Pathologic Quiz Case

A 42-Year-Old Man With Right Facial Swelling and Weakness

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A 42-year-old white man presented with right facial swelling that had progressed for 5 months. The patient was unable to shut his eye fully or smile; pain and pressure in this region required the use of narcotics. His medical and surgical histories were unremarkable. A physical examination of this area revealed a 2.0-cm mass in the right parotid gland. A diagnostic fine-needle aspiration of the mass showed a markedly cellular aspirate consisting of cohesive syncytial fragments of malignant epithelial cells; the favored diagnosis at that time was a poorly differentiated nonkeratinizing squamous cell carcinoma. A preoperative neck and nasal computed tomography with contrast demonstrated an enhancing mass in the superficial lobe of the right parotid gland measuring 2.6 × 3.4 cm. Several borderline enlarged lymph nodes were identified in the level II location, one immediately posterior to the parotid gland measuring 12 mm and another at the mandibular angle measuring 13 mm. Magnetic resonance imaging of the region suggested involvement of the facial nerve.

The patient subsequently underwent a right parotidectomy with a facial nerve resection. A frozen section of the facial nerve was positive for malignancy, while the other surgical margins were uninvolved by the tumor. Serial sectioning of the parotidectomy specimen, which measured 7.5 × 6.5 × 4.5 cm, showed the presence of a mass, which represented more than 90% of the total specimen. The lesion was tan-gray and homogeneous with a pushing border. There were a few areas of yellow discoloration measuring from 0.6 to 1.0 cm in maximum diameter within the lesion; however, no areas of overt necrosis or hemorrhage were identified.

Microscopically, the tumor consisted of large cells arranged in sheets with marked nuclear pleomorphism (Figure 1). Mitoses were frequent with areas of necrosis (Figure 2). In some areas, the tumor was more differentiated and formed glandular structures. The cells in these areas had vesicular nuclei with granular basophilic cytoplasm (Figure 3). Periodic acid–Schiff-positive diastase-resistant intracytoplasmic granules were focally present (Figure 4). Mitoses were also seen in these areas.

What is your diagnosis?

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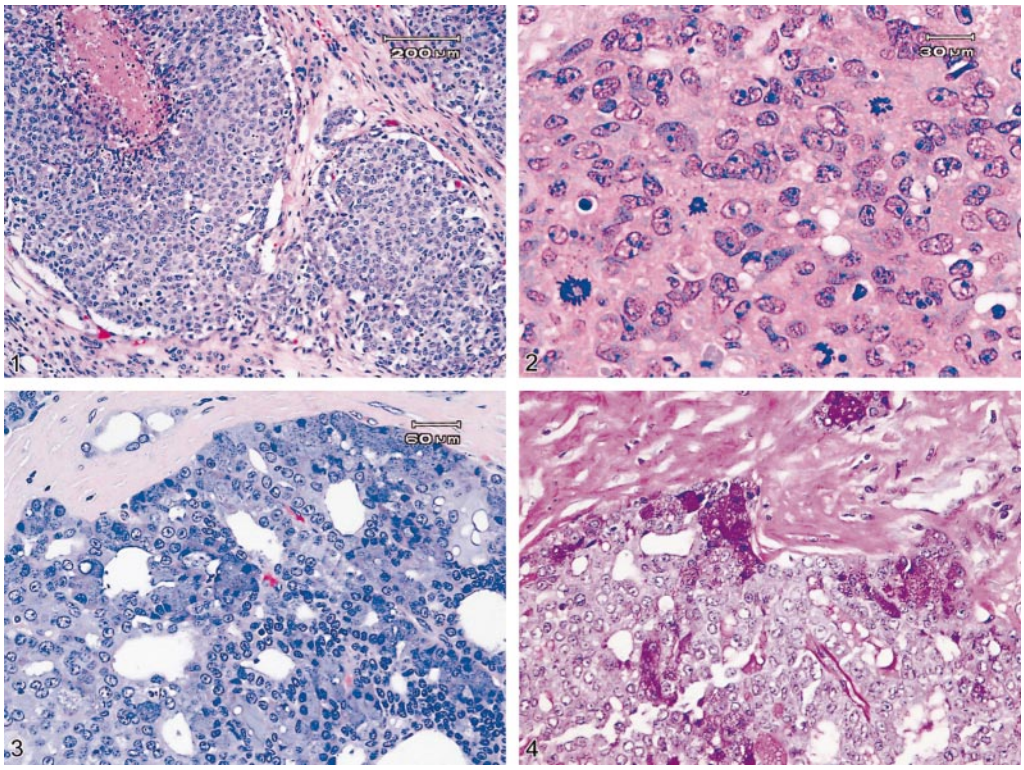

