Ewing's Sarcoma of the Petrous Temporal Bone: Case Report and Literature Review

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ABSTRACT

Ewing's sarcoma, which accounts for 6 to 9% of malignant bone neoplasms in children, typically affects the trunk and long bones and less often affects the skull (i.e., maxilla, frontal, parietal, ethmoid, temporal bones). Adding to literature of five previously reported cases, we now describe the case of the oldest child, a 16-year-old boy, with a primary Ewing's sarcoma of the petrous temporal bone. When this patient presented after 1 week of right-sided facial paralysis and new-onset headache, imaging studies showed a mass that originated in the right petrous temporal bone. During biopsy and surgical excision, the mass was found to involve the facial nerve, which then required nerve grafting. Postoperatively, he then underwent radiotherapy with both induction and adjuvant chemotherapy. Although an uncommon tumor of the temporal bone, physicians should consider Ewing's sarcoma in the differential diagnosis of children and adolescents who present with facial nerve paralysis.

KEYWORDS: Ewing's sarcoma, petrous temporal bone, facial nerve paralysis

Ewing's sarcoma is a malignant, small, round cell tumor arising from bone and primarily affects children and adolescents. Ewing's sarcoma is an aggressive malignant tumor of childhood and the second most common primary bone cancer. Among pediatric patients, Ewing's sarcoma accounts for 6 to 9% of malignant bone neoplasms.^{1,2} Although the trunk and long bones are typically affected, involvement of bones throughout the body has been reported. In 1 to 6% of total Ewing's sarcoma cases, this malignant tumor affects the skull (i.e., maxilla, frontal, parietal, ethmoid, or temporal bones).^{3,4} We describe the oldest child affected by a primary Ewing's sarcoma of the petrous temporal bone and discuss this patient's presentation and management relative to the five previously reported cases.⁴⁻⁸

CASE REPORT

A 16-year-old boy presented to an outside hospital for evaluation of a right-sided facial weakness. After an initial diagnosis of Bell's palsy for which medical treatment with acyclovir and oral steroids was prescribed, no response to the antiviral and steroid therapy was observed. One week later, he presented to the emergency room when he developed a right-sided, lancinating headache that was associated with nausea and vomiting. When a noncontrast computed tomography (CT) scan of the head revealed an intracranial lesion, he was transferred to our facility for treatment.

Magnetic resonance imaging demonstrated a well-defined extra-axial lesion that measured $2.8 \times 2.7 \times 2.1$ cm. The mass originated from the right petrous

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Figure 1 Axial T1-weighted magnetic resonance scan with flair sequence, postcontrast, with fat saturation. Tumor is seen involving the geniculate ganglion and extending into the tympanic segment of the facial nerve (arrowhead).

apex and extended laterally into the internal auditory canal and posteriorly along the right tentorial leaflet in the cerebellopontine angle (Fig. 1). The patient's medical history, physical examination, and audiological testing were normal with the exception of intermittent right-sided tinnitus, headache, and facial weakness (House-Brackmann grade V). Magnetic resonance angiographic studies further delineated the vascularity of the tumor. The mass was found to abut the right internal carotid artery and the right posterior communicating arteries without associated arterial stenosis or irregularity. The tumor's primary arterial supply was from a proximal branch of the middle meningeal artery. A dedicated temporal bone CT scan demonstrated abnormal periosteal reaction and new bone formation of the petrous bone at the level of the cochlea and abutting the fallopian canal (Fig. 2).

Based on the characteristics observed on the imaging studies, potential diagnoses included meningioma and sarcoma. After counseling the patient and his family, biopsy with possible resection of the lesion was planned using a temporal craniotomy with middle cranial fossa and transmastoid approaches. Operative findings revealed the mass had involved the petrous apex, compressed the temporal lobe, and extended into the proximal tympanic segment, geniculate ganglion, and labyrinthine segment via the facial hiatus (Fig. 3). Frozen-section analysis showed a neoplasm that was composed predominantly of small round cells. After resection of the facial nerve because of tumor involve-

Figure 2 Coronal noncontrast temporal bone computed tomography scan showing evidence of marked bone destruction and periosteal reaction.

ment in the perigeniculate region, a greater auricular nerve cable graft was used for reconstruction. A near total gross resection (95%) was achieved as confirmed by the intraoperative findings and postoperative imaging studies. With an uneventful postoperative course, the patient was discharged on postoperative day 8.

Final histopathology revealed a malignant, small round cell tumor most consistent with Ewing's sarcoma (Fig. 4). Analysis of the specimen with reverse transcriptase polymerase chain reaction and sequencing identified the presence of the *EWS-FLI1* fusion gene resulting from translocation $t(11;22)(q24;q12)$; overexpression of the membrane protein CD99 was also present.

As complete resection could not be achieved without substantial additional morbidity, the postoperative protocol included induction chemotherapy with radiotherapy and adjuvant chemotherapy. Four cycles of induction chemotherapy with vincristine, cyclophosphamide, doxorubicin, ifosfamide, and etoposide were administered and tolerated by the patient.⁹ Radiotherapy of 54 Gy was administered in 30 fractionations. After

Figure 3 Intraoperative photograph demonstrating dilation of tympanic segment of facial nerve (arrowhead).

