Concurrent Cochlear Implantation with Resection of Skull Base Hemangiopericytoma following Sudden Deafness in an Only Hearing Ear

Evan R. Ransom, M.D.,¹ Kevin D. Judy, M.D.,² and Douglas C. Bigelow, M.D.¹

ABSTRACT

A 72-year-old man with a known left acoustic neuroma, left-sided deafness, and a recently diagnosed right infratemporal fossa (ITF) hemangiopericytoma, presented with sudden deafness in his right ear. Imaging revealed right-sided skull base extension and a large intracranial tumor component. The patient underwent a frontotemporal craniotomy with concomitant ITF approach. Complete tumor resection was possible, though invasion of the otic capsule was present. Immediately postresection, a cochlear implant (CI) was performed via a transmastoid approach. Full electrode insertion was achieved and confirmed by visualization through the dehiscent middle fossa floor. Mastoid obliteration was then performed with a free fat graft. Postoperative imaging confirmed complete tumor resection (Simpson grade I) and adequate CI placement. Follow-up magnetic resonance imaging was performed at 6 and 12 months, and no tumor recurrence was seen. Prior to CI activation, the patient was completely deaf bilaterally. At 18-month follow-up, however, excellent hearing was achieved with the right CI (16 of 22 electrodes active), and the patient is now conversational with no obvious deficit. His cognitive function is excellent, corresponding to preoperative status, and he is independent in his activities of daily living. Following adjuvant radiation, our patient remains disease free at 18 months.

KEYWORDS: Hemangiopericytoma, cochlear implant, infratemporal fossa, skull base tumor, sudden hearing loss

Hemangiopericytoma (HPC) is a rare and highly vascular soft tissue sarcoma first described by Stout and Murray in 1942.¹ These tumors are derived from pericytes, a specialized contractile cell beneath the vascular endothelial layer, which contribute to local blood flow regulation. HPCs represent only ~3% of

soft tissue sarcomas.¹⁻⁴ Approximately one quarter of HPCs occur in the head and neck region, though most of these are found in the nasal cavity or paranasal sinuses.⁵⁻¹⁰ HPC involving the skull base is very rare, with less than 15 cases reported in the literature, only two of which had significant intracranial extension.¹¹⁻¹⁶

Departments of Otorhinolaryngology, ¹Head and Neck Surgery and ²Neurosurgery, University of Pennsylvania, Philadelphia, Pennsylvania.

Address for correspondence and reprint requests: Douglas C. Bigelow, M.D., Associate Professor, Dept. Otorhinolaryngology, Head and Neck Surgery, University of Pennsylvania, 3400 Spruce Street, 5th Floor Silverstein Building, Philadelphia, PA 19103 (e-mail: douglas.bigelow@uphs.upenn.edu).

Skull Base 2010;20:279-284. Copyright © 2010 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.

Received: October 8, 2009. Accepted after revision: November 25, 2009. Published online: January 27, 2010.
DOI: <http://dx.doi.org/10.1055/s-0030-1247633>.
ISSN 1531-5010.

Hearing loss attributable to skull base HPC has been described in only two cases, both low-grade tumors occurring in the middle ear space.^{15,16}

Cochlear implantation (CI) at the time of tumor resection is well characterized in the case of acoustic neuroma, though there are few reports of concurrent CI following ablative surgery for other tumors.^{17–20} Here we present a case of invasive skull base HPC, extending from the preauricular soft tissues into the temporal bone and middle cranial fossa, and causing sudden deafness in a patient's only hearing ear. This lesion was resected via a combined temporal craniotomy and infratemporal fossa (ITF) approach. Hearing was rehabilitated simultaneously with CI.

CASE REPORT

A 72-year-old man with a history of unilateral deafness secondary to a previously diagnosed and untreated left vestibular schwannoma presented to an outside hospital for evaluation of new-onset imbalance and right-sided hearing loss. Hearing loss occurred suddenly, with a loud pop, while the patient was chewing. Evaluation at the outside institution resulted in transtympanic steroid injections for sudden sensorineural hearing loss (SSNHL); oral steroids were not prescribed secondary to concerns regarding the patient's diabetes. Hearing loss on the right side improved modestly, and the patient was referred to our institution for further diagnosis and treatment.

