Total Removal of an Intramedullary Meningioma of the Medulla Oblongata

—Case Report—

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Abstract

A 12-year-old boy had central von Recklinghausen's neurofibromatosis with multiple intracranial meningiomas and bilateral acoustic neurinomas. One of the meningiomas was found in the parenchyma of the medulla oblongata and was successfully removed. To our knowledge, this is the first reported case of surgical removal of an intramedullary meningioma of the medulla oblongata. Details of the clinical, neuroradiological, and operative findings are described.

Key words: intramedullary meningioma, medulla oblongata, neurofibromatosis, childhood, brain neoplasms

Introduction

Although intracranial meningiomas in children are rare,^{6,15,26)} meningiomas are relatively common among children with central von Recklinghausen's neurofibromatosis.^{7,21)} Recently, we encountered the extremely rare instance of neurofibromatosis with multiple meningiomas and bilateral acoustic neurinomas. One of the meningiomas was entirely embedded in the parenchyma of the medulla oblongata and was successfully removed. We believe this to be the first reported case of surgical removal of an intramedullary meningioma of the medulla oblongata.

Case Report

This 12-year-old male patient was the younger of uniovular twins. The older twin had multiple meningiomas — a frontotemporal convexity meningioma, a foramen magnum meningioma, and a choroid plexus meningioma in the right lateral ventricle —

and bilateral acoustic neurinomas. Also, the older twin was blind in the right eye because of congenital maldevelopment of the papilla. There were no other known abnormalities in his family. The patient was admitted to Ehime University Hospital on February 22, 1985 for evaluation of intracranial lesions, although his only complaint was blindness in the right eye.

He was fully active and had no neurological abnormalities. He was blind in the right eye as a result of congenital maldevelopment of the papilla. There were several unpigmented cutaneous nodules in the right hypochondrial region, and biopsies of these nodules yielded a histological diagnosis of neurofibromatosis. Routine blood studies and chromosomal analysis were normal. Computed tomography (CT) showed moderately high density lesions with homogeneous enhancement, one in the right frontal parasagittal region (4.8 × 4.5 cm) and another in the region of the foramen magnum (1.7 × 1.3 cm) (Fig. 1). The location of the latter was accurately demonstrated on coronal and sagittal reconstruction

Received June 11, 1986; Accepted April 6, 1987

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Fig. 1 CT scan showing moderately high density lesions with homogeneous enhancement. *left*: A 4.8×4.5 cm mass in the right frontal parasagittal region. *right*: A 1.7×1.3 cm mass in the region of the foramen magnum.

Fig. 4 *left*: Right carotid angiogram shows tumor staining of the right middle meningeal artery in the right parasagittal region. *right*: Vascularity in the region of the foramen magnum is not abnormal, on vertebral angiogram.

Fig. 2 Coronal (*upper*) and sagittal (*lower*) reconstruction views accurately showing the location of the tumor in the region of the foramen magnum and indicating an intramedullary tumor of the medulla oblongata.

Fig. 3 *left*: A calcified lesion is seen in the left lateral ventricle, suggesting an intraventricular meningioma. *right*: The bilateral internal acoustic meatus are markedly dilated, suggesting bilateral acoustic neurinomas.

views, which indicated that it was an intramedullary tumor of the medulla oblongata (Fig. 2). A calcified lesion was also found in the left lateral ventricle. In addition to these multiple meningiomas, there were slightly dense, enhanced lesions in the bilateral, dilated internal acoustic meatus, suggestive of acoustic neurinomas (Fig. 3). Angiograms showed tumor staining of the right middle meningeal artery in the right frontal region, but no abnormal vascularity in the region of the foramen magnum (Fig. 4).

The preoperative diagnosis was multiple meningiomas (right parasagittal meningioma and choroid plexus meningioma of the left lateral ventricle), intramedullary tumor of the medulla oblongata, and bilateral acoustic neurinomas associated with von Recklinghausen's neurofibromatosis.

First operation: On March 18, 1985, a suboccipital craniectomy with a laminectomy of the C1 was performed through a midline skin incision with the patient in a prone position. The exposed dura mater over the cerebellum and the upper cervical spinal cord appeared normal and had good pulsation. The dura mater was opened with a Y-shaped incision. The medulla oblongata was markedly swollen, with a normal surface, which suggested an intramedullary tumor (Fig. 5). We separated and retracted the cerebellar tonsils and found the caudal portion of the floor of the fourth ventricle close to the obex to be bulging, with dark red discoloration. However, the surface of the ventricular floor appeared to be completely covered by an ependymal layer. The choroid plexus of the fourth ventricle did not adhere to the bulging floor. The medulla oblongata was incised to about 1.3 cm caudally from the obex on the midline, where we discovered a well demarcated, finely nodular tumor less than 2 mm from the surface of

Fig. 5 *left*: Intraoperative photograph showing the swollen medulla oblongata (MO) with a normal surface. The floor of the fourth ventricle (IV) looks normal in the left side of the picture. The arrow indicates the obex. *right*: Schematic drawing of the operative findings.

the medulla oblongata.

Having received the frozen section diagnosis of meningioma during the operation, we decided to totally remove the tumor, which was solid and encapsulated. Soft, gliotic tissue with numerous fine vessels separated the tumor capsule from the surrounding neural tissue, providing a plane for dissection of the tumor. While carefully monitoring cardiac function and blood pressure, we removed the tumor completely, piece by piece. The pathological diagnosis was meningotheliomatous meningioma (Fig. 6).

