Kobe J. Med. Sci., Vol. 49, No. 2, pp. 45-49, 2003

Central Poorly Differentiated Adenocarcinoma of the Maxilla: Report of a Case

MASAHIRO UMEDA[1)], SATOSHI YOKOO[1)], YASUYUKI SHIBUYA[1)], TAKAHIDE KOMORI[1)], and CHIHO OBAYASHI[2)]

Department of Oral and Maxillofacial Surgery, Kobe University Graduate School of Medicine[1)]
Division of Surgical Pathology, Kobe University Hospital[2)]

Received 3 February 2003/ Accepted 14 April 2003

Key words: central carcinoma of the maxilla; poorly differentiated adenocarcinoma; minor salivary gland; treatment and prognosis

Central adenocarcinoma of the jaws is an extremely rare malignant tumor. We reported a case of poorly differentiated adenocarcinoma occurring intraosseously in the maxilla.

A 62-year-old male was referred to our hospital because of swelling of the palate. MRI showed a central tumorous lesion in the maxilla. He underwent maxillectomy combined with neck and parapharyngeal dissection. Histologic examinations of the surgical specimen revealed poorly differentiated adenocaricinoma showing a positive reaction for PAS, CEA and cytokeratin. He underwent adjuvant chemotherapy with cis-platinum diamminodichloride, 5-fluorouracil, and pirarubicin, but he died of multiple distant metastases 7 months after the surgery.

Most carcinomas of the oral cavity have their origin in the oral mucosa, but rarely develop intraosseously from malignant change in the epithelium lining of odontogenic cysts, epithelial remnants, or the epithelium of ectopic salivary glands. This intraosseous carcinoma (central carcinoma) sometimes occurs in the mandible but rarely in the maxilla.

Histologically, the majority of central carcinomas of the jaws are squamous cell carcinoma, mucoepidermoid carcinoma, and adenoid cystic carcinoma. We encountered a case of central carcinoma of the maxilla which was finally diagnosed as poorly differentiated adenocarcinoma histologically. We report the clinical and histological findings, and treatment of this extremely rare malignancy.

CASE REPORT

A 62-year-old male visited an otolaryngologist with a chief complaint of swelling of the right palate which he noticed a month ago. Biopsies were performed two times under the clinical diagnosis of a palatal tumor, but no definitive diagnosis was obtained. He was referred to our hospital.

Clinical examination revealed slight, diffuse swelling of the right palate with almost normal mucosa, except the biopsy wound (Fig. 1). MRI findings showed an intraosseous, tumorous lesion in the maxilla (Fig. 2). The maxillary sinus and nasal cavity were intact. There was no lymphadenopathy in the neck. A biopsy of the intraosseous lesion suggested poorly differentiated carcinoma.

He underwent radical operation about a month after the first visit. After a functional neck dissection of the right side, parapharyngeal dissection was performed using the mandibular

Phone: +81-78-382-5111 Fax: +81-78-382-6229 E-mail: ume@med.kobe-u.ac.jp

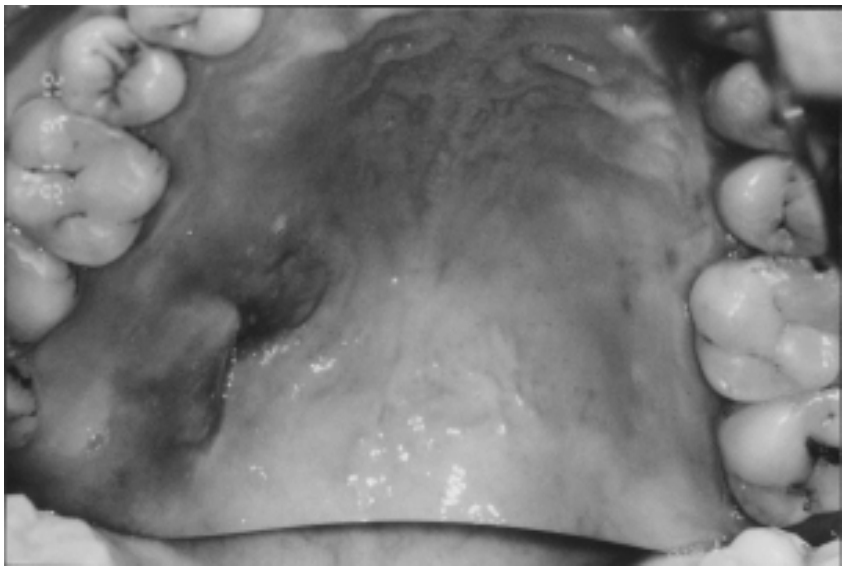

Fig. 1. Intraoral views show slight swelling of the palate. The ulceration was caused by the biopsy procedure performed at the former hospital.

