Anaplastic Ependymoma Metastatic to the Neck
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Introduction
Ependymomas are neuro-ectodermal tumors derived from ependymal cells lining the ventricles of the central nervous system.1 They represent 2-5% of all brain tumors and 5-10% of all brain tumors in children. They tend to be locally aggressive within 5 years after diagnosis in 40-70% of cases. The World Health Organization classifies all ependymomas as either subependymoma (grade I), ependymoma (grade II), or anaplastic ependymoma (grade III).2 Surgical resection is the primary treatment. Despite aggressive treatment, often with multiple treatment modalities, the 5-year survival rate for all ependymomas does not exceed 60%.1 There are only two previously reported cases of anaplastic ependymoma metastatic to the neck in the literature. We present the third case of this rare sequela in a patient with metastases to the ipsilateral parotid gland and cervical lymph nodes.

Case Presentation
This patient is a 57-year-old male with a history of recurrent anaplastic ependymoma of the right parietal lobe first diagnosed in 1998. After having undergone a right parietal craniotomy with subtotal resection of a malignant ependymoma at that time, he received chemotherapy and a full course of postoperative local field irradiation. He presented with recurrence in the right parietal lobe five times in the next 7 years. The first four of these recurrences were treated with parietal craniotomies and gross total resections. The fifth recurrence was treated with gamma knife radiosurgery. At this time, the patient began taking the oral chemotherapeutic agent temozolomide (330 mg five days per month). The patient tolerated only four rounds secondary to toxicity in the form of nausea and vomiting. Four months later, the patient noted a firm nodule involving the inferior aspect of his craniotomy scar. Excision of the subgaleal mass under local anesthesia revealed anaplastic ependymoma. A similar nodule was again removed from the right parietal scalp four months later. In December 2006, he had yet another craniotomy for recurrence at the primary site as well as excision of another scalp nodule. In January 2007, the patient was referred to the head and neck oncology clinic with rapid-onset right suboccipital, postauricular, parotid, and lymphadenopathy.

On examination, there was a subcentimeter nodule in the suboccipital region, two firm 0.5-cm nodules just posterior to the right postauricular skin crease, a 1-cm nodule within the tail of the right parotid gland, and another 1-cm nodule in the right level II neck. He was admitted to the hospital within one week for planned superficial parotidectomy with facial nerve preservation and extended modified radical neck dissection. In the immediate preoperative period, examination showed enlargement of his level IIB lymph node as well as a new 0.5-cm nodule in level IV. The patient tolerated his surgical procedure well and had an uneventful postoperative course. Histological analysis revealed anaplastic ependymoma within multiple lymph nodes in the right superficial parotid region, level IIB, and level V. One lymph node in level IIB showed extranodal involvement.

Since that time, the patient has received the chemotherapeutic agents bevacizumab and irinotecan, which he has tolerated well. He again developed a recurrence in his subgaleal scalp measuring 7 cm by 3 cm noted on a PET scan in August 2007 requiring surgical resection. At the time of this writing, the patient remains active with an acceptable quality of life. He does complain of right-sided hearing loss and has a spastic left hemiparesis.

Discussion
Ependymomas comprise a relative paucity of brain tumors overall, and the incidence of metastasis is rarer still. Low-grade ependymomas tend to occur infratentorially while high-grade tumors of this histology have a predilection for the cerebral hemispheres.2 Pediatric cases are often present in the cerebrum and exhibit high-grade features, while the adult manifestation of ependymoma is usually marked by a spinal location and a more well-differentiated tumor.1,2 Though they most commonly originate from the floor of the fourth ventricle, ependymomas can occur throughout the CNS. Prognostic factors for ependymoma include radiation strategy, chemotherapy, histologic grade, and location; the most significant factor, however, remains the extent of resection, and location often precludes complete removal of tumor.2 Local field irradiation has been deemed suitable for adjuvant therapy, as the vast majority of recurrences occur at the site of the primary tumor.1 While dissemination of intracranial ependymomas occurs frequently, especially in high-grade lesions, the incidence of extracranial metastasis to the neck for this type of brain tumor remains exceedingly rare.1,2

As previously mentioned, our patient represents the third documented case of anaplastic ependymoma metastasis to the cervical lymph nodes. This uncommon manifestation possesses relevance not only as an esoteric curiosity but also because it illustrates another pattern of disease progression, albeit infrequent, with which clinicians should be familiar. It perhaps also demonstrates the significance of brain extracellular fluid drainage through deep cervical lymphatic channels. This evidence of drainage to the cervical lymphatics would seem to challenge the traditional assumption that the afferent arm of the immune response to brain antigens is interrupted by a lack of conventional lymphatic outflow.

In the case of our patient, his tumor exhibited the aggressive features characteristic of an intracranial ependymoma. Unlike the majority of anaplastic ependymoma patients, however, his age is far removed from the normal pediatric demographic. The pattern of local intracranial recurrence in this patient is quite common, as dissemination occurs in 10% of patients and location often limits resectability.1 While this patient’s most recent metastasis to the scalp also correlates well with previous reports of extra-CNS ependymoma metastasis to the skin, lungs, and scalp, the presence of disease in his cervical lymph nodes remains an uncommon and noteworthy presentation.

Conclusions
Anaplastic ependymomas are aggressive tumors which have a propensity for local recurrence as well as regional metastases. Gross total resection initially and also with each recurrence is recommended, as our patient maintained adequate function and quality of life through multiple intracranial procedures. Local field radiation, chemotherapy, and gamma knife surgery play a role as adjunctive treatments. Development of cervical metastases requires immediate referral to an otolaryngologist.

References