Extradural Temporal Meningioma Directly Extended to Cervical Bone

—Case Report—

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Abstract

A 42-year-old male presented with a rare extracranial meningioma manifesting as right facial swelling, facial nerve paresis, and hearing disturbance. Neurological imaging revealed a primarily extra-axial tumor extending from the temporal base to the subcutaneous space in the parietal region, and partially into the intra-axial region of the temporal lobe through the dura mater, with a small intracerebral hematoma and a small mass in the porus acusticus destroying the temporal bone, the floor of the middle fossa, zygomatic arch, and porus acusticus. Malignant meningioma was confirmed by histological examination after subtotal tumor removal *via* the transzygomatic approach. Subdural fluid accumulation like hematoma occurred 5 days after surgery and malignant cells were found in the fluid drained by skull trephination. Subsequently, radiation therapy with a total dose of 60 Gy was administered. One year later, he complained of head and neck pain with motor and sensory disturbances in the right upper extremity, and then developed tetraparesis. Cervical x-ray films demonstrated a compression fracture of the C3 vertebral bone and radiolucent areas at C2-4 levels, while computed tomography showed the tumor invading the subcutaneous space and compressing the spinal cord. He died of respiratory distress. Autopsy revealed that the tumor originated in the temporal region and had directly extended to the cervical region.

Key words: malignant meningioma, extracranial extension, cervical spine

Introduction

Meningiomas total about 15% of all primary intracranial tumors, but extracranial extension is rare. Possible mechanisms for extracranial meningioma have been proposed.^{6,12)} Meningioma involving the temporal bone extends extracranially in 43% of cases, most frequently through the jugular and lacerated foramina into the parapharyngeal area.¹²⁾

Here, we describe a patient with an extracranial malignant meningioma located in the middle fossa which extended directly to the cervical region, not through the foramina, causing a compression fracture of the cervical bone.

Case Report

A 42-year-old male was referred to our hospital with facial swelling and symptoms of right facial nerve paresis persisting for 8 months. He was admitted on January 4, 1990.

Neurological examination on admission revealed right peripheral facial nerve paresis, facial swelling, facial sensory disturbance (2nd and 3rd divisions of the trigeminal nerve), hearing disturbance on the right, and bilateral papilledema. Plain skull x-ray films demonstrated destructive changes in the temporal bone and zygomatic arch. Axial skull x-ray films showed disappearance of the right round, ovale, and lacerated foramina. Coronal plain skull tomograms demonstrated destruction of the middle fossa and destructive enlargement of the right porus acusticus internus (Fig. 1), but no enlargement of the jugular foramen. Technetium-99m bone scin-

Fig. 1 Preoperative coronal skull tomogram, demonstrating an enlargement of the porus and meatus acusticus internus (arrow).

Fig. 2 Preoperative coronal postcontrast CT scans, revealing a heterogeneously enhanced lesion with bony destruction extending from the middle fossa to the infratemporal fossa and parietal region.

tigrams showed abnormal accumulation at the temporal bone and zygomatic arch. Postcontrast computed tomographic (CT) scans showed a heterogeneously enhanced mass extending from the right temporal base to the parietal region with marked destruction and thinning of the temporal bone (Fig. 2). Magnetic resonance (MR) imaging demonstrated a tumor located mainly extradurally and extending partially into the intra-axial region of the temporal lobe through the dura mater, which appeared iso- or slightly hypointense on short SE and IR images, and iso- or slightly hyperintense on long SE images. The tumor was enhanced by gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA). MR images also showed a small tumor in the right cerebellopontine angle and a small intracerebral hematoma (Fig. 3). Right external carotid angiograms demonstrated an irregular hypervascular tumor shadow.

The right external carotid artery was artificially thrombosed with spongel preoperatively, then ex-

posed and ligated. The tumor was subtotally removed *via* the right transzygomatic approach on January 17, 1990. The tumor was located beneath the temporal muscle, involving the temporal bone and zygomatic arch extradurally, and extending to the pterygopalatine fossa through destruction of the bone of the middle fossa. A small incision of the dura mater found no tumor extension into the intradural space, and the cortical surface of the temporal lobe looked normal.