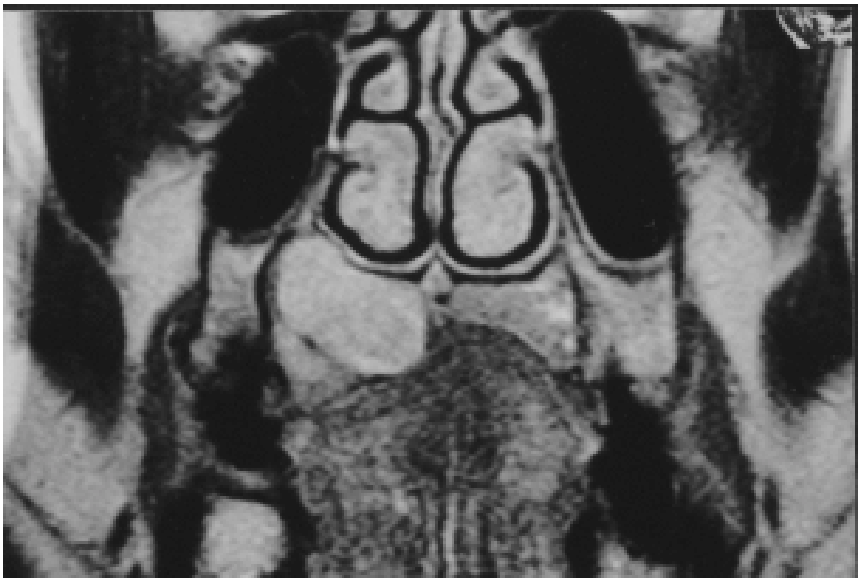

Fig. 2. MRI shows an intraosseous tumorous lesion with high signals in T1-weighted image contrasted by gadolinium.

swing approach, and right partial maxillectomy, except the orbital floor, was done en block with the neck and parapharyngeal tissues. Free forearm flap was transplanted to the raw surface of the cheek to minimize facial disfigurement and trismus. He further underwent two courses of adjuvant chemotherapy with cis-platinum diamminodichloride (CDDP), 5-fluorouracil (5FU), and pirarubicin (THP). Five months after the surgery, motor paralysis of the right hand appeared, and MRI examination revealed multiple brain metastases. Radiotherapy was performed for the brain metastases, but he died of multiple metastases 7 months after the surgery.

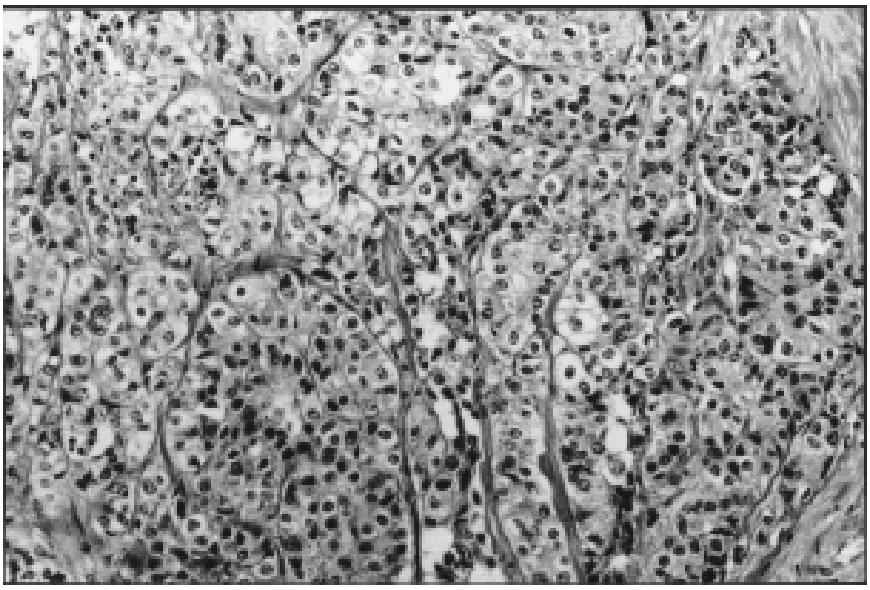

Fig. 3. Microscopic features showing proliferation of nested poorly differentiated caricnoma cells (HE stain, origical magnification x100).

