Pineocytoma With Massive Intratumoral Hemorrhage After Ventriculoperitoneal Shunt

—Case Report—

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Abstract

A 58-year-old male suffered a massive intratumoral hemorrhage after placement of a ventriculoperitoneal shunt before treatment for pineocytoma. At admission, he complained of headache, urinary incontinence, gait disturbance, and recent memory disturbance. His neurological examination revealed impaired memory and papilledema from hydrocephalus. Two days after the placement of the ventriculoperitoneal shunt, he suddenly became stuporous and Parinaud’s sign appeared. Computed tomography showed a massive intratumoral hemorrhage occupying the third ventricle which had extended into the lateral ventricle. The infratentorial supracerebellar approach was used to remove the hematoma and the tumor. The histological diagnosis was pineocytoma. Intratumoral hemorrhage in pineocytoma after the placement of a ventriculoperitoneal shunt is rare, but should be kept in mind when a ventriculoperitoneal shunt is placed before a craniotomy.

Keywords: pineocytoma, intratumoral hemorrhage, ventriculoperitoneal shunt

Introduction

Primary pineal tumors are rare, comprising only 0.4% to 1% of primary brain tumors. Consequently, their clinical picture remains unclear, but pineocytomas tend to occur in adults, grow slowly, and often cause hydrocephalus. A pineocytoma associated with hemorrhage is extremely rare. We report a case of massive intratumoral hemorrhage after the placement of a ventriculoperitoneal shunt in a patient with a pineocytoma. The hemorrhage caused the patient’s clinical condition to deteriorate suddenly.

Case Report

A 58-year-old male was referred to our service with persistent headache. On admission, he complained of headache, urinary incontinence, gait disturbance, and recent memory disturbance. Neurological examination revealed impaired memory, gait ataxia, and bilateral papilledema but his ocular movements and pupillary function were normal. Computed tomography (CT) showed moderate hydrocephalus and an isodense lesion in the pineal region (Fig. 1). T1-weighted magnetic resonance (MR) imaging showed the lesion as a low-intensity area which was homogeneously enhanced after the administration of gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA). T2-weighted MR imaging showed a high-intensity area (Fig. 2). Laboratory examination, including the bleeding time, were within normal limits. Tumor markers, such as human chorionic gonadotropin, alpha-fetoprotein, carcinoembryonic antigen, and placental alkaline phosphatase, were normal in both serum and cerebrospinal fluid. Carotid and vertebral angiography showed no vascular staining in the pineal region, and no aneurysm or arteriovenous malformation. A ventriculoperitoneal shunt was placed to alleviate the patient’s hydrocephalus (Codman-Medos programmable valve shunt system; Medos, Lelocle, Switzerland). The initial pressure was 20 cmH2O and the programmed pressure was 7 cmH2O.

The patient’s symptoms markedly improved after the shunt was placed, and he became well oriented and recovered his memory. Two days after the shunt procedure, the patient became stuporous and paresis of the conjugate upward gaze (Parinaud’s sign) appeared. Immediate CT showed a massive in-
tratumoral hemorrhage in the third ventricle which had ruptured into the lateral ventricle, causing obstructive hydrocephalus (Fig. 3). An external ventricular drain was placed in the right lateral ventricle, and the patient's level of consciousness improved. MR imaging 2 days after ventricular drainage showed reduction in the ventricular size and an intratumoral hematoma in the third ventricle (Fig. 4). Sagittal T₁-weighted MR imaging with Gd-DTPA disclosed an enlarged and enhanced capsule, and the tumor itself was displaced posteriorly (Fig. 5).

A week after the external ventricular drainage, the lesion was removed via the infratentorial supracerebellar approach with the patient in the Concord position. Incision of the arachnoid membrane disclosed the brownish-gray tumor enclosed in a capsule. Removal of the posterior portion of the tumor allowed easy evacuation of the hematoma. Both the tumor and hematoma were totally removed. The histological diagnosis was pineocytoma with uniform, proliferating spherical cells in a lobular pat-
tern. No abnormal proliferation of vessels was observed. Postoperative MR imaging showed that the tumor was totally removed (Fig. 6).

Postoperatively, the patient was alert and his gaze paresis was completely resolved. One month after surgery, he was fully ambulatory and underwent whole-brain radiation therapy to treat any potential seeded tumors (40 Gy to the entire ventricular system followed by 10 Gy boost to the tumor bed). The patient was discharged 3 months after surgery with no neurological deficits.

