Ceruminous Gland Adenoid Cystic Carcinoma With Contralateral Metastasis to the Brain

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We present the case of a 38-year-old man with an adenoid cystic carcinoma originating from the ceruminous glands of the external ear canal. The patient subsequently presented with a contralateral brain mass that was also diagnosed as adenoid cystic carcinoma. To our knowledge, contralateral metastasis to the brain of a patient with an adenoid cystic carcinoma of the ceruminous glands has not been reported previously. This rare neoplasm should be considered in the differential diagnosis of poorly differentiated carcinomas metastatic to the central nervous system in patients with occult malignant neoplasms.

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REPORT OF A CASE

A 38-year-old black man presented to the Otolaryngology Clinic with a 4- to 6-month history of bloody drainage from the left external ear canal. Radiologic evaluation demonstrated some slight thickening within the skin of the external ear canal, as well as possible lytic areas in the underlying bone. A lesion obliterating the external canal was biopsied and diagnosed as poorly differentiated carcinoma. The patient subsequently returned for definitive surgical therapy, which consisted of resection of the mass, including the external auditory canal, tympanic membrane, malleus, and incus, as well as a portion of the left temporal bone and the left parotid gland. At this time, the tumor was diagnosed as a primary adenoid cystic carcinoma of the ceruminous glands. Eight months later, the patient began to complain of right temporal headaches. Magnetic resonance imaging performed at that time failed to show any intracranial lesions or recurrent disease. After 4 months, a computed tomographic scan demonstrated a large contralateral mass in the parenchyma of the parietal lobe of the brain, as well as an overlying soft tissue mass, which was palpable on clinical examination. No ipsilateral lesions were identified. The brain lesion was subsequently biopsied and diagnosed as metastatic adenoid cystic carcinoma.

PATHOLOGIC FINDINGS

Material received for pathologic review at the time of the definitive resection included an irregular fragment of soft tissue consisting of the external ear canal, which was enclosed by 2 irregular fragments of bone and cartilage. The external ear canal was almost entirely occluded by a neoplasm measuring up to 1.3 cm in maximum diameter. The neoplasm did not appear to infiltrate the surrounding soft tissues or extend into the salivary gland tissue (Figure 1). The salivary gland tissue was uninvolved. Biopsies from the brain mass were composed of a 6 × 5.5 × 4.2-cm piece of tan to brown variegated necrotic tissue with an attached piece of fibroconnective tissue.

Microscopic examination of the external ear canal lesion demonstrated an infiltrative malignant neoplasm composed of basaloid cells exhibiting indistinct cell borders, scant amphophilic cytoplasm, and enlarged hyperchromatic nuclei. Numerous mitotic figures were also present. The mass tended to infiltrate in solid aggregates of tumor cells with focal areas of cribriform and tubular components. Some of the tubular and cribriform areas had lumina containing basophilic to eosinophilic basement membrane–like material (Figure 2). No lymphovascular or perineural invasion was identified. Immunohistochemical studies demonstrated strong positive immunoreactivity of tumor cells with antibodies to cytokeratins AE1–AE3, epithelial membrane antigen, and vimentin. The intraluminal basement membrane–like material, as well as the extracellular matrix stroma, exhibited strong positive staining with Alcian blue. Additional immunostains with antibodies to chromogranin, synaptophysin, S100 protein, and muscle-specific actin were negative. Biopsies from the brain mass contained a tumor with similar histologic features and with extensive necrosis (Figure 3). Biopsy material from the brain tumor demonstrated an immunohistochemical staining pattern similar to that of the auditory canal tumor.

COMMENT

The true origin of the glandular neoplasms of the external ear canal is controversial. It has been proposed that these tumors arise from the ceruminous glands, which are modified apocrine glands with some eccrine functions. Immunohistochemical and electron microscopic studies of these tumors have demonstrated histologic and ultrastructural features similar to those found in ceruminous glands. Others have suggested that the tumors arise from ectopic minor salivary glands located in the external ear, although this opinion has not been well documented.

In 1972, Wetli et al described the nomenclature that has become widely accepted to differentiate benign from malignant tumors that were previously all designated as ceruminoma. They separated the tumors into 4 types: (1)
ceruminous gland adenocarcinoma, (2) ceruminous adenoma, (3) adenoid cystic carcinoma, and (4) pleomorphic adenoma. Others have proposed the addition of more rare tumors, such as mucoepidermoid carcinoma and syringocystadenoma papilliferum to this classification system.

Adenoid cystic carcinomas of the ceruminous gland (ACCCGs) are morphologically similar to the more commonly encountered salivary gland tumors. As do their more common salivary gland analogs, ACCCGs have a propensity for perineural growth. This tendency accounts for most cases of intracranial involvement by ACCCG, as the neoplasm grows along the nerves and by direct extension involves the central nervous system. It has been proposed for salivary gland neoplasms that the propensity for perineural involvement is related to the expression of neural cell adhesion molecules by the neoplastic tumor cells. To our knowledge, contralateral metastatic central nervous system involvement has not been reported previously.

Treatment of ACCCG involves complete surgical resection with clear surgical margins. This objective can be difficult to achieve because of the perineural involvement commonly encountered in these neoplasms. Perzin et al reviewed a series of 16 cases of adenoid cystic carcinomas of the external ear canal region, 8 of which were definitively ACCCG. After 20 years of follow-up, 7 of their patients had no evidence of recurrence following surgical resection, 2 patients were living with recurrent unresectable tumors, 5 died of the disease, 1 died of other causes, and 1 was lost to follow-up. Most recurrences occurred within 2 years of initial diagnosis; however, recurrences also were reported to occur up to 14 years after the initial diagnosis.

In summary, this article presents the case of an adenoid cystic carcinoma of the ceruminous glands of the external ear canal with contralateral central nervous system metastasis, a hitherto unreported phenomenon. Although the likelihood of a similar disease progression may be low, the possibility is worthy of consideration in the differential diagnosis of poorly differentiated central nervous system carcinomas in patients with occult malignancies.
References