Pathologic Diagnosis: Dedifferentiated Acinic Cell Carcinoma of the Parotid Gland

This rare variant of acinic cell carcinoma was first defined by Stanley et al[1] in 1988 as a complex of typical low-grade acinic cell carcinoma and areas of a high-grade, poorly differentiated adenocarcinoma or undifferentiated carcinoma. The importance of this variant lies in its predictably aggressive nature when compared with the usual acinic cell carcinoma.

The median age for presentation is 58 years with a 5:4 male-female ratio. It is not unusual for patients to present with signs of facial nerve involvement, as in our case. Pain is also a typical feature.

On gross examination, the areas of dedifferentiated tumor cannot be distinguished from acinic cell carcinoma. However, these areas have differing histologic features, which may be present in varying amounts. The low-grade areas consist of round-to-polyhedral cells that can be arranged in acinar groups, solid cords, or diffuse sheets. Nuclei are round to oval, and the cytoplasm contains fine basophilic granules. Mitotic figures are rare. In contrast, the dedifferentiated portion of the tumor is composed of sheets of large, atypical cells that may appear syncytial. Nuclei contain one or more nucleoli, and there is abundant clear cytoplasm. Mitotic activity and central necrosis are frequent. Vascular and perineural invasion are also more common with dedifferentiated acinic cell carcinoma. Recently, Piana et al[2] described a case of dedifferentiation with morphologic and immunohistochemical features of myoepithelium.

Stanley et al[1] proposed that dedifferentiated acinic cell carcinoma develops from a preexisting low-grade acinic cell carcinoma through a process of dedifferentiation. Both El-Naggar et al,[3] using DNA flow cytometry, and Di Palma et al,[4] using image analysis, demonstrated an aneuploid DNA content in the dedifferentiated component of the tumor in contrast to classic acinic cell carcinoma, which is diploid. However, the molecular events surrounding this transformation are unknown. In 1997, Henley et al[5] investigated *p53* as a possible involved gene; however, p53 oncoprotein expression and point mutation analysis by polymerase chain reaction–single strand conformational polymorphism were negative in both areas of the tumor. Their work has been supported by Di Palma et al,[4] who were unable to demonstrate *TP53* mutations, microsatellite instability, or loss of heterozygosity at the *p53* locus.

Because these tumors behave in a more aggressive manner than is usual in acinic cell carcinoma, they should be treated as such with a radical resection that often includes the facial nerve; it may be necessary to follow this resection with a neck node dissection and/or radiotherapy. Prognosis is poor in comparison to classic acinic cell carcinoma; however, the exact biologic behavior of these tumors is uncertain, as, to our knowledge, only 10 cases[1,3,5] (including the current case) of this entity have been published in English literature.

References

1. Stanley RJ, Weiland LH, Olsen KD, et al. Dedifferentiated acinic cell (acinous) carcinoma of the parotid gland. *Otolaryngol Head Neck Surg.* 1988;98:155–161.
2. Piana S, Cavazza A, Pedroni C, et al. Dedifferentiated acinic cell carcinoma of the parotid gland with myoepithelial features. *Arch Pathol Lab Med.* 2002;126:1104–1105.
3. El-Naggar AK, Batsakis JG, Luna MA, et al. DNA flow cytometry of acinic cell carcinomas of major salivary glands. *J Laryngol Otol.* 1990;104:410–416.
4. Di Palma S, Corletto V, Lavarino C, et al. Unilateral aneuploid dedifferentiated acinic cell carcinoma associated with bilateral low-grade diploid acinic cell carcinoma of the parotid gland. *Virchows Arch.* 1999;23:465–472.
5. Henley JD, Geary WA, Jackson C, Wu CD, Gnepp DR. Dedifferentiated acinic cell carcinoma of the parotid gland: a distinct rarely described entity. *Hum Pathol.* 1997;28:869–873.