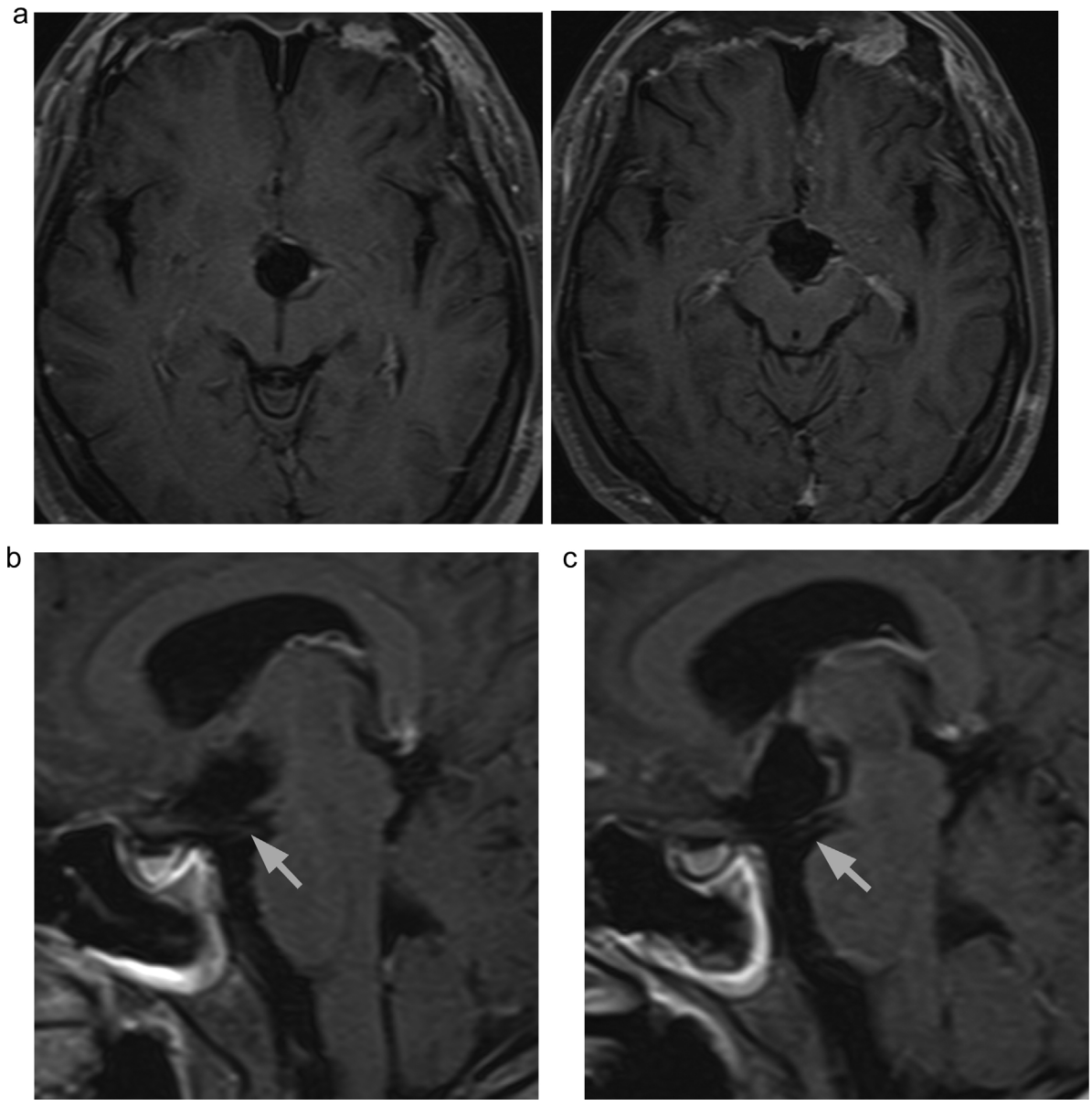

Fig. 4 – Postoperative MRI. (a) Postcontrast T1-weighted axial images showing that the tumor was completely removed. (b) and (c) Postcontrast T1-weighted sagittal images demonstrating preservation of the floor of the third ventricle (arrows).

that seen in the two cases described above. Interestingly, based on the endoscopic findings, the tumor developed in the floor of the third ventricle, without growth or progression into the ventricle. Postoperative MRI findings showed that the third ventricle floor was preserved, consistent with our insight regarding the primary site of the tumor.

3.2. *Imaging studies and differential diagnosis*

Intracranial CM typically shows hyperdensity on CT images, due to calcification and recent hemorrhage. Intracranial cavernous malformations exhibit mixed-intensity signals in T1- and T2-weighted MRI and are typically surrounded by a hypointense area (hemosiderin) in T2-weighted MRI [9]. Although CM is relatively easy to diagnose when located within the parenchyma, suprasellar CMs can be difficult to distinguish from other tumors because of their uncommon locations. Radiographic differential diagnoses for suprasellar lesions include craniopharyngioma, teratoma, and astrocytoma [10,11]. Sinson et al. [10] remarked on the difficulty experienced in distinguishing between CM and papillary craniopharyngioma. The short-term memory loss and disorientation observed in this case are not typical initial symptoms of craniopharyngioma. Teratomata are usually localized to pineal and suprasellar regions and cause hyperurea with hypothalamic dysfunction. Neuroimaging shows a mixed density mass with calcification and cysts [12]. Tumor biomarkers, such as beta-human chorionic gonadotropin and alpha-fetoprotein,

may help to diagnose nongerminomatous germ cell tumors such as embryonal carcinoma, choriocarcinoma, endodermal sinus tumor. Although pilocytic astrocytomata are rare in adults, they mimic the appearance of craniopharyngiomata, which is a potential radiological and diagnostic pitfall [13].

3.3. *Treatment and outcome*

Although close follow up for observation is indicated for asymptomatic CMs, the typical treatment for a symptomatic supratentorial CM is surgical resection [14]. In our case, surgical removal was performed successfully using an interhemispheric translamina terminalis approach. The third ventricle floor and lateral wall are closely associated with memory impairment [15]. Memory dysfunction can also be caused by damage to the thalamus, mammillary bodies, mammillothalamic tract, fornix, and internal medullary lamina [16]. Recovery of short-term memory is considered difficult, even if a tumor is completely removed [4,15].

In conclusion, CM should be included in the differential diagnosis of tumors located in the floor of the third ventricle. The short-term memory deficit and mixed intensity observed in T2-weighted imaging may support preoperative diagnosis. Surgical removal of large CMs originating in the floor of the third ventricle should be considered in symptomatic patients.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article was conducted in accordance with the World Medical Association (Declaration of Helsinki) Ethical Principles for Medical Research Involving Human Subjects, and uniform requirements for manuscripts submitted to Biomedical Journals.

Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:10.1016/j.pjnns.2015.08.004.

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