Dedifferentiated Acinic Cell Carcinoma of the Parotid Gland With Myoepithelial Features

Simonetta Piana, MD; Alberto Cavazza, MD; Corrado Pedroni, MD; Rosa Scotti, MD; Luigi Serra, MD; Giorgio Gardini, MD

- Dedifferentiated acinic cell carcinoma of the salivary gland is an uncommon variant of acinic cell carcinoma, characterized by the coexistence of both an usual low-grade acinic cell carcinoma and a high-grade dedifferentiated component, as well as by an accelerated clinical course. We describe a case of acinic cell carcinoma of the parotid gland in a 67-year-old woman, which recurred 4 times after surgery and radiotherapy. The recurrences consisted of residual foci of acinic cell carcinoma intermingled with a high-grade epithelial proliferation; the latter was focally constituted by cells with morphologic and immunohistochemical features of myoepithelium. (Arch Pathol Lab Med. 2002;126:1104–1105)

Microscopically, the nodules were only partially encapsulated and consisted of a proliferation of medium-sized polygonal cells, showing round nuclei with evident nucleoli and wide, granular, basophilic cytoplasm; nuclear atypia was minimal. Intermingled vacuolated cells were often noted. The neoplastic cells were arranged in a solid and microfollicular pattern. Periodic acid–Schiff stain highlighted cytoplasmic basophilic granules, which appeared diffuse and were resistant to diastase predigestion. This phenomenon has never been described in dedifferentiated acinic cell carcinoma.

REPORT OF A CASE

A 46-year-old woman underwent a right radical parotidectomy, with preservation of the facial nerve, at another institution in 1977. In 1991, she presented with a well-circumscribed nodule, 2.5 cm in diameter, in the right parotid region and underwent a surgical resection. No residual salivary parenchyma was noted during surgery. The nodule was resected and the patient was treated subsequently with local radiotherapy. In 1997, a re-resection of a 1-cm nodule in the right parotid region was performed, with a partial resection of adjacent muscular and adipose tissue. In 1998, a 2.5-cm nodule was found adherent to the right facial nerve; the facial nerve and the nodule were resected. In January 1999, the patient underwent another surgical intervention, with the resection of a 1.5-cm nodule localized near the surgical scar, followed by radiotherapy. In August 1999, a nodule (2 cm in diameter) was noted next to the mandible angle on ultrasonography and was partially excised.

The patient is alive with a local recurrence; a radical surgical intervention has been planned.

PATHOLOGIC FINDINGS

The original resection, performed in 1977 at another institution, involved the superior and deep lobe of the right parotid gland. A review of 2 slides from this resection showed mild chronic inflammation with no evidence of neoplasia.

In all 5 subsequent interventions, from 1991 to 1999, the specimens grossly consisted of well-defined nodules, ranging from 3 to 0.6 cm in diameter, with a smooth surface. All excisions were marginal, and salivary parenchyma was not macroscopically apparent.

Microscopically, the nodules were only partially encapsulated and consisted of a proliferation of medium-sized polygonal cells, showing round nuclei with evident nucleoli and wide, granular, basophilic cytoplasm; nuclear atypia was minimal. Intermingled vacuolated cells were often noted. The neoplastic cells were arranged in a solid and microfollicular pattern. Periodic acid–Schiff stain highlighted cytoplasmic basophilic granules, which appeared diffuse and were resistant to diastase predigestion. This was the only neoplastic component present in the nodule excised in 1991 and in the first recurrence in 1997.

In the second, third, and fourth recurrences, the neoplasm consisted mainly of solid nests of atypical cells with large nuclei and abundant clear cytoplasm and with a high mitotic index. The cells failed to reveal any obvious duct differentiation. An additional feature was the presence of an irregular desmoplastic stroma associated with foci of myxoid stroma, in which the clear cells imperceptibly merged. Furthermore, a minor proliferation, morphologically superimposable to the one described in the first tumors, was present (Figure 1) and was highlighted by periodic acid–Schiff.

Immunohistochemical reactions were performed on the third and fourth recurrences using the avidin-biotin complex (ABC) technique. Neoplastic cells showed strong and diffuse reactivity for S100 protein (Menarini, Italy; Figure 2) and focal positivity for smooth muscle actin (Menarini; Figure 3) and cytokeratin (Dakopatts A/S, Glostrup, Denmark; Figure 4). Chromogranin immunostaining was negative.
Since the original report by Stanley et al in 1988, only a few cases of dedifferentiated acinic cell carcinoma have been described or briefly mentioned in the English literature. Dedifferentiated acinic cell carcinoma is composed of an usual acinic cell carcinoma and a poorly differentiated, highly malignant carcinomatous component in variable proportions. The high-grade carcinoma is often non–mucin-producing, but little is reported about its possible pattern of differentiation. The present report describes an acinic cell carcinoma that recurred with poorly differentiated areas and therefore can be designated “dedifferentiated” acinic cell carcinoma; immunohistochemistry revealed that the poorly differentiated areas were diffusely positive to S100 protein and focally positive to smooth muscle actin and cytokeratin. They had, in other words, a myoepithelial immunoprofile.

Myoepithelial differentiation is common in salivary gland neoplasms. By definition, it is present in epithelial–myoepithelial carcinoma, myoepithelioma, and myoepithelial carcinoma, and it often occurs in benign mixed tumors, adenoid cystic carcinoma, and salivary duct carcinoma. To our knowledge, no cases of myoepithelial features in a dedifferentiated acinic cell carcinoma have been reported to date.

Although dedifferentiation is always associated with tumor progression, little is known about the molecular events that regulate it; immunohistochemical and molecular studies on the expression of p53 oncoprotein have not provided conclusive results.

The prognostic value of dedifferentiation is still to be defined, and statistically significant conclusions on the biological behavior of these tumors is not possible because of the small number of cases described. It is generally accepted that dedifferentiated carcinomas are associated with a poor clinical outcome, as they tend to involve the whole parotid gland and invade the facial nerve.

The current case expands the morphologic spectrum of dedifferentiated acinic cell carcinoma and emphasizes that myoepithelial differentiation should bring to mind differential diagnoses other than the usual myoepithelial salivary gland tumors. When such a myoepithelial differentiation is an expression of dedifferentiation, as in our case, its recognition may have prognostic significance.

References