Initial evaluation revealed a history of resection of a right preauricular mass ~10 years prior, with unknown pathology. No chemotherapy had been given, though the patient remembered that radiation had been considered but eventually ruled out. Further medical records, including pathology and operative reports, were not available. In addition to the recent episode of worsening right-sided SNHL, the patient reported mild dizziness and imbalance. Physical exam was significant for a mild right facial palsy (House-Brackman 2/6). No cervical lymphadenopathy was present. Magnetic resonance imaging (MRI) of the head, neck, and brain confirmed the presence of right-sided inhomogeneously enhancing ITF mass, measuring $\sim 3.4 \times 3.1 \times 3.0$ cm, and extending from the right preauricular soft tissue through the temporal bone into the right middle fossa. No change from the previous MRI was appreciated in the left acoustic neuroma size. Computed tomography (CT) scan obtained at this time confirmed erosion into the superior aspect of the cochlea and the right temporomandibular joint (TMJ).

A plan for surgery and arrangements for preoperative medical evaluation were made. During this interval, however, the patient experienced a second episode of SSNHL on the right side, leaving him completely deaf. This was attributed to tumor erosion through the

otic capsule and was accompanied by worsening imbalance and right facial palsy (House-Brackman 3/6). Repeat MRI at this time showed tumor enlargement, now measuring $3.8 \times 3.1 \times 4.0$ cm, with further intracranial extension (Fig. 1). Cytotoxic edema was noted in the right temporal lobe, and there was mass effect at the brain stem along with moderate effacement of the right lateral ventricle. The patient was started on corticosteroids with some improvement in his balance only, and surgery was scheduled for 2 weeks later. Given the uncertainty of the tumor type, no preoperative embolization was performed.

The surgical approach selected was a temporal craniotomy from above in continuation with an ITF approach from below, as described by Fisch.²¹ This began with a modified "lazy S" incision, essentially a combination of a hemicoronal scalp incision and a postauricular otologic incision. A frontotemporal craniotomy with zygomatic arch osteotomy was performed, with careful preservation of the temporalis muscle, to access the intradural portion of the tumor. Microdissection was performed with bipolar cautery, and the arachnoid and pia mater of the temporal lobe were easily separated from the tumor mass. A frozen section was sent, which confirmed a diagnosis of malignant tumor, favoring HPC.

Resection of the intradural and intracranial-extradural portions of the tumor then proceeded with the use of a Cavitron ultrasonic surgical aspirator

Figure 1 Preoperative T1 postgadolinium magnetic resonance imaging, axial section at the level of the cerebral peduncles. Hyperintense, inhomogeneously enhancing, right-sided infratemporal fossa tumor is seen extending from the preauricular soft tissue, with erosion of the right temporal bone and extension to the middle cranial fossa.

(CUSA, Integra NeuroSciences, Plainsboro, NJ). This required drilling of the middle fossa floor and microdissection medially to the cavernous sinus. Medial dissection to the level of the internal auditory canal was performed. The petrous carotid was freed from its bony canal, and removal of the involved portions of the petrous bone was performed with a diamond burr. This resulted in communication of the middle ear space and middle fossa, and also revealed focal erosion at the superior aspect of the cochlea. Gross tumor involvement was noted at the head of the malleus, in the area of the geniculate ganglion, and rostrally to the capsule of the TMJ. This was removed and then followed into the ITF to complete the extracranial resection. The eustachian tube was packed with fat and oversewn. Unfortunately, the facial nerve did not stimulate at this point.

After microscopic inspection of the resection cavity to ensure complete tumor extirpation, preparation was made for CI. Mastoidectomy and facial recess approach were performed through the postauricular incision. The round window was identified and was free of tumor. A cochleostomy was drilled anteroinferior to the round window and the Nucleus Freedom implant (Cochlear Corp., Lane Cove, Australia) was inserted with jeweler's forceps. Full insertion was achieved, and the electrode was visualized in the cochlea through the eroded middle fossa floor. The middle ear and mastoid cavity were packed with free abdominal fat. The implant was then placed in a previously drilled superoposterior well, with the magnet removed. Two Prolene (Ethicon, Somerville, NJ) sutures placed through paired tie-down holes and tied in a figure of eight were used to secure the implant.

