

## A Case of Medullomyoblastoma of Cerebellopontine Angle Mimicking Acoustic Neuroma

Sang-Yoo Park<sup>1</sup>, Jin Hyung Kim<sup>1</sup>, Ki Taek Kim<sup>1</sup>, Yoo Jung Kim<sup>1</sup>, Tae Hwan Kim<sup>1</sup>, Keum Hwang<sup>2</sup>,  
Ki Jun Sung<sup>3</sup>, and Kwang Hwa Park<sup>4</sup>

*Department of <sup>1</sup>Otolaryngology-Head and Neck Surgery, <sup>2</sup>Neurosurgery, <sup>3</sup>Radiology, and <sup>4</sup>Pathology, Yonsei University Wonju College of Medicine, Wonju, Korea.*

Medulloblastoma is a common malignant central nervous system neoplasm found mainly in children. On the contrary, medulloblastoma of the cerebellopontine angle, the location of the tumor is very unusual. This is the first case of the medullomyoblastoma, a rare form of medulloblastoma, occurring in the cerebellopontine angle.

A 15-year-old boy experienced a sudden hearing loss in the left ear. Conservative medical treatment failed, and temporal MR imaging revealed a heterogeneously enhancing mass at the left cerebellopontine angle cistern and in the internal auditory canal; therefore, the lesion was regarded as a typical acoustic neuroma. Few days before surgery, an ipsilateral facial palsy developed, and a follow-up MR imaging showed a rapid growth of the previous lesion. The extended translabyrinthine approach permitted surgical removal. And under pathological diagnosis of malignancy, radiation therapy and series of chemotherapy was performed.

**Key Words:** Medullomyoblastoma, cerebellopontine angle

### INTRODUCTION

Medullomyoblastoma, a unique variant of medulloblastoma, is known as a cerebellar tumor found predominantly in children. It has the histological characteristics of a medulloblastoma, within which smooth and striated muscle fibers are found.<sup>1</sup> The term medullomyoblastoma was first coined by Marinesco and Goldstein in 1933

in their report describing the two histological components of these tumors: a medulloblastic component and skeletal muscle.<sup>2</sup> The muscle elements usually present a malignant appearance.<sup>1</sup> Until 1990, only 22 cases of medullomyoblastoma had been reported in the literature.<sup>3</sup> Its typical location is the cerebellar vermis, and it often causes rapid progressive symptoms and has poor outcomes. In our review of the literature, no previous case of the medullomyoblastoma existed on the cerebellopontine angle. Thereby, we present this case of medullomyoblastoma, mimicking clinical features of the acoustic neuroma.

### CASE REPORT

A 15-year-old boy visited the OPD with a history of hearing difficulty in the left ear for 3 weeks. He was treated at another otology practice clinic on the diagnostic basis of unilateral sudden sensorineural hearing loss (SSNHL), but conservative medical management did not improve hearing but rather worsened it. The otoscopic findings of both ears were normal. Audiological tests revealed that he was deaf in the left ear, and by the bithermal caloric test, canal paresis (52%) was seen in the left ear. However, he did not complaint of any symptoms like vertigo or imbalance. Temporal magnetic resonance imaging revealed a heterogeneously enhancing mass located on the left cerebellopontine angle cistern and in the internal auditory canal (Fig. 1A-C). Otherwise, his examination was unremarkable. At the same time, through present symptoms and

Received August 18, 2003

Accepted December 26, 2003

Reprint address: requests to Dr. Sang-Yoo Park, Department of Otolaryngology-Head and Neck Surgery, Wonju College of Medicine, Yonsei University, 162 Ilsan-dong, Wonju, Kangwon-do 220-701, Korea. Tel: 82-33-741-0642, Fax: 82-33-732-8287, E-mail: sangyoo3@wonju.yonsei.ac.kr



**Fig. 1.** Pre-operative Imaging. (A) Axial T1-weighted Gd enhanced image reveals a heterogeneously enhancing mass within in the left CPA cistern and IAC. (B, C) Axial T1-weighted images show slightly hypo-intense mass compared with the gray mass. (D) Axial T1-weighted enhanced image shows a ice-cream cone (arrows) shaped irregularly enhancing the extra-axial mass in the left CPA cistern and IAC. The marked enlargement of the mass compare with previously noted. (E) 3D FSE T2-weighted image shows a heterogeneously hypo-intense mass. A slight compression of the brain stem (arrows) and the left brachium pontis is suggested. The canalicular component fills the entire internal auditory canal. (F) Axial CT shows the widening of the left internal auditory canal.

imaging results, the authors made a diagnosis of the acoustic schwannoma, and early surgical operation was planned.

