INTRODUCTION

Rare mixed tumors of the head and neck often present a diagnostic dilemma when patient history, tumor location and histology provide a confusing clinical picture. This case report describes a patient with a benign mixed tumor (pleomorphic adenoma) of the parotid gland that was complicated by multiple recurrences—locally within the parotid bed as well as regional metastases to the scalp. These recurrences all displayed the same benign mixed tumor morphology, but their location within two different subsites of the head and neck and similar histologic appearance led to an uncertain pathologic diagnosis of chondroid syringoma or benign metastasizing pleomorphic adenoma. However, given the entire clinicopathologic picture, we believe this represents a rare case of benign metastasizing pleomorphic adenoma.

DISCUSSION

Both pleomorphic adenomas and chondroid syringomas are referred to as “mixed tumors” because they have microscopic features of epithelial and mesenchymal differentiation. As such, their morphological appearances are similar and the two can be easily confused. Indeed, chondroid syringoma was referred to as “mixed tumor of the skin” until the current terminology was introduced in 1961. However, these two tumors arise from different glands and have different distributions within the head and neck which can help to differentiate them. Pleomorphic adenomas derive from salivary glands and classically exhibit benign ductal structures with myoepithelial cells of salivary gland origin; they can be found anywhere salivary gland tissue exists throughout the upper aerodigestive tract and deep neck. Chondroid syringomas arise from sweat glands and are subclassified into apocrine and eccrine types. They have parenchyma that typically shows pilosebaceous-apocrine differentiation along with the presence of cartilage-like stroma. They are typically found on the nose, cheek, upper lip, scalp, forehead and chin.

Although most mixed tumors of salivary gland origin are found within the glands themselves, certain rare forms can metastasize. These include carcinoma ex-pleomorphic adenoma, malignant mixed tumor (carcinoma sarcoma) and metastasizing mixed tumor (or benign metastasizing pleomorphic adenoma). The first two are true malignant tumors with histological evidence of malignant cells and aggressive clinical behavior. The latter is a poorly understood variant that occurs in patients with both primary and metastatic tumor deposits that appear histologically benign. However, its clinical behavior seems to suggest that it should be considered a low-grade malignancy. It typically presents after multiple local recurrences and often adjuvant radiation of the primary tumor. The interval between diagnosis of the primary lesion and the metastases varies between 3 and 52 years, with the most common sites being bone (45%), head and neck (43%) and lung (36%). Metastatic deposits have also been discovered in regional lymph nodes, the oral cavity, pharynx, skin, liver, retroperitoneum, kidney, calvarium and central nervous system. Because it can be found in locations rare for primary pleomorphic adenomas, it can easily be mistaken by the pathologist for chondroid syringoma (as initially occurred in our case) without the appropriate historical information.

Surgical management is the primary therapeutic intervention for all benign mixed tumors and requires adequate surgical excision with a surrounding rim of normal tissue. Recurrence is associated with incomplete removal of the tumor or the presence of unrecognized satellite tumor nodules.

REFERENCES


CASE REPORT

The patient is a 28-year-old man who presented with a 13-year history of multiply recurrent pleomorphic adenoma of the right parotid gland. He had undergone two surgical resections at outside hospitals when he presented in 2006 with several palpable masses within the right parotid bed. A contrast-enhanced MRI revealed a multifocal recurrence in the superficial lobe of the parotid and the patient underwent revision parotidectomy.

During the surgery five separate foci of tumor were identified within the superficial lobe of the gland. A capsule of normal parotid tissue was left around the specimen and it was excised en bloc with preservation of the underlying facial nerve branches. During the postoperative period the patient had normal facial nerve function and healed without complications. Because of his history of multiple recurrences, the patient was referred for adjuvant radiation therapy. A course to the right parotid bed was abbreviated to 2000 cGy due to patient noncompliance.

One year later he returned with 2 small well circumscribed nodules at the central forehead and right parietal scalp. FNA cytology was read as benign skin adnexal lesions, consistent with chondroid syringomas. The patient underwent simple excision of the lesions, which were both subcutaneous, well encapsulated and less than 1 cm in diameter. The final pathology was read as benign mixed tumors, consistent with chondroid syringomas or benign metastasizing pleomorphic adenomas. A contrast-enhanced MRI performed 5 months later showed no evidence of recurrence in the head and neck.

Parotid lesion (hematoxylin and eosin): A. Low-power micrograph showing an intact capsule with benign morphology; B. Medium-power micrograph showing mixed tumor composed of ductal structures with surrounding myoepithelium and adjacent chondroid myxoid stroma.

Scalp lesion (hematoxylin and eosin): A. Low-power micrograph showing benign morphology and no capsular invasion; B. High-power micrograph showing mixed tumor with ductal elements without pilosebaceous-apocrine differentiation.