ABSTRACT

Objectives
Carcinoma ex pleomorphic adenoma (CXPA) falls under the category of malignant transformations that can occur in a benign pleomorphic adenoma. It is the most common of the transformations, with carcinomas (malignant mixed tumor) and metastasizing adenoma being much less likely. CXPA accounts for 3.6-4% of all salivary tumors and 5-15% of salivary malignancies. CXPA typically arises in the sixth to eighth decades of life, and seldom occurs in young adults. The late onset is thought to be secondary to the protracted transformation into malignancy, which can take an average of 20 years and up to 50 years. Another study suggests that the rate of malignant conversion is 1% for the first year and up to 9.5% if left untreated in 15 years.

The exact etiology for the transformation of pleomorphic adenoma to CXPA is not well understood; however various studies suggest that radiation, genetic instability within the tumor, and increased expression of the cytoplasm of p16 protein by promoter methylation of the p16 gene may all contribute. A molecular study suggests a multi-step model with the progressive loss of heterozygosity at chromosomal arms 8q, then 12q and finally 17p.

CXPA can also be divided into those with only epithelial (luminal) malignancy and those with myoepithelial (non-luminal) malignancy. Myoepithelial cells are contractile cells that serve to expel the secretions of exocrine glands. One study revealed that myoepithelial cells are encouraged by malignant cells to provide fuel for tumor progression.

CASE REPORT
A 28-year-old female presented with expanding painful left-sided soft tissue swelling of the face. The mass was present for only a few months and without any associated facial nerve palsy. Examination revealed a round, well-circumscribed mass in the region of the parotid gland without overlying erythema, edema, or skin breakdown. Intraoral examination showed intact mucosa without evidence of invasion or erosion. CT scan images (figure 1) revealed a heterogeneous mass of the left parotid gland measuring approximately 4.0 x 2.7 x 3.9 cm with hypodense foci suggesting necrosis. A nonspecific level II lymph node was also visualized. Figure 2 displays the ultrasound images of the mass.

Histological findings displayed an intermediate grade myoepithelial carcinoma arising from a pleomorphic adenoma (figures 5 and 6). The carcinoma shows greater than 1.5 mm invasion in to the surrounding adipose tissue (invasive type). The parotid lymph node did not show any evidence of malignancy. The final diagnosis of myoepithelial carcinoma ex-pleomorphic adenoma was made. The tumor inked margins were positive, necessitating adjuvant radiation therapy to the patient successfully underwent at a goal of 60 Gy.

INTRODUCTION

Carcinoma ex pleomorphic adenoma (CXPA) falls under the category of malignant transformations that can occur in a benign pleomorphic adenoma. It is the most common of the transformations, with carcinomas (malignant mixed tumor) and metastasizing adenoma being much less likely. CXPA accounts for 3.6-4% of all salivary tumors and 5-15% of salivary malignancies. CXPA typically arises in the sixth to eighth decades of life, and seldom occurs in young adults. The late onset is thought to be secondary to the protracted transformation into malignancy, which can take an average of 20 years and up to 50 years. Another study suggests that the rate of malignant conversion is 1% for the first year and up to 9.5% if left untreated in 15 years.

The exact etiology for the transformation of pleomorphic adenoma to CXPA is not well understood; however various studies suggest that radiation, genetic instability within the tumor, and increased expression of the cytoplasm of p16 protein by promoter methylation of the p16 gene may all contribute. A molecular study suggests a multi-step model with the progressive loss of heterozygosity at chromosomal arms 8q, then 12q and finally 17p.

CXPA can also be divided into those with only epithelial (luminal) malignancy and those with myoepithelial (non-luminal) malignancy. Myoepithelial cells are contractile cells that serve to expel the secretions of exocrine glands. One study revealed that myoepithelial cells are encouraged by malignant cells to provide fuel for tumor progression.

CASE REPORT
A 28-year-old female presented with expanding painful left-sided soft tissue swelling of the face. The mass was present for only a few months and without any associated facial nerve palsy. Examination revealed a round, well-circumscribed mass in the region of the parotid gland without overlying erythema, edema, or skin breakdown. Intraoral examination showed intact mucosa without evidence of invasion or erosion. CT scan images (figure 1) revealed a heterogeneous mass of the left parotid gland measuring approximately 4.0 x 2.7 x 3.9 cm with hypodense foci suggesting necrosis. A nonspecific level II lymph node was also visualized. Figure 2 displays the ultrasound images of the mass.

Histological findings displayed an intermediate grade myoepithelial carcinoma arising from a pleomorphic adenoma (figures 5 and 6). The carcinoma shows greater than 1.5 mm invasion in to the surrounding adipose tissue (invasive type). The parotid lymph node did not show any evidence of malignancy. The final diagnosis of myoepithelial carcinoma ex-pleomorphic adenoma was made. The tumor inked margins were positive, necessitating adjuvant radiation therapy to the patient successfully underwent at a goal of 60 Gy.

REFERENCES