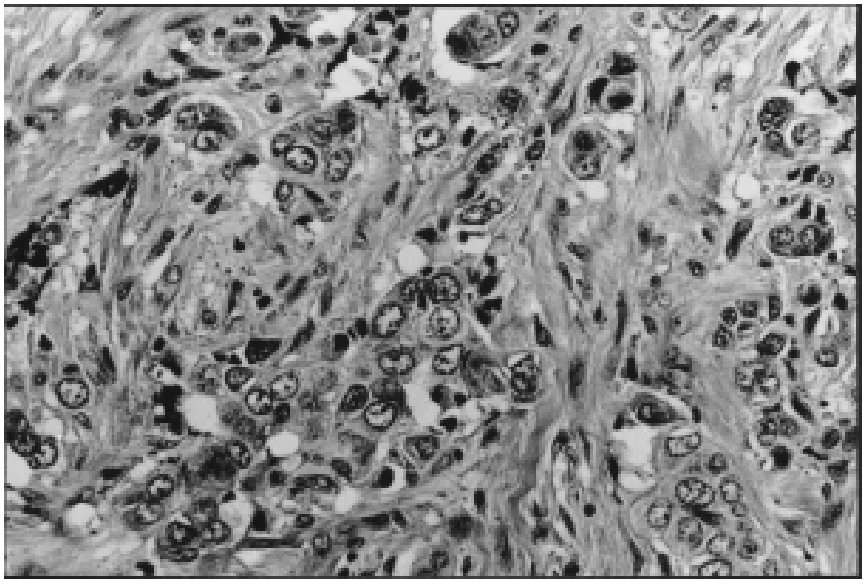

Fig. 4. Individually proliferating tumor cells with marked atypia (HE stain, origical magnification x200).

Histological examination of the surgical specimen revealed proliferation of poorly differentiated polygonal tumor cells with eosinophilic cytoplasm and round nucli showing marked atypia and many mitoses. Tumor cells formed subepithelial large nests but sometimes invaded individually in the proliferative spreadhead (Figs. 3 and 4). They showed glycogen granules by PAS stain (Fig. 5A), while intercellular bridge or keratinization, and intraepithelial replacemental growth of the tumor, corresponding to carcinoma in situ, were not observed.

Tumor cells showed a positive reaction for low-molecular weight cytokeratin (CAM 5.2) and CEA (Fig. 5B), but negative for α SMA. Although these histological and immunohistochemical findings did not indicate clearly the origin of the tumor, this lesion was finally diagnosed as poorly differentiated adenocarcinoma, probably originating in the minor salivary gland.

