Pineoblastoma Showing Unusual Ventricular Extension in a Young Adult
—Case Report—

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Abstract

A 19-year-old male presented with a 4-week history of headache. Neurological examination showed bilateral papilledema. Computed tomography revealed a pineal region mass with remarkable obstructive hydrocephalus. Magnetic resonance imaging showed a pineal region tumor continuously invading through the tectum into the cerebral aqueduct and the fourth ventricle with the preservation of the adjacent structures. The tumor appeared an iso- to hypointense mass on T<sub>1</sub>-weighted images, a heterogeneous iso- to hyperintense mass on T<sub>2</sub>-weighted images, and a heterogeneously enhanced mass after administration of contrast medium. Histological examination after endoscopic biopsy confirmed that the tumor was a pineoblastoma. Radiotherapy was given to the whole brain and the spinal cord, and magnetic resonance imaging showed complete remission of the tumor. Pineoblastomas are highly malignant tumors with seeding potential through the neighboring ventricle or along the meninges, and this type of tumor becomes larger with local extension. We found no previous reports of the continuous extension into the fourth ventricle. The present case showed ventricular extension with minimal mass effect to adjacent structures, and did not disturb ventricular configuration. According to the unusual finding of ventricular extension, this rare case of pineoblastoma requires adjuvant chemotherapy.

Key words: pineoblastoma, pineal region tumor, magnetic resonance imaging

Introduction

Pineal region tumors are uncommon, accounting for 3–8% of all intracranial tumors in children, and 0.1–1.0% in adults. The most common types of tumors are the germ cell tumors that occur in young males. Pineal region masses can be classified histologically into four groups: germ cell tumors, pineal parenchymal tumors, glial tumors, and non-neoplastic masses. Pineal parenchymal tumors account for only less than 15% of all pineal region masses. Pineal tumors pose a diagnostic problem due to the pathological variation and deep internal location. The computed tomography (CT) features of pineal region neoplasms are known, but the magnetic resonance (MR) imaging characteristics of pineoblastomas are unclear. MR imaging is extremely important for the diagnosis of pineal region neoplasms, and is useful for the early detection of both intracranial and spinal seeding. We describe a rare case of pineoblastoma that invaded through the tectum into the cerebral aqueduct and the fourth ventricle with minimal displacement of the quadrigeminal plate, and the MR imaging features of pineoblastoma.

Case Report

A 19-year-old male presented with a 4-week history of mild headache. Physical examination found se-
vere bilateral chronic papilledema without optic atrophy.

CT revealed a pineal region mass invading the fourth ventricle with a small amount of calcification at the pineal gland, and remarkable obstructive hydrocephalus without dilation of the cerebral aqueduct and fourth ventricle (Fig. 1). MR imaging showed an iso- to hypointense mass on T₁-weighted images (Fig. 2A) and a heterogeneous iso- to hyperintense mass on T₂-weighted images (Fig. 2B). The mass was heterogeneously enhanced after intravenous administration of 0.1 mmol/kg of gadopenitetate dimeglumine. Sagittal MR imaging showed the tumor extended continuously to the cerebral aqueduct through the tectum from the pineal gland to the fourth ventricle. The ventricular configuration was preserved without displacement of the quadrigeminal plate (Fig. 2C, D). There was no evidence of dissemination to the rest of the central nervous system or any systemic metastatic lesions. Bilateral vertebral angiography showed no tumor staining. Serum levels of the tumor markers, alphafetoprotein and human chorionic gonadotropin β were <0.1 ng/ml (normal <0.1) and 1.2 ng/ml (normal <10.0), respectively. These features suggested a pineal region tumor or a tumor of ventricular origin such as plastic ependymoma.

Endoscopic biopsy was performed via the right anterior horn and the third ventricle. A third ventriculostomy was performed at the same time without complications. No tumor cells appeared in the cerebrospinal fluid (CSF) obtained by the initial ventricular puncture performed at surgery. Intraoperative endoscopic findings revealed a well-demarcated tumor extending from the posterior part of the third ventricle into the cerebral aqueduct. Three specimens were obtained with biopsy forceps from different parts of the tumor. Histological examination found a highly cellular neoplasm composed of small and round tumor cells (Fig. 3 upper). The cells had hyperchromatic oval nuclei and scant cytoplasm. The tumor cells were partially arranged...
in Homer-Wright rosettes (Fig. 3 lower). All the three specimens from different parts of the tumor showed same findings. Immunohistochemical staining found no cells positive for glial fibrillary acidic protein, synaptophysin, or S-100 protein, but some cells were positive for neuron-specific enolase. Based upon these findings, the diagnosis was pineoblastoma.

