Malignant intraventricular schwannoma

Case Report

JIN-MYUNG JUNG, M.D., HYUNG-JIN SHIN, M.D., JE G. CHI, M.D., IN SUNG PARK, M.D., EUN SANG KIM, M.D., AND JONG WOO HAN, M.D.

Department of Neurosurgery, Gyeongsang National University College of Medicine, Chinju, Korea; and Department of Pathology, Seoul National University College of Medicine, Seoul, Korea

The authors present the clinical, radiological, and pathological features of a malignant schwannoma occurring in the right lateral ventricle of a 40-year-old man. Metastasis to both cerebellopontine angles and to the cerebellum was found 7 months after subtotal removal of the tumor.

KEY WORDS • schwannoma • cerebellopontine angle • metastasis

Intracerebral schwannomas are uncommon, with 42 cases reported to date; only two of these cases were malignant. Intraventricular schwannomas are even rarer; six cases have been reported with no malignancies. We describe a case in which a malignant schwannoma was subtotally removed from the right lateral ventricle, with metastasis to both cerebellopontine angles and the cerebellum found 7 months later. The relevant literature is reviewed.

Case Report

This 40-year-old man was admitted to our department of neurosurgery in December, 1991, for evaluation of a bursting headache, vomiting for 20 days, and a slightly drowsy mentality which began a few days before presentation.

Examination. On admission, the patient was slightly drowsy and funduscopied revealed bilateral papilledema and retinal hemorrhage. There were no stigmata of neurofibromatosis. Computerized tomography (CT) showed a 5 × 5 × 6-cm cystic mass with peripheral edema and heterogeneous enhancement in the right posterior temporal area (Fig. 1). Magnetic resonance (MR) imaging on a 1.0-tesla system demonstrated slightly low signal intensity on T1-weighted images, mixed signal intensity on T2-weighted images, and considerable enhancement with gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA) (Fig. 2). Cerebral angiography revealed a hypervascular mass fed by the right anterior choroidal and right lateral posterior choroidal arteries.

First Operation. A middle temporal gyrus approach with subtotal tumor removal was performed in December, 1991. The mass was yellowish, firm, and rubbery, and bled easily. The tumor was relatively well circumscribed superficially, but not in the deeper portion. A ventriculoperitoneal shunt was placed on the 9th postoperative day because of entrapment of the temporal horn of the right lateral ventricle. Whole-brain irradiation was performed using 55 Gy.

Pathological Examination. Microscopically, the tumor cells were arranged in interlacing fascicles of spindle cells (Fig. 3 left). Intense cellularity and frequent mitotic figures were seen (Fig. 3 center). Reticulin staining revealed an extensive network of reticulin (Fig. 3 right). Glial fibrillary acidic protein and epithelial membrane antigen staining was negative; however, the tumor stained diffusely positive for S-100 protein and vimentin. Electron microscopy revealed spindle cells with long, thin, interwining processes, a moderate amount of common organelles, and distinct external laminae that were discontinuous (Fig. 4). On the basis of these findings, the tumor was diagnosed as a malignant schwannoma.

Second Operation. In March, 1992, the patient began to complain of mild headache and vertigo which slowly progressed. In July, 1992, he was readmitted for evaluation of severe headache, vertigo, and confusion. Tumor recurrence and metastasis to both cerebellopontine angles and the surface of the left cerebellar hemisphere were revealed on MR imaging (Fig. 5). Left suboccipital craniectomy with total removal of the cerebellar and left cerebellopontine angle masses was performed; however, the patient...
died of bacterial meningitis and fungal pneumonia on the 15th postoperative day. The pathological findings were the same as those of intraventricular mass.

Postmortem Examination. Examination revealed a hard recurrent mass in the right temporal lobe and two hard masses (1.7 × 1.8 × 1.4 cm and 1.0 × 0.9 × 0.3 cm) attached to the dura at the posterosuperior lip of the right internal acoustic meatus and superior lip of the right jugular foramen. No residual tumor was found in the cerebellopontine angle or at the surface of the cerebellar hemisphere. The microscopic findings of these metastatic nodules were the same as those of the main mass. No other metastatic focus could be demonstrated.

Discussion

Acoustic neurinomas are reported to demonstrate iso- to hypointensity on T₁-weighted MR images, hyperintensity on T₂-weighted images, and considerable enhancement on Gd-DTPA administration. In our patient, hypointensity was demonstrated on the T₁-weighted images and mixed signal intensity on the T₂-weighted images, with considerable enhancement by Gd-DTPA. These findings are very similar to those of previously reported cases of intracerebral schwannoma.¹⁻⁸

The origin of intraventricular schwannoma is still controversial. As far back as 1874, Benedict identified nerve fibers in the choroid plexus of the fourth ventricle, an observation that was confirmed by Stöhr.²⁹ Because the autonomic nervous system is known to include Schwann cells, it appears most likely that primary intraventricular schwannomas arise from the autonomic nerve supply of the choroid plexus.³⁰

Many electron microscopy studies clearly reveal intercellular collagen fibrils (Luse bodies) and the lamellar pattern of thin, elongated cell processes covered by a basal lamina and separated by basement membrane material.²⁴ However, these features are highly variable depending on the degree of differentiation of the neoplasm.²⁴ Bruner, et al.,³ reported a case of intracerebral malignant schwannoma that showed sparse basement membrane and abundant intermediate filaments. These findings were very similar to those in our case.

Intracerebral schwannoma is a rare event in which the majority of patients are between 6 and 26 years of age and are male.²⁸ With the exception of three cases,⁴⁻⁶,¹³ intracerebral schwannomas were not associated with neurofibromatosis. Malignant intracerebral schwannoma is even more rare, with only two reported cases. We describe a patient in whom we suspect that drop metastases occurred. Liwnicz¹⁸ reported a case of bilateral trigeminal neurofibrosarcoma, but the drop metastasis of malignant intracranial schwannoma is extremely unusual. Han, et al.,¹⁴ reported drop metastasis in the case of a malignant triton tumor of the acoustic nerve.

We believe that this is the first report of an intraventricular malignant schwannoma with drop metastasis. In our
Malignant intraventricular schwannoma

Fig. 3. Photomicrographs of tumor specimens in this study. Left: Tumor cells are arranged in interlacing fascicles of spindle cells. H & E, original magnification × 200. Center: High cellularity and frequent mitotic figures are seen. H & E, original magnification × 200. Right: Reticulin mesh surrounds individual tumor cells. Reticulin, × 133.

Fig. 4. Electron micrograph demonstrating tumor cells containing a moderate amount of common organelles and discontinuous external laminae. Collagen fibrils are seen in the stroma. Original magnification × 12,000.
case, the initial clinical features and appearance of the diagnostic image resembled those of previously reported intraventricular benign schwannomas, but the age and the rapid clinical course were somewhat different.

References


Fig. 5. Contrast-enhanced axial T1-weighted magnetic resonance images obtained 7 months after the first operation showing tumor recurrence in the right temporal area (left) and metastasis to the surface of the left cerebellar hemisphere (center) and both cerebellopontine angles (right).