Dural repair and skull base reconstruction were performed in a layered fashion with DuraGuard (Synovis Surgical Innovations, St. Paul, MN) underlay, followed by interposed free abdominal fat, then DuraGen (Integra NeuroSciences, Plainsboro, NJ), and finally a pedicled temporalis muscle rotation flap. The bone flap was then replaced and the zygomatic arch plated. Three-layer soft tissue closure was performed to ensure watertight repair. Blood loss during the procedure was moderate, requiring transfusion of 1 U of packed red cells. The

patient was extubated in the operating room and taken to the neurointensive care unit (NICU) in stable condition. Postoperative MRI confirmed Simpson grade I resection.

On gross pathology, the tumor was soft and red-tan colored with multiple areas of hemorrhage and with fragments of irregular-appearing calvarial and petrous bone. After formalin fixation, hematoxylin and eosin stains showed tightly packed, spindle-shaped and epithelioid cells with oval or irregular nuclei and occasional visible nucleoli (Fig. 2A). Numerous mitotic figures were seen, and the characteristic "staghorn" vessels were present (Fig. 2B). Reticulin preparation showed abundant pericellular reticulin (Fig. 2C). Immunohistochemistry was strongly positive for CD 34 and negative for other stains, including epithelial membrane antigen and factor XIIIa. Ki-67 immunostaining was performed, and a proliferation index of ~10% was calculated, indicating aggressive tumor growth. A small area of fibrosis at the lateral edge corresponded to the site of previous biopsy. No necrosis was found.

The patient had an arduous postoperative course, with significant disorientation, combative behavior, and waxing and waning mental status. This was complicated by the patient's now complete deafness, as the CI was not activated until postoperative day 18. Serial non-contrast head CT scans, however, did not reveal intracranial bleed or hydrocephalus. The patient required NICU management for 8 days, including one episode of reintubation for oversedation. Feeding tube placement was required for aspiration prophylaxis and nutrition deficit. In addition, right-sided facial nerve function was House-Brackman 6/6. This contributed significantly to the patient's initial difficulty with intelligible speech and oral competence.

Mental status slowly cleared in the second postoperative week, and the patient's behavior improved significantly after CI activation. He was able to ambulate and work with physical therapy. On postoperative day 20, the patient was discharged to inpatient rehabilitation. Since that time, the patient has made a significant recovery. Swallowing function returned with time and therapy, and the patient's feeding tube was removed

Figure 2 (A) Hematoxylin and eosin preparation demonstrates tightly packed, spindle-shaped and epithelioid cells with oval or irregular nuclei and occasional visible nucleoli. (B) Typical "staghorn" vascular channels associated with hemangiopericytoma (HPC). (C) a reticulin preparation demonstrating abundant pericellular reticulin, which is characteristic of HPC.

~13 months after surgery. Vision and extraocular movements remain intact. A right canthoplasty was required for cornea protection, though facial nerve function has now improved to House-Brackman 3/6. Interrogation of the CI shows 16 of 22 electrodes active and hint scores around 40%. The patient is pleasant and conversant, with some reliance on lip reading but without obvious language deficit. Cognitive function has returned to preoperative baseline, and he is independent in his activities of daily living.

Adjuvant radiation was given to the skull base with a full course dose of 60 Gy. No chemotherapy was given. Follow-up MRIs performed at 6 and 12 months show no recurrence of tumor. No regional or distant metastases have occurred, and the patient is disease free at 18 months.

DISCUSSION

HPCs are a rare subtype of soft tissue sarcoma derived from Zimmerman's pericytes and were first described by Stout and Murray in 1942.¹ Pericytes are a specialized type of mesenchymal cells that surround the endothelial layer of capillaries and venules and contribute to local blood flow regulation.²² These tumors represent only ~1% of all vascular neoplasms and only ~3% of soft tissue sarcomas.¹⁻⁴ Most occur in the fifth decade, though some cases in infants and children have been reported.²³⁻²⁵ Presentation is usually prompted by persistent pain or an obvious, sometimes pulsatile or discolored, mass lesion.

