Subarachnoid Hemorrhage Associated With Clival Chordoma

—Case Report—

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Abstract

A 66-year-old man presented with clival chordoma associated with subarachnoid hemorrhage. Computed tomography showed subarachnoid hemorrhage in the right ambient cistern and a well-enhanced tumor in the petroclival region. Surgical exploration performed on the day of admission showed a clot in the tumor. The tumor was totally removed. Histological examination showed hemorrhage between the tumor and the dura. The diagnosis was clival chordoma. Subarachnoid hemorrhage in chordoma seems to occur by spreading of intratumoral hemorrhage into the subarachnoid space due to dural invasion.

Key words: clival chordoma, subarachnoid hemorrhage, dural invasion

Introduction

Chordomas are uncommon, slowly-growing tumors that arise from notochordal remnants. Chordomas are characterized by their invasive nature and aggressive recurrence. Recurrence usually occurs within 3 years of treatment and the patients are likely to die soon thereafter. Both radical surgical removal and high-dose radiation therapy have been effective for tumor control.^{2,5)} Patients with clival chordomas typically present with a history of headache and progressive cranial nerve pareses of several months duration. Here we describe a case of chordoma which manifested as apoplectic syndrome secondary to the intracranial hemorrhage.

Case Report

A 66-year-old man complained of sudden onset of severe headache, vomiting, and mild disturbance of consciousness. He was a little drowsy but well oriented. No other neurological deficit was found. The platelet count and clotting time were normal.

Computed tomography (CT) of his head showed subarachnoid hemorrhage (SAH) in the right am-

bient cistern (Fig. 1A), but no other obvious signs of any other hemorrhage, for example intratumoral hemorrhage, and magnetic resonance (MR) imaging was not performed for diagnosis. Selective cerebral four-vessel angiography demonstrated a left carotid cave aneurysm measuring 5 mm in diameter, but no vertebrobasilar arterial aneurysm. The basilar artery deviated posteriorly (Fig. 2). CT after the angiography showed a well-enhanced tumor in the petroclival region (Fig. 1B).

Surgical exploration was performed on the day of admission through the combined supra- and infratentorial approach. A diffuse clot was found in the ambient cistern. The tumor originated in the upper clivus and invaded the dura and the basilar plexus. A massive clot was found in the tumor. After sufficient internal decompression, the tumor capsule was dissected from the sixth cranial nerve and the brain stem.

Gross examination found the tumor was gelatinous, grayish, and lobulated. Histological examination found fibrous strands dividing chordomas into lobules, which contained abundant mucin-containing tumor cells. Physaliferous cells with large intracytoplasmic vacuoles were seen

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Fig. 1 A: Computed tomography (CT) scan showing subarachnoid hemorrhage in the right ambient cistern. B: CT scan after angiography showing a well-enhanced tumor in the petroclival region.

Fig. 2 Left vertebral angiogram demonstrating the basilar artery deviating posteriorly (arrow).

(Fig. 3A). Hemorrhage was present not only in the tumor but also between the tumor and the dura (Fig. 3B). The histological diagnosis was typical clival chordoma.

Postoperative MR imaging revealed gross total removal of the tumor. Cranial nerve examination demonstrated transient left sixth cranial nerve paresis but no permanent neurological deficits. Recurrence occurred after 2 years 6 months of treatment. The recurrence was associated with hydrocephalus, so surgical removal and ventriculoperitoneal shunting was performed. Aggres-

Fig. 3 A: Photomicrograph showing fibrous strands dividing chordomas into lobules and characteristic physaliferous cells. HE stain, original magnification $\times 400$. B: Photomicrograph showing hemorrhage (arrow) between the tumor and the dura. HE stain, original magnification $\times 80$.

sive recurrence associated with hemorrhage in the temporal lobe occurred at 1 year 8 months after the second operation. Surgical treatment was performed five times in all for tumor control during 4 years 2 months.

Discussion

Only six cases of clival chordomas associated with intracranial hemorrhage have been reported.^{1,3,7-9)} Cases occurring before 1991 had a fatal outcome and no correct antemortem diagnosis was established. The cause of death was hemorrhage diffusing into the white matter tracts of the brain stem. The first case of clival chordoma associated with hemorrhage into the brain stem was reported in 1991.⁸⁾ The preoperative diagnosis was established using MR imaging. The patient survived. The present case is only the second of its type. Surgical exploration was performed in the acute phase and showed a clot in the tumor. Histological findings showed hemorrhage between the tumor and the dura.

Intratumoral hemorrhage in chordoma may result from rupture of the thin-walled vessels or hemorrhagic infarction due to rapid tumor growth. Hemorrhage in malignant gliomas occurs by various mechanisms.⁶⁾ Vascular endothelial proliferation with subsequent obliteration of the lumen of the vessels is a common cause of necrosis, resulting in hemorrhage into the tumor. Thin-walled, poorly

formed vessels in a tumor may also become distorted due to tumor growth, and such tumors may easily rupture and hemorrhage. Necrosis, with subsequent loss of vessel support, may also be a factor in the development of hemorrhages. Tumor erosion of vessels is another probable cause.

Two of three cases of chordomas demonstrated microscopic hemorrhages.⁴⁾ In contrast, clinical hemorrhage is rare. All six cases of clival chordomas associated with intracranial hemorrhage demonstrated an intraaxial component of the tumor. Therefore, the mechanism of clinical hemorrhage may also be associated with dural invasion. Tumor extension and invasion of the dura eventually caused mechanical destruction of the blood vessels surrounding the tumor. The mechanism of SAH seems to be the spread of the intratumoral hemorrhage into the subarachnoid space in association with dural invasion.

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