Then, facial paralysis on the left side (House-Brackmann grade III, 27.6% denervation ratio by ENG study) developed a few days before surgery. A follow-up MRI showed a rapid growth of the mass (Fig. 1D) found to be the result of bleeding hematoma or possible malignancy. Angiographic evaluation was done for tumor vascularity. It was not hypervascular mass. Surgical removal was performed on the mass by the extended trans-labyrinthine approach. The tumor had a soft, reddish and easy-bleeding nature (Fig. 2). A sharp dissection was difficult because the mass adhered to the nerve bundles of the internal auditory canal. The tumor mass with the involved audio-vestibular nerve was carefully extirpated, except the high-risk area attached to the pons, a  $0.7 \times 0.7$  cm sized remnant.

Histopathologically, the specimen was compatible with the medulloblastoma, and it also showed smooth and striated muscle fibers within

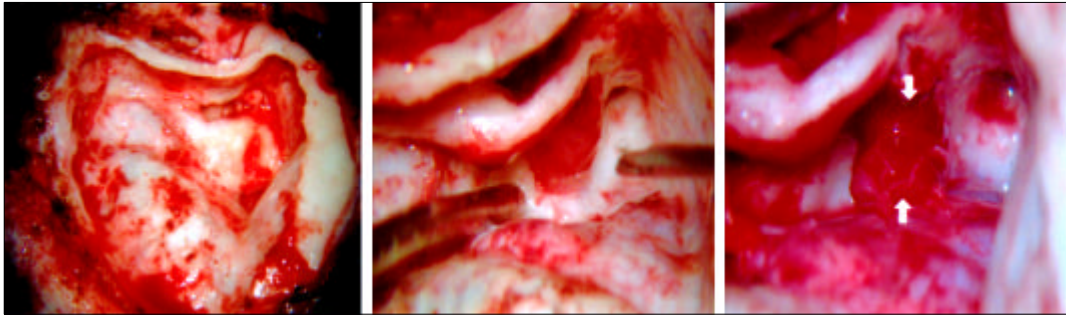
(Fig. 3). In immunohistochemical studies, the tumor cells were found to be positive for the desmin stain (Fig. 4). In addition, the immunohistochemical stain for myoglobin is also positive (Fig. 5). So the final diagnosis of the medulloblastoma was made. The patient was discharged without notable complications and with post-operative radiation therapy of a total of 5400 cGy on the primary site and a total of 3060 cGy on the craniospinal irradiation was done. After 6 series of chemotherapy with vincristine, CDDP and CCNU, the patient is now on the remissive state for one year post-operatively, and the authors are considering the appropriate facial reanimation procedure.

## DISCUSSION

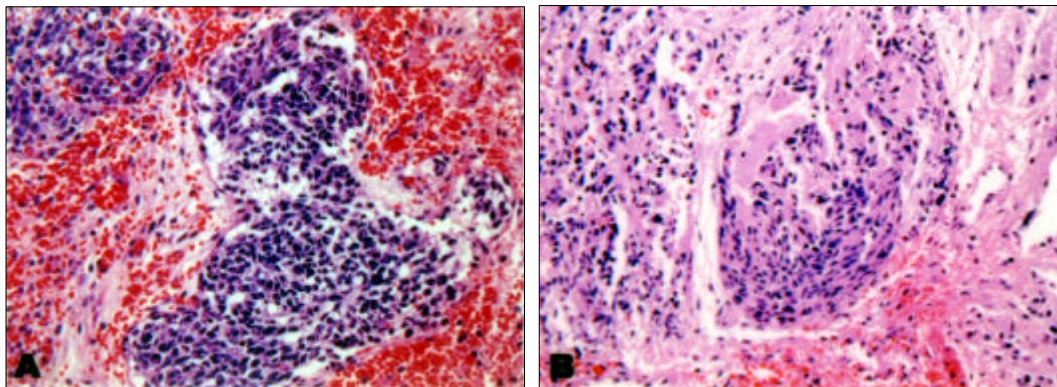
The medulloblastoma is a rare cerebellar tumor, a unique variant of the medulloblastoma. The histogenesis of the myogenic component of the medulloblastoma has been a subject of

controversy. Russell and Rubinstein, Misugi and Liss, and Walter and Brucher proposed the hypothesis that this tumor should be regarded as

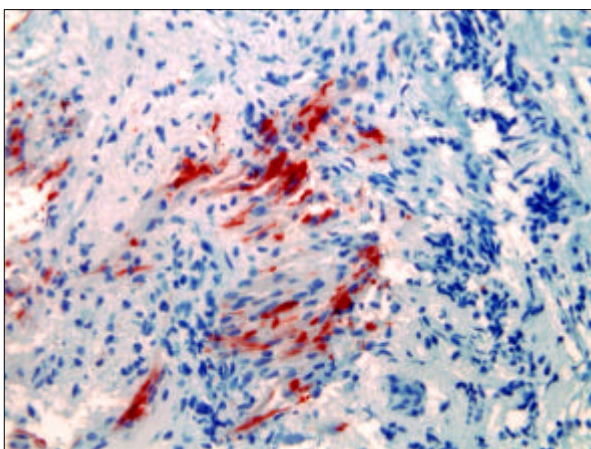
a teratoma, consisting of a neuroectodermal element as medulloblastoma and mesenchymal rhabdomyosarcoma component. The second hypo