HPC may occur in a wide variety of anatomic locations, most commonly in the lower extremities, followed by regions of the trunk and the upper extremities.^{5,7,22,25-28} Approximately one-quarter of HPCs occur in the head and neck region, with studies quoting incidences from 7.5 to 30%.^{5,7-10} Skull base or temporal bone HPCs are very rare, with fewer than 15 cases reported in the literature.¹¹⁻¹⁶ Only two cases with significant intracranial extension of an ITF HPC are reported in the literature.^{13,14} This is to be distinguished from primary intracranial HPC or distant metastasis.^{27,29,30} Interestingly, in addition to the skull base HPC, our patient had a contralateral vestibular schwannoma. He does not carry a diagnosis of neurofibromatosis (NF-2). Although it is tempting to find a common etiology for the two lesions, clinical connections between these tumor types have not been established. Further, our patient's HPC seems to have originated in the preauricular soft tissue and extended medially to involve the skull base and middle fossa, rather than originating intracranially and growing outward.

HPCs have widely variable malignant potential. Though they generally follow an indolent course, more aggressive and invasive tumors have been reported.^{12,13,30,31} In addition, acute expansion of pre-

viously quiescent lesions is a known phenomenon, as seems to have occurred in our case.³¹ Treatment is primarily surgical, with the roles of neoadjuvant and adjuvant therapy not clearly defined.^{12,32} Studies of radiation alone have shown poor efficacy, though adjuvant radiation to the surgical bed is supported by good evidence from at least two studies.^{33,34} Radiation is important particularly in cases of incomplete resection and in cases where recurrence would either be difficult to detect or extremely morbid, as in the skull base. Our patient received a standard full course of adjuvant radiation, in addition to complete resection. The utility of chemotherapy in the management of HPC remains unclear, and to date, has been negligible.¹² Regimens including doxorubicin, interferon- α , and a variety of other agents have been used for recurrence, widespread metastases, unresectable disease, and palliation, but little survival benefit is apparent and the systemic toxicities of these drugs are significant.^{12,32,33,35,36}

Local recurrence is expected in up to 40% of cases.^{12,37} Metastasis is common and is frequently delayed more than 3 years from treatment of the primary lesion.⁷ Lung, bone, and liver are common sites for distant metastases. Survival in HPC appears to be related to histological grade (with tumors divided into low, intermediate, and high grade), in addition to tumor size (>6.5 cm diameter), surgical margin status, necrosis, and mitotic activity.^{5,7,12,32,38,39} Our patient had a medium-sized lesion without necrosis, though the significant mitotic activity observed renders this a high-grade lesion.

Hearing loss attributable to skull base HPC has been described in only two cases in the literature, both low-grade tumors occurring in the middle ear space.^{15,16} These lesions caused a predominantly conductive loss, however, as inner ear tumor was not present. In our case, as in other invasive processes and malignancies of the temporal bone or ITF, hearing loss resulted from disruption of the fluid spaces of the cochlea or direct infiltration of the cochlear artery or nerve. Interestingly, hearing loss has not been a prominent presenting feature in cases of primary intracranial hemangiopericytoma, even though most of these lesions occur in the temporal region or cerebellopontine angle.^{29,30}

Simultaneous tumor resection and CI is possible and, in select cases, can be both safe and effective. The overwhelming majority of the literature supporting concurrent CI comes from acoustic neuroma surgery, and in particular, patients with neurofibromatosis type II.¹⁷⁻²⁰ Little if any data exist regarding CI at the time of malignant tumor extirpation. The scenario encountered in our case was extremely unique. The patient was already deaf in the contralateral ear secondary to an untreated but stable acoustic neuroma. Further, the patient experienced SSNHL just 2 weeks before tumor resection. Though corticosteroids were given and

did result in reduced intraparenchymal edema and improvement of vestibular symptoms, no hearing recovery was noted preoperatively.

Perhaps the most complicated issues with concurrent CI with skull base malignancies surround adjuvant radiation therapy and the need for surveillance imaging over an extended time. There are some data evaluating the effects of radiation on CIs, with adequate device function noted in therapeutic range of external beam radiation.⁴⁰ In our single experience, no complication or device malfunction occurred. To perform post-operative and surveillance MRIs, the magnet normally used to hold the CI to the patient's scalp was replaced with a nonmagnetic plug. Though this required the use of a liquid fixative to keep the extracranial components in place, adequate device function was apparent at each visit. Given the probability of late recurrence or metastasis, this patient will likely never have the magnet replaced.