The patient awoke from anesthesia immediately after the operation. His respiration was somewhat shallow but regular and spontaneous, so the endotracheal tube was left in place for several hours postoperatively. By the following day, his respiration had recovered and his vital and neurological functions were all normal.

Second operation: We performed a right fronto-temporal craniotomy on May 2, 1985 and removed the entire parasagittal meningioma *en bloc*. The pathological diagnosis was meningotheliomatous meningioma.

The patient had mild hemiparesis following the second surgery, but it gradually improved and he was discharged on June 2, 1985 with no neurological deficits.

Discussion

Intracranial meningiomas in children are rare. Ingraham and Matson¹⁵⁾ found only three meningiomas (0.4%) among 750 intracranial tumors in children under 14 years of age. Taptas,²⁶⁾ in a review,

Fig. 6 Photomicrographs of the intramedullary tumor of the medulla oblongata. The tumor consists of poorly defined oval or spindle-shaped cells. The nuclei are relatively large and spheroidal. The tumor cells are arranged in sheets and are divided by a small amount of connective tissue (*upper*). There are interlacing bundles of long spindle-shaped cells (*lower*). HE stains, $\times 200$.

reported that there were 19 meningiomas (1.1%) among 1760 brain tumors in children. Childhood meningiomas often arise in association with von Recklinghausen's neurofibromatosis. Crouse and Berg⁷⁾ reported three meningiomas associated with von Recklinghausen's neurofibromatosis among 13 children and adolescents with primary meningeal tumors. Merten *et al.*²¹⁾ discovered, in data collected from medical centers in the United States and Canada, 11 cases of neurofibromatosis among 48 children and adolescents with meningiomas. Of particular interest is that five of the six cases of multiple meningiomas in their series were associated with neurofibromatosis. Rodriguez and Berthrong²⁴⁾ reviewed 49 cases of von Recklinghausen's neurofibromatosis with single or bilateral acoustic neuromas and multiple meningiomas and found that six of 45 patients (13.3%) were in the second decade, the youngest being 16 years old. Forty of 49 pa-

tients (81.6%) had bilateral acoustic neurinomas, 26 of 48 (54.2%) had intracranial meningiomas, 20 (41.7%) had intracranial and intraspinal meningiomas, and eight of the last group had intraventricular meningiomas in addition to the intracranial and intraspinal meningiomas.⁸⁾

Our patient had von Recklinghausen's neurofibromatosis and multiple meningiomas: right frontal parasagittal and left intraventricular meningiomas, and an intramedullary meningioma of the medulla oblongata associated with bilateral acoustic neurinomas. The intraventricular meningioma and neurinomas were not, however, histologically verified. To our knowledge, there has been no previous report of surgical excision of an intramedullary meningioma of the medulla oblongata in patients with or without von Recklinghausen's neurofibromatosis.

According to Lichtenstein,¹⁸⁾ the basic pathological lesions of von Recklinghausen's neurofibromatosis are foci of hyperplasia and neoplasia of the supportive derivatives of the primitive ectoderm in association with hyperplasia of mesodermal elements.^{4,10,18,24,27)} When meningiomas occur, they are commonly multiple and adherent to the inner aspect of the dura mater. They frequently occur in the choroid plexus. Focal areas of hyperplasia or neoplasia are also observed in the cerebrospinal arachnoid. In our patient, it was difficult to identify the origin of the tumor, since it had already grown quite large. It was entirely contained in the lower medulla oblongata, the dorsal surface of which was markedly swollen and thinly covered with brain parenchyma. The bulging caudal floor of the fourth ventricle had a normal ependymal layer. During its dissection the tumor adhered strongly to the surrounding neural tissue. Its feeding and draining vessels were just under the obex, which is perhaps where tumor originated. It likely arose from the inferior tela choroidea or from the meningeal structures around the obex and developed in the parenchyma of the medulla oblongata.^{1,5,25,28)}

Surgical intervention in patients with brain stem tumors is controversial. However, since the introduction of steroid therapy and the operating microscope, some neurosurgeons have biopsied such tumors and used the histological diagnosis to decide the appropriate irradiation therapy.^{2,9,14,23)} Also, many patients have been successfully treated for large neoplastic cysts in the brain stem by surgical evacuation followed by radiation therapy.^{11,16,17,19,22)} In our clinic, the primary treatment for brain stem tumors has been radiation therapy, except when the neuroradiological findings indicate benign, well demarcated tumors or cystic lesions. In the patient

reported here we totally removed an intramedullary meningioma from the medulla oblongata without causing neurological deficits. The tumor mainly involved the lower and probably the dorsal portions of the medulla oblongata. There was a soft, gliotic interface between the tumor capsule and the functionally important brain parenchyma; thus, a dissecting plane was easily found with the aid of a surgical microscope and the small vascular adhesions were safely cut with microsurgical instruments.^{12,13,20)} For these reasons we did not encounter abnormal changes in vital functions during or after surgery.³⁾

We conclude that this surgical approach can be safely attempted in patients with brain stem tumors, provided the tumor is well demarcated and mainly involves the lower medulla oblongata, preferably dorsal to the central canal.

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