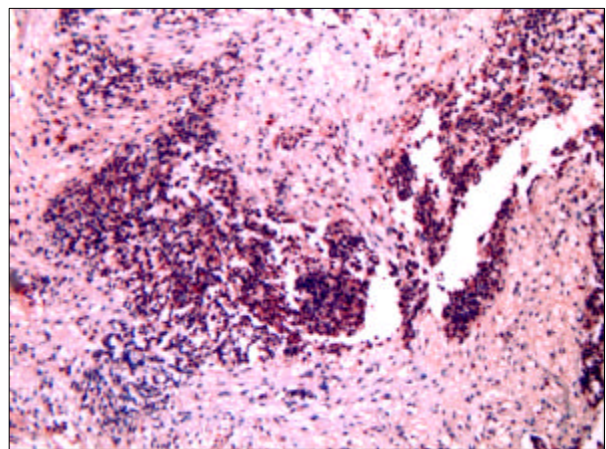
**Fig. 2.** Operative photograph shows a soft reddish mass of about  $0.7 \times 0.5 \times 0.5$  cm filling the internal auditory canal. (arrows)



**Fig. 3.** (A) There are solid sheets of anaplastic cells showing scanty or multi-focally vacuolated cytoplasm and hyperchromatic nuclei in the hemorrhagic background (H&E,  $\times 200$ ). (B) Some cells have relatively abundant eosinophilic cytoplasm and eccentrically located nuclei that showing rhabdomyoblastic differentiation (H&E,  $\times 200$ ).



**Fig. 4.** The tumor cells were found to be partially positive for the desmin. (Immunohistochemical stain,  $\times 400$ )



**Fig. 5.** The immunohistochemical stain for myoglobin is positive. (Immunohistochemical stain,  $\times 200$ )

thesis is based on the explanation put forward by Willis, saying that the myoblastoma component originates from the highly plastic embryonal pluripotential mesenchymal cells present in or near the tumor. The third hypothesis describes that the myogenic component derive from the multipotential endothelial cells, based on the ultrastructural similarities between endothelial cells and undifferentiated myogenic cells. The last hypothesis is based on the concept that primitive neuroectodermal cells are pluripotential and possess myogenic capabilities.<sup>1,3,4</sup> The pathological diagnosis of the medulloblastoma was based on the presence of a densely cellular primitive neuroectodermal component. It contained less-cellular areas in which striated muscle fibers were found.<sup>5</sup>

Biological behaviors, recurrence rates, clinical characteristics and metastatic pattern of the medulloblastoma are probably the same as those of the classical medulloblastoma.<sup>1</sup> Its typical clinical characteristics on the CPA are headaches, hearing loss, facial nerve paralysis and cerebellar signs. The clinical symptoms before diagnosis usually last for a short period from a few weeks to 3 months. Regarding the differential diagnosis of the cerebellopontine angle lesions, one must consider the most common lesion that occurs there, namely, the acoustic neuroma which extends from the internal auditory canal into the cerebellopontine angle.<sup>5</sup> Other primary cerebellopontine angle lesions, such as cholesteatoma, meningioma, and arachnoid cyst, may occur. Only 15 cases of CPA medulloblastoma have been reported.<sup>6</sup> There seems to be no clinical, neuro-otological, or neuroradiological findings peculiar to CPA medulloblastomas. Several features, however, may help to separate them from other CPA tumors, especially from the most common acoustic neuroma. The diminution of hearing or 7<sup>th</sup> nerve involvement is uncommon in medulloblastoma on CPA. Only two of the 15 reported cases, presented with diminution of hearing as the initial symptom. These symptoms may help to differentiate the medulloblastoma from acoustic schwannoma of the same size. Because of these subtle clinical signs, the diagnosis of CPA medul-

loblastoma or medulloblastoma is difficult.

The current treatment of choice for the cerebellopontine angle medulloblastoma is considered to be a surgical removal followed by post-operative irradiation.<sup>7</sup> Chemotherapy is indicated in cases where there is tumor recurrence or dissemination of tumor in the cerebrospinal fluid.<sup>8</sup> Treatment combining surgery and post-operative irradiation offer a 30% chance of a five-year survival.<sup>3,4,7</sup> In case of the medulloblastoma, total excision followed by radiation seems to be equally useful treatment. However, the survival rate for patients with medulloblastoma is lower than with medulloblastoma. Rao et al. reviewed the literature up to the 1990 and found that 10 of 21 patients lived for shorter than one month. And, only 5 of 21 patients having survived for longer than 6 months.<sup>1,3</sup>

The medulloblastoma of the cerebellopontine angle is very rare and reported about 15 cases in the literature. This is the the first case of the medulloblastoma, a rare form of medulloblastoma, occurring in the cerebellopontine angle.

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