The extreme circumstances in this case make it difficult to draw generalized conclusions. However, there is a need for more studies of CI at the time of skull base surgery, particularly for tumors other than acoustic neuromas. HPCs remain a very rare skull base malignancy, though complete resection and adjuvant radiation can lead to excellent outcomes in even the most complicated of cases.

REFERENCES

1. Stout AP, Murray MR. Hemangiopericytoma: A vascular tumor featuring Zimmermann's pericytes. *Ann Surg* 1942; 116:26–33
2. Stout AP. Hemangiopericytoma; a study of 25 cases. *Cancer* 1949;2:1027–1054; illust
3. Dehner LP, Askin FB. Tumors of fibrous tissue origin in childhood. A clinicopathologic study of cutaneous and soft tissue neoplasms in 66 children. *Cancer* 1976;38:888–900
4. Russell WO, Cohen J, Enzinger F, et al. A clinical and pathological staging system for soft tissue sarcomas. *Cancer* 1977;40:1562–1570
5. Batsakis JG, Rice DH. The pathology of head and neck tumors: vasoformative tumors, part 9B. *Head Neck Surg* 1981;3:326–339
6. de Campora E, Calabrese V, Bianchi PM, Camaioni A, Corradini C. Malignant hemangiopericytoma of the nasal cavity. Report of a case and review of the literature. *J Laryngol Otol* 1983;97:963–968
7. Enzinger FM, Smith BH. Hemangiopericytoma. An analysis of 106 cases. *Hum Pathol* 1976;7:61–82
8. Gührin R. Haemangiopericytoma in otolaryngology. *J Laryngol Otol* 1979;93:477–494
9. McMaster MJ, Soule EH, Ivins JC. Hemangiopericytoma. A clinicopathologic study and long-term followup of 60 patients. *Cancer* 1975;36:2232–2244
10. Sabini P, Josephson GD, Yung RT, Dolitsky JN. Hemangiopericytoma presenting as a congenital midline nasal mass. *Arch Otolaryngol Head Neck Surg* 1998;124: 202–204
11. Abdel-Fattah HM, Adams GL, Wick MR. Hemangiopericytoma of the maxillary sinus and skull base. *Head Neck* 1990;12:77–83
12. Carew JF, Singh B, Kraus DH. Hemangiopericytoma of the head and neck. *Laryngoscope* 1999;109:1409–1411
13. Diensthuber M, Götz F, Länger F, Lenarz T, Lenarz M. Extra- and intracranial dumbbell-shaped hemangiopericytoma. *Eur Arch Otorhinolaryngol* 2008;265:481–484
14. Kanazawa T, Nishino H, Miyata M, Kuriki K, Abe K, Ichimura K. Haemangiopericytoma of infratemporal fossa. *J Laryngol Otol* 2001;115:77–79
15. Karasen RM, Sutbeyaz Y, Gundogdu C, Eren S, Sirin S. Haemangiopericytoma of the middle ear with benign histological features. *J Laryngol Otol* 1999;113:911–913
16. Sutbeyaz Y, Selimoglu E, Karasen M, Ciftcioglu A, Ozturk A. Haemangiopericytoma of the middle ear: case report and literature review. *J Laryngol Otol* 1995;109:977–979
17. Ahsan S, Telischi F, Hodges A, Balkany T. Cochlear implantation concurrent with translabyrinthine acoustic neuroma resection. *Laryngoscope* 2003;113:472–474
18. Aristegui M, Denia A. Simultaneous cochlear implantation and translabyrinthine removal of vestibular schwannoma in an only hearing ear: report of two cases (neurofibromatosis type 2 and unilateral vestibular schwannoma). *Otol Neurotol* 2005;26:205–210
19. Arriaga MA, Marks S. Simultaneous cochlear implantation and acoustic neuroma resection: imaging considerations, technique, and functional outcome. *Otolaryngol Head Neck Surg* 1995;112:325–328
20. Zanetti D, Campovecchi CB, Pasini S, Nassif N. Simultaneous translabyrinthine removal of acoustic neuroma and cochlear implantation. *Auris Nasus Larynx* 2008;35:562–568
21. Fisch U. Infratemporal fossa approach for glomus tumors of the temporal bone. *Ann Otol Rhinol Laryngol* 1982; 91(5 Pt 1):474–479
22. Koch M, Nielsen GP, Yoon SS. Malignant tumors of blood vessels: angiosarcomas, hemangioendotheliomas, and hemangiopericytomas. *J Surg Oncol* 2008;97:321–329
23. Atkinson JB, Mahour GH, Isaacs H Jr, Ortega JA. Hemangiopericytoma in infants and children. A report of six patients. *Am J Surg* 1984;148:372–374
24. Carvalho JR, Haddad L, Leonhardt FD, et al. Head and neck hemangiopericytoma in a child: case report. *Sao Paulo Med J* 2004;122:223–226
25. Weiss SWGJ. Enzinger and Weiss's Soft Tissue Tumours. 4th ed. St. Louis, MO: C.V. Mosby; 2001
26. Chnaris A, Barbetakis N, Efstathiou A, Fessatidis I. Primary mediastinal hemangiopericytoma. *World J Surg Oncol* 2006;4:23
27. Epat NJ, Bilsky M, Lewis JJ, Leung D, Brennan MF. Soft tissue sarcoma brain metastases. Prevalence in a cohort of 3829 patients. *Cancer* 2002;94:2706–2711
28. Epat NJ, Lewis JJ, Leung D, et al. Conventional hemangiopericytoma: modern analysis of outcome. *Cancer* 2002;95:1746–1751
29. Heiser MA, Waldron JS, Tihan T, Parsa AT, Cheung SW. Temporal fossa hemangiopericytoma: a case series. *Otol Neurotol* 2009; (Epub ahead of print)
30. Tashjian VS, Khanlou N, Vinters HV, Canalis RF, Becker DP. Hemangiopericytoma of the cerebellopontine angle: a case report and review of the literature. *Surg Neurol* 2009; 72:290–295