The postoperative course was uneventful. However, CT scans demonstrated right subdural fluid collection 5 days later, which was drained by trephination. The fluid resembled dark motor oil and looked like hematoma. Cytological examination discovered malignant cells. The region from the jaw joint to the C3 vertebra was irradiated with

Fig. 3 Preoperative coronal T₁-weighted Gd-DTPA-enhanced MR images, demonstrating the lesion mainly located extradurally and extending partially into the intra-axial region (arrow), with a small tumor in the cerebellopontine angle (arrowhead) and an intracerebral hematoma (double arrowhead).

Fig. 4 Follow-up coronal T₁-weighted Gd-DTPA-enhanced MR images, showing tumor recurrence in the right jaw joint (arrow).

a total dose of 60 Gy. Lumbar puncture on March 10, 1990 showed no malignant cells in the cerebrospinal fluid (CSF). He was discharged on March 14, 1990, and returned to work 1 month later.

In January, 1991, he complained of head and neck pain, difficulty in opening his mouth, and paresthesia in the right upper extremity. MR images on February 25 revealed no tumor recurrence in the intracranial space, but recurrence in the right jaw joint (Fig. 4). On March 4, he suddenly developed tetraparesis, and his general condition worsened. He was admitted to our hospital on March 7.

Neurological examination revealed tetraparesis, worse on the right than on the left, and hypesthesia in the bilateral upper and lower extremities. Plain x-ray films of the cervical spine showed a compression fracture at C3 and C4 levels and radiolucent areas at C2–4 levels, suggesting tumor invasion to the bone (Fig. 5). A Halo Vest (Toray Medical, Osaka) was immediately applied to maintain correct cervical alignment. CT scans demonstrated tumor invasion to the C2–4 vertebral bones and the extradural space of the C3–4 spinal canals, compressing the spinal cord (Fig. 6).

His condition deteriorated daily. Ten days after admission, he could no longer move his extremities. A second radiation therapy was considered, but not pursued because the C3 region was included in the field of the first radiation therapy. He died of respiratory dysfunction on March 16, 1991. Autopsy revealed that the tumor in the right jaw joint extended continuously to the cervical vertebral bones at C2–3 levels (the main lesion was C3), and that the tumor in the extradural space of the cervical region markedly compressed the spinal cord.

Fragments of surgical specimens and autopsy specimens of the cervical bone were embedded in

Fig. 5 Plain cervical radiogram on the second admission, demonstrating the compression fracture at the C3 level (arrowhead) and radiolucent areas at the C4 level (arrow) with disalignment of the cervical bone.

Fig. 6 Postcontrast CT scans at the C2 (left) and C3 levels (right), demonstrating an irregular heterogeneously enhanced lesion invading the spinal canal through the intervertebral foramen with destruction of the bones and compression of the spinal cord.

paraffin and stained with HE, phosphotungstic acid-hematoxylin, periodic acid-Schiff, Congo red, and the Watanabe method for silver impregnation. Immunohistochemical staining using vimentin, neurofilament, synaptophysin, epithelial membrane antigen, glial fibrillary acidic protein, keratin, S-100 protein, von Willebrand factor (VWF), and desmin were also performed. All specimens showed diffuse staining by vimentin and slight staining by VWF and S-100 protein. However, immunohistochemical stain-

Fig. 7 Photomicrographs of the operative specimen, showing meningioma with atypical cells and marked mitoses. HE stain, $\times 50$.

ing gave no useful indications. Both specimens had the same histological appearance. The tumor was highly cellular: cells were atypical and fibrous, spindle- or oval-shaped, with increased mitotic activity and an invasive proliferative appearance. Cells also showed syncytial and whorl formation, although not complete (Fig. 7). The tumor appeared sarcomatous findings in most parts, but partial findings of meningioma resulted in the final diagnosis of anaplastic meningioma.

Discussion

This case included several rare and interesting findings in the meningioma: 1) the histology of malignant meningioma, 2) the tumor was located mainly in the extradural region, 3) the tumor directly extended to the cervical region and caused depressed fracture at C3 level, 4) the CT and MR imaging findings of malignant nature of the tumor, and 5) the possibility of CSF metastasis and the formation of the subdural hematoma; malignant cells were confirmed in the subdural hematoma after surgery.