Discussion

Pineal tumors include a diverse group of tumor types, ranging from very benign to highly malignant. Identification of tumor histology is essential for determining optimal tumor-specific management, and some patients may benefit from radiation therapy or chemotherapy specifically directed.
against the tumor. We treat germinoma by radiotherapy following stereotactic biopsy, meningioma and mature teratoma by surgical resection, and the other malignant tumors by multimodality treatment including surgery, radiotherapy, and chemotherapy. The treatment of pineocytoma is controversial because of the behavioral variability. Most authors include pineocytomas among malignant neoplasms of the pineal region and advocate surgical intervention with the intention of aggressive surgical resection followed by radiation therapy, modifying the planning of adjuvant therapy, including the extent of the field of irradiation, depending on the differentiation pattern of the tumor.\textsuperscript{4,5,8)}

Spontaneous intratumoral bleeding is rare in primary tumors of the central nervous system.\textsuperscript{1,2,9,10)} Most cases have occurred in patients with highly vascular tumors, such as pituitary adenomas, metastatic lesions, and glioblastoma multiforme.\textsuperscript{7)} Hemorrhage associated with a pineocytoma is quite rare.\textsuperscript{2,9)} A case of apoplexy due to hemorrhage into a pineal cyst occurred during anticoagulant therapy,\textsuperscript{11)} but there are no other cases of hemorrhage from a pineal tumor. Two cases of subarachnoid hemorrhage occurred in patients with initial symptoms resembling a subarachnoid hemorrhage.\textsuperscript{3)} The underlying tumor became obvious only later in the course of the disease, and the tumor was diagnosed postmortem. MR imaging detected asymptomatic intratumoral hemorrhage in a patient with a pineocytoma 10 days after the placement of a ventriculoperitoneal shunt.\textsuperscript{3)} The hemorrhage was small and limited. We have treated four patients with pineocytoma in the past 30 years, but this patient was the first to suffer intratumoral hemorrhage after the placement of a ventriculoperitoneal shunt before craniotomy.

Intratumoral hemorrhage after the placement of a ventriculoperitoneal shunt is rare, and most often associated with posterior fossa tumors.\textsuperscript{9)} One case of fatal intratumoral hemorrhage occurred after the placement of a ventriculoperitoneal shunt in a patient with a posterior fossa ependymoma.\textsuperscript{11)} The sudden and dramatic decline in the patient's clinical state after ventricular decompression suggests that inserting the ventriculoperitoneal shunt caused the bleeding even though the valvular pressure of the

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**Fig. 5** Sagittal magnetic resonance images with gadolinium-diethylenetriaminepenta-acetic acid before (left) and after (right) the intratumoral hemorrhage showing an enlarged and enhanced capsule and displacement of the tumor posteriorly.

**Fig. 6** Postoperative axial magnetic resonance images showing total removal of the tumor, $T_1$-weighted image (left), $T_2$-weighted image (center), $T_1$-weighted image with gadolinium-diethylenetriaminepenta-acetic acid (right).
The ventricles in our patient were considerably smaller after the shunt was placed. We believe that this abrupt reduction in pressure affected the tumor mechanically, causing its distortion. After the ventriculoperitoneal shunt was placed and the ventricles were reduced in size, the tumor shifted away from the surrounding structures. This shift may have altered the venous circulation in the tumor, causing the intratumoral hematoma. Immediate CT after hemorrhage showed an intimate relation between the drain and the hematoma. This suggests another possibility for the origin, that the hemorrhage was caused by laceration of the tumor by the drain.

Hydrocephalus is often associated with pineal region tumors, and this problem must be addressed before surgery. Many neurosurgical services have established the practice of placing a ventriculoperitoneal shunt or an external ventricular drain one or more days before a direct surgical approach to posterior fossa tumors or pineal region tumors. This practice is accepted as useful in facilitating a direct surgical approach and lowering mortality. However, the surgeon must prevent excessive reduction of the cerebral ventricles by modifying the opening pressures in patients with obstructive hydrocephalus. The possibility of intratumoral hemorrhage in a pineocytoma after the placement of a ventriculoperitoneal shunt should be considered when a shunt is placed before a craniotomy, although this does not affect the clinical decision making in a patient with pineal region tumor and hydrocephalus.

References


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