31. Musacchio M, Mont'Alverne F, Belzile F, Lenz V, Riquelme C, Tournade A. Posterior cervical haemangiopericytoma with intracranial and skull base extension. Diagnostic and therapeutic challenge of a rare hypervascular neoplasm. *J Neuroradiol* 2003;30:180-187
32. Chamberlain MC, Glantz MJ. Sequential salvage chemotherapy for recurrent intracranial hemangiopericytoma. *Neurosurgery* 2008;63:720-726; author reply 726-727
33. Galanis E, Buckner JC, Scheithauer BW, Kimmel DW, Schomberg PJ, Piepgras DG. Management of recurrent meningeal hemangiopericytoma. *Cancer* 1998;82:1915-1920
34. Staples JJ, Robinson RA, Wen BC, Hussey DH. Hemangiopericytoma—the role of radiotherapy. *Int J Radiat Oncol Biol Phys* 1990;19:445-451
35. Kirn DH, Kramer A. Long-term freedom from disease progression with interferon alfa therapy in two patients with malignant hemangiopericytoma. *J Natl Cancer Inst* 1996;88:764-765
36. Ortega JA, Finklestein JZ, Isaacs HJr, Hittle R, Hastings N. Chemotherapy of malignant hemangiopericytoma of childhood. Report of a case and review of the literature. *Cancer* 1971;27:730-735
37. Walike JW, Bailey BJ. Head and neck hemangiopericytoma. *Arch Otolaryngol* 1971;93:345-353
38. Fletcher CD, McKee PH. Sarcomas—a clinicopathological guide with particular reference to cutaneous manifestation. III. Angiosarcoma, malignant haemangiopericytoma, fibrosarcoma and synovial sarcoma. *Clin Exp Dermatol* 1985;10:332-349
39. Kraus DH, Dubner S, Harrison LB, et al. Prognostic factors for recurrence and survival in head and neck soft tissue sarcomas. *Cancer* 1994;74:697-702
40. Ralston A, Stevens G, Mahomudally E, Ibrahim I, Leckie E. Cochlear implants: response to therapeutic irradiation. *Int J Radiat Oncol Biol Phys* 1999;44:227-231