2–10% of meningiomas are malignant. The histological criteria for malignant meningioma are high mitotic rates, high cellularity, brain infiltration, and poor differentiation. The most striking common characteristic is the loss of the typical architectural pattern.¹⁾ In our case, these histological findings were confirmed, although the infiltration to the brain was only shown on CT scans and MR images. The diagnosis of meningioma was confirmed by various stains, and dural metastasis of cancer in systemic organs such as the breast, lung, or prostate eliminated.

The unusual location calls into question the origin

of this meningioma. Meningiomas generally arise from fragments of the arachnoid or embryonal arachnoid cell rests which occur extradurally or rarely in the intraosseous region.^{7,13)} The MR images and CT scans showed that this meningioma originated within the temporal bone, particularly the anterior surface of the petrous bone or within the petrous pyramid. Meningiomas within the petrous pyramid originate in: 1) the meatus acusticus internus, 2) the jugular foramen, 3) the geniculate ganglion region, and 4) the sulcus of the greater and lesser superficial petrosal nerves.¹²⁾ In our case, the origin of the meningioma may have been the sulcus. The extensive destruction of the temporal and cervical bones suggests a strong affinity for bony structures, and may be a feature of meningioma originating from arachnoid cells in a bony structure.

The most common route for extratemporal extension is through the jugular and lacerated foramina. Other routes, as in our case, are through the squamous portion of the temporal bone into the temporal fossa, through the zygoma into the preauricular region, and through the mastoid cortex into the postauricular area.¹²⁾

Extracranial meningioma may form in four ways: direct extracranial extension, extracranial growth from the arachnoid within the cranial nerve sheaths, extracranial growth from an ectopic or embryonic arachnoid cell rest, and distant metastasis from the intracranial meningioma.^{6,12)} In the present meningioma, extracranial extension was direct.

CT and MR imaging are very helpful in the diagnosis of extracranial meningiomas. CT achieves superior definition of bone and tumor, while MR imaging has no artifacts due to the surrounding bony structure, teeth, or other materials, while the multiplanar image options achieve easy detection of any bone replacement by tumor. Therefore, MR imaging is the most useful diagnostic method for the evaluation of extracranial meningiomas.⁵⁾ In this case, CT scans demonstrated signs of malignant meningioma, including moderate enhancement post-contrast compared to benign meningioma,²¹⁾ hypodense lesions within the tumor,²¹⁾ irregular border (including infiltration to brain parenchyma),^{4,14,21)} osteolysis by bone involvement,¹⁹⁾ absence of calcifications,²¹⁾ and the "mushrooming pattern."¹⁴⁾ MR imaging also showed an intracerebral hematoma, which may be a sign of a malignant tumor.

MR imaging showed a tumor shadow at the porus acusticus internus, possibly due to CSF metastasis of the malignant meningioma, as malignant cells were found in the subdural fluid accumulation. However, direct extension of the tumor through the

meatus acusticus internus is also possible because the meatus acusticus internus was enlarged. Subdural fluid accumulation, which looked like chronic subdural hematoma, occurred 5 days after surgery, and is very rare. The mechanism of formation of the subdural hematoma in this case is same as that caused by dural metastasis from systemic cancer: bleeding or hemorrhagic effusion secondary to either an occlusion of the dural capillary bed with tumor cells or a nodular deposit in the dura mater.^{9,10,20)}

The first choice for the treatment of meningioma is surgical removal, and postoperative radiation therapy when the meningioma is malignant^{2,8)} or incompletely removed.^{11,18)} However, radiation therapy to avoid recurrence of incompletely removed benign meningioma is controversial.^{2,8,15,17)} In our case, the brain from the jaw joint to C3 was irradiated. However, the tumor recurred in the irradiated area 1 year later, so the meningioma may have been radioresistant.

Various adjuvant therapies for meningioma have been reported. Recently, radiation therapy combined with administration of 2-deoxy-D-glucose has achieved improved results.³⁾ The role of hormone receptors in reducing the growth rate of meningiomas has also been investigated.¹⁶⁾ However, hormone therapy is not established at present. Malignant meningioma, incomplete removable meningioma, and recurrent meningioma require a more effective therapy than is currently available.

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