Acoustic neuroma with malignant transformation

Case report

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The authors describe the case of a 57-year-old woman who had a right-sided hearing disturbance that had remained untreated for 1 year. The diagnosis was of a right cerebellopontine angle tumor, and the patient underwent its removal via retrosigmoid approach. Pathologically, the tumor was a benign neuroma. Growth of residual tumor was detected 4 years after the initial operation, and it was treated with gamma knife surgery (GKS). Six months later, the tumor had grown, and the patient underwent surgery via a combined retrosigmoid–translabyrinthine approach. Abnormal mitotic figures were observed on histological studies, indicating that the tumor had become malignant. Thereafter, the tumor grew rapidly, and the patient died 6.5 years after the initial treatment. It cannot be ruled out that GKS affected the outcome, but the causal sequence was unclear. Because such a patient is rare, documentation of the case was considered clinically important.

KEY WORDS • acoustic neuroma • malignant transformation • gamma knife surgery • radiation complication

Malignant cranial nerve tumors are very rare, and most often occur in the fifth cranial nerve, followed in frequency by the seventh and eighth cranial nerves.2-4,7-10 Although malignant peripheral nerve sheath tumors are often observed as complications of von Recklinghausen disease, such complications are rare in cranial nerve tumors.5 Acoustic neuroma is usually benign, except for patients in whom this disease is accompanied by neurofibromatosis; malignant transformation and recurrence has been reported by McLean, et al.,11 in only one patient. We encountered a patient in whom the acoustic neurinoma was benign at onset, but it later became malignant, as confirmed by pathological studies, and then grew rapidly. We discuss this case in connection with the use of GKS and also review the literature.

Case Report

History and Examination. This 51-year-old woman had no contributory anamnesis and no family history of von Recklinghausen disease. She presented at our hospital with a chief complaint of progressive hearing disturbance in the right ear, which had remained untreated for approximately 1 year. Neurological examination demonstrated that the patient had hearing loss in the right ear. A tumor with a clear boundary exhibiting uneven high-intensity signals was observed in the right cerebellopontine angle on T1-weighted Gd-enhanced MR images (Fig. 1 left).
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enolase–positive and glial fibrillary acidic protein–negative on immunohistological staining. The patient underwent surgical connection of the right facial and hypoglossal nerves 2 months after the second surgery and was ambulatory on discharge. A second GKS (lesion volume 9.2 ml, peripheral dose 14 Gy [50%], maximum core dose 28 Gy) was targeted to the residual tumor in July 1996.

Postoperative Course and Outcome. The patient was hospitalized 6 months after the second GKS because of difficulty walking, and she experienced gradual aggravation of dizziness and unsteadiness. Neurologically, the patient scored 15 on the Glasgow Coma Scale and had paralysis of the right third to 12th cranial nerves, reduction of muscular strength on the left side, and poor coordination on the right side. The irregular tumor observed on MR images had grown rapidly and was clearly pressing on the brainstem (Fig. 3). The patient’s consciousness level gradually declined, and tumor excision was performed twice to save her life. Despite these efforts, she died in May 1997, 6.5 years after the initial treatment.

Postmortem Examination. On macroscopic observation, we found that the tumor extended to the entire brainstem, onto the tentorium, and to the cerebral ventricles; marked necrosis was observed at autopsy. Microscopically, necrosed lesions with pseudopallisading, pleomorphic nuclei, and abnormal mitotic figures were observed (Fig. 4). The tumor was diagnosed as an acoustic neuroma that had undergone malignant transformation (sarcomatous change).

Discussion
Schwannoma is generally benign and rarely becomes malignant. In 1994, Woodruff, et al., summarized the cases of nine patients with schwannoma that became malignant. Of those nine, schwannoma in the skull occurred only in one patient, who had been reported on by Franks in 1985. This case was not a neuroma but an epithelioid neurilemmoma in the trigeminal nerve. Malignant cranial nerve tumor occurs most often in the fifth cranial nerve,
followed in frequency by the seventh and eighth cranial nerves. Malignant peripheral nerve sheath tumors are often observed in conjunction with von Recklinghausen disease, but a tumor of the cranial nerve is rare. Malignant nerve sheath tumor of the eighth cranial nerve has been reported in only six patients. Of these six, four had malignant triton tumor, and one had malignant triton tumor with neurofibromatosis Type 2. Malignant triton tumor, which is a variant of malignant nerve sheath tumors, is a rare soft-tissue sarcoma with rhabdomyoblastic differentiation. This tumor generally occurs in the peripheral nerve, and approximately 70% of these tumors are related to von Recklinghausen disease.

In our patient, no rhabdoid elements characteristic of triton tumor were detected on the first pathological examination; the tumor was a typical benign acoustic neuroma with both Antoni type A and type B areas. McLean, et al., reported on a rare case in which the patient had recurrent tumor with malignant transformation 11 months after excision of the entire acoustic neuroma. To our knowledge, that is the only case reported before ours of a patient who demonstrated recurrent acoustic neuroma with malignant transformation.

In our patient, the tumor regrew 4 years after the first surgery, and GKS was then performed. It is therefore necessary to evaluate the relationship between transformation and GKS. Stereotactic radiosurgery has been shown to be an effective alternative to microsurgical removal of most small- and medium-sized acoustic tumors. Tumor control rates average 93 to 98%. Anniko, et al., performed histological examinations of acoustic tumor cells in tissue cultures that were exposed to single doses of gamma radiation ranging from 30 to 150 Gy. Histological analysis of irradiated cells revealed early cytoplasmic vacuolization, followed by either early cell death or cell survival with electron-dense intracytoplasmic inclusion bodies. No evidence of malignant transformation was noted. Linsky and colleagues carefully studied the histological features of human acoustic tumor xenografts subjected to stereotactic radiosurgery. In their studies, irradiated tumor tissue revealed areas of blood vessel wall hyalinization and scattered foci of hemosiderin not seen in control specimens. No evidence of increased cellular atypia or malignant transformation was observed.

Patients with eighth cranial nerve sheath tumor that became malignant after radiosurgery have been reported, although such cases are very rare. At the meeting of the Stereotactic Radiosurgery Society held at Madrid, Spain in 1997, Comey, et al., reported that a tumor became malignant 6 years after adjuvant radiosurgery of residual tumor following subtotal resection of a benign eighth cranial nerve sheath tumor, and the patient died 3 months later.

In our patient, mitotic figures were observed on pathological studies approximately 6 months after GKS, which may indicate that the tumor had become malignant. The transformation, however, would necessarily have been very rapid, compared with that in previously reported cases, for the malignancy to have been caused by the GKS. Although it cannot be ruled out that GKS affect-
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ed the disease course, it seems more likely that this patient was a very rare one in whom benign acoustic neuroma spontaneously became malignant.

References