The postoperative course was uneventful and his headaches resolved immediately after treatment. The patient underwent craniospinal irradiation (30 Gy to the whole brain with a 24 Gy boost to the posterior fossa and 30 Gy to the spinal cord). MR imaging revealed complete remission of the tumor after craniospinal irradiation was completed (Fig. 4). The third ventriculostomy had improved the obstructive hydrocephalus. Implantation of a ventriculoperitoneal shunt and the subsequent risk of systemic metastasis through the shunt were avoided by performing the endoscopic third ventriculostomy. In addition, the patient will receive adjuvant chemotherapy.

Discussion

Pineal parenchymal tumors can be divided into three histological groups: pineocytomas, which are benign; pineoblastomas, which tend to metastasize to the cerebrospinal axis and directly invade the adjacent structures; and mixed pineocytomas and pineoblastomas, which have intermediate characteristics. Pineoblastomas are highly cellular and consist of poorly differentiated cells with scant cytoplasm and processes, and appear as patternless sheets, similar to those of medulloblastomas, retinoblastomas, and other embryonal or primitive neuroectodermal tumors.

Only a small number of cases of pineoblastomas have been described, so the clinical, histological, and radiographic features are still being defined. The largest published series of pineoblastomas in adults found that the major presenting symptoms are headache, blurred vision, nausea, and vomiting. Definitive common signs include papilledema, ataxia, and upward gaze palsy. The most relevant CT feature of pineal parenchymal tumors is peripheral
displacement of preexisting native pineal calcifications, a disease process referred to as the exploded pineal pattern. The presence of peripheral tumor calcification or central calcification (calcification produced by the tumor) may suggest a pineocytoma rather than a germinoma, as there is no calcification in pineoblastomas. Review of the reported features of 26 pineoblastomas found that pineoblastomas tend to be large, lobulated, and heterogeneously enhanced with infrequent calcifications. In addition, pineoblastomas present with a greater degree of hydrocephalus than do pineocytomas. CT of our case revealed a round, isodense tumor with peripheral calcifications and remarkable hydrocephalus.

MR imaging is extremely important for the detection of pineal region neoplasms and provides specific signal characteristics associated with pineoblastomas. MR imaging is also useful for the early detection of both intracranial and spinal seeding. Unfortunately, MR imaging experience of pineoblastomas is very limited, so the specific characteristics have not determined. Review of all 15 reported cases revealed that: T1-weighted imaging showed five hypointense, four hypo- to isointense, and six iso to hyperintense masses; and T2-weighted imaging showed one hypointense, one hypo- to isointense, two iso intense, six iso- to hyperintense, one hyperintense, and four mixed intensity masses. Administration of contrast medium showed six cases with heterogeneous and two with homogeneous enhancement. Therefore, pineoblastoma is indicated by hypointensity or hypo- to isointensity on T1-weighted images, isointensity or iso- to hyperintensity on T2-weighted images, and heterogeneous enhancement in the pineal region of young adults. In the present case of a young adult with a pineoblastoma, the pineal region neoplasm appeared as iso- to hypointensity on T1-weighted images, iso- to hyperintensity on T2-weighted images, and heterogeneously enhanced, and in agreement with previous reports.

The present case had an unusual extension into the fourth ventricle that nevertheless preserved the adjacent structures. Pineoblastomas tend to spread into the subarachnoid space and into the adjacent structures. Therefore, MR imaging study is useful for detecting the extent of tumors when decisions about initial treatment are made. This type of tumor normally becomes larger with local extension; inferiorly compresses the superior part of the vermis, tectum, and superior medullary velum; and can invade into the third ventricle. Pineoblastomas tend to be large (>4 cm), and generally infiltrate the splenium of the corpus callosum, tectum, tegmen-

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