Figure 4 Light microscopy (400 $\times$). (A) Hematoxylin and eosin staining shows sheets of uniform undifferentiated cells. (B) A strong and diffuse membranous staining pattern on CD99 antibody stain.

completion of this treatment protocol, the patient was doing well with no sign of tumor growth or recurrence.

DISCUSSION

Ewing's sarcoma that affects the skull accounts for only 1 to 6% of total cases and usually develops in patients younger than 20 years; however, one report involved a 54-year-old man.¹⁰ Primary Ewing's sarcoma of the skull typically respects dural planes and often presents as an expansile mass causing symptoms of increased intracranial pressure. Within the skull, this sarcoma typically affects the frontal and parietal bones, as well as the temporal (squamosa and mastoid), ethmoid, orbital, and maxillary bones. On the basis of our review of the world literature, we identified Ewing's sarcoma with involvement of the petrous temporal bone in only five previous reports.⁴⁻⁸ With the exception of age, characteristics of our 16-year-old patient were similar to the other five patients who ranged in age from 5 months to 9 years at presentation (Table 1). Considering these six patients with petrosal Ewing's sarcoma, presentation included lower motor neuron facial palsy in

Table 1 Clinical Summary of Six Patients with Ewing's Sarcoma of the Petrous Temporal Bone

Author and Reference	Age	Presentation	Surgery	Adjuvant Therapy	Follow-up
Kadar et al	16 y	Right facial palsy, ipsilateral tinnitus	Temporal craniotomy, petrosectomy, nerve graft	Chemotherapy and radiotherapy	9 mo
Pfeiffer et al ⁸	5 mo	Transient facial palsy at age 2 mo, subsequent complete facial palsy	Stereotactic biopsy	Chemotherapy and radiotherapy	25 mo, developmentally normal
Carlotti et al ⁶	5 mo	Facial paralysis, signs of raised ICP	Petrosectomy, craniotomy	Chemotherapy	8 y, no evidence of recurrence
Desai et al ⁷	18 mo	Syncope, emesis, no focal neurological deficit	Lateral subtemporal approach	Chemotherapy and radiotherapy	6 mo, no sign of residual tumor
Steinbok et al ⁴	3 y	Headache, emesis, left temporal swelling	Subtotal en bloc removal	Chemotherapy	Not mentioned
Fitzer and Steffey ⁵	9 y	Otorrhea, hearing loss, facial weakness	Facial nerve decompression through middle fossa approach, procedure abandoned	Chemotherapy and radiotherapy	Not mentioned

ICP, intracranial pressure.

four patients; headache, nausea, and vomiting in three patients; temporal swelling in one patient; and otorrhea in one patient.

Histopathologic features of Ewing's sarcoma show monotonous sheets of monomorphic round cells with scant cytoplasm, round nuclei, and inconspicuous nucleoli.^{11,12} Molecular genetic analysis for chromosomal translocation t(11:22)(q24; q12) is pathognomonic.¹³ Overexpression of CD99, a transmembrane protein encoded by the *MIC-2* gene, is another helpful adjunct.^{3,13} Rhabdomyosarcoma, which is high on the differential as the most common skull base sarcoma in the pediatric population, is also a small, round, blue cell tumor that can be easily confused with Ewing sarcoma. In both cases, initial biopsy of an infiltrating skull base tumor is usually the most appropriate intervention because these tumors typically respond quickly and dramatically to chemotherapy. Local control with surgery or radiotherapy is ideally delayed until after this initial response to chemotherapy.

Treatment options for Ewing's sarcoma include induction chemotherapy, local control, and adjuvant chemotherapy. Chemotherapy, using a multimodal therapeutic regimen, has greatly increased survival from less than 10% to more than 50%.¹⁴ Chemotherapeutic agents commonly used include vincristine, doxorubicin, cyclophosphamide, etoposide, and ifosfamide.¹⁴⁻¹⁶ Before the introduction of chemotherapy, most patients died of metastatic disease within 2 years. Options for local control include surgical excision and radiation. Radiation is the primary local control modality when tumors are difficult to access or when surgical excision would lead to unacceptable morbidity. In contrast with patients with Ewing's sarcoma affecting the pelvis or lower body, patients with cranial lesions often undergo urgent surgical treatment because of elevated intracranial pressure and impending neurological deficits; consequently, induction radiation or chemotherapy is not commonly used. Tumors arising from the calvarium may be amenable to complete surgical resection. However, complete excision of skull base lesions is often impossible without substantial morbidity.

CONCLUSION

This case report of a 16-year-old boy adds to the five previously reported cases of this rare form of Ewing's sarcoma affecting the petrous temporal bone. When this bone is affected, the tumor presents unique management considerations because of its intimate relationship with critical neurovascular structures in this region. Given its rarity for affecting the skull, particularly the petrous temporal bone, our findings add to our global understanding of the treatment options and outcomes for this uncommon subset of patients affected by Ewing's sarcoma.

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