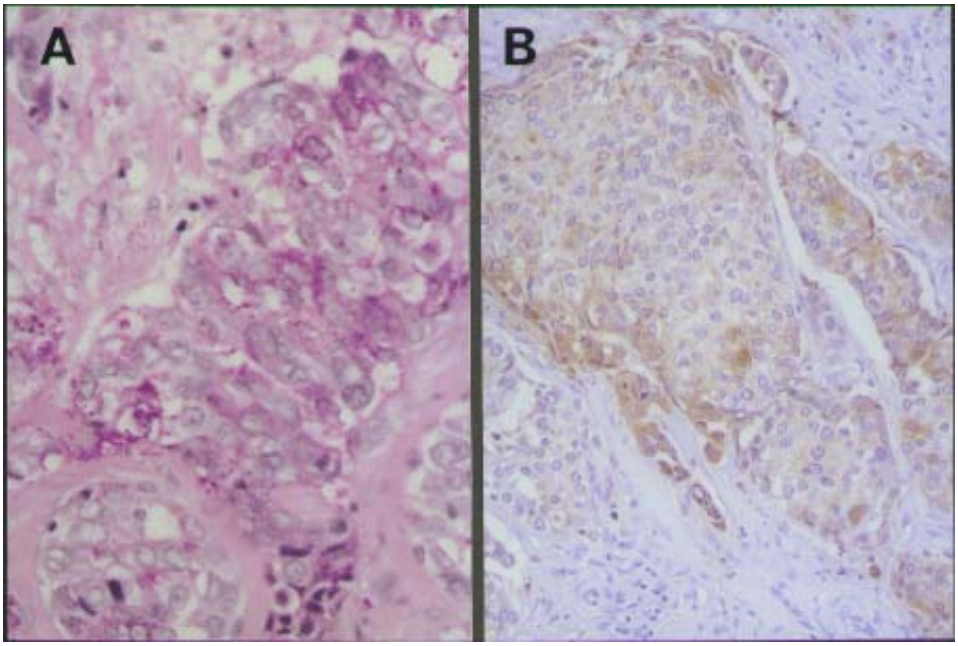

Fig. 5. The tumor cells showed glycogen granules by PAS stain (A) and positive reaction for CEA (B). (origical magnification, A x200, B x100).

DISCUSSION

Although most carcinomas of the oral and maxillofacial regions originate from the oral, nasal, or antrum epithelium, they rarely occur intraosseously and are designated as central carcinomas. Eversole et al. (2) reviewed 63 cases of central carcinoma of the jaws, 36 squamous cell and 27 mucoepidermoid carcinomas, and stated that only 7 occurred in the maxilla and the others occurred in the mandible. Brookstone et al. (1) and Ohtake et al. (4) also reported that central carcinomas occurred predominantly in the mandible rather than the maxilla.

Central carcinoma of the jaws originates from odontogenic cysts, remnants of odontogenic epithelium, or epithelium of ectopic salivary glands. The current case probably originated from the ectopic salivary gland because the tumor occurred in the palate where odontogenic cysts or odontogenic epithelium rarely exists. Although the cancer cells showed minimal differentiation toward both adenocarcinoma and squamous cell carcinoma, the tumor was finally diagsosed as poorly differentiated adenocarcinoma probably originating in the ectopic minor salivary glands.

According to the review by Brookstone et al. (1), the most frequent histology of 138 cases of central salivary gland carcinoma of the jaws was mucoepidermoid carcinoma followed by adenoid cystic carcinoma, while central adenocaricnoma was seen in only 6 patients. In 28 cases of central carcinoma of the jaws surveyed by Ohtake et al. (4) in the Japanese literature,

there were no cases of central adenocarcinoma. To our knowledge, this is only the second reported case of a central adenocarcinoma occurring in the maxilla; The first case was reported by Morgan et al. (3) in 1968.

En block resection of the primary tumor and metastasic lymph nodes is the principle surgical procedure for patients with oral cancer. However, in cases of maxillary cancer, the tumor and nodes are usually resected separately for anatomical reasons, so recurrence sometimes occurs in the parapharyngeal space located between the two regions. We reported previously the necessity of en block resection for patients with maxillary cancer with N+ necks (5). The current case was diagnosed to have an N0 neck, but considering the extremely aggressive biologic behavior of this type of malignancy, we performed radical surgery, including wide resection of the tumor combined with elective neck and parapharyngeal dissection. Further, postoperative chemotherapy was performed after a final diagnosis of poorly differentiated adenocarcinoma was made, but we could not cure the patient because of the aggressive dissemination of cancer cells to the multiple organs.

REFERENCES

1. **Brookstone, M.S. and Huvos, A.G.** 1992. Central salivary gland tumors of the maxilla and mandible: A clinicopathologic study of 11 cases with an analysis of the literature. J Oral Maxillofac Surg **50**:229-236.
2. **Eversole, L.R., Sabes, W.R., and Robin, S.** 1975. Aggressive growth and neoplastic potential of odontogenic cysts. With special reference to central epidermoid and mucoepidermoid carcinomas. Cancer **35**:270-282.
3. **Morgan, G.A., Morgan, P.R., and Murphy, H.J.** 1968. Adenocarcinoma of the maxilla, report of a case. Oral Surg **26**:651-653.
4. **Ohtake, K., Yokobayashi, Y., Shingaki, S., Nakajima, T., Ishiki, T., and Kawasaki, T.** 1989. Central carcinoma of the jaw. A survey of 28 cases in the Japanese literature. J Craniomaxillofac Surg **17**:155-161.
5. **Umeda, M., Minamikawa, T., Yokoo, S., and Komori, T.** 2002. Metastasis of maxillary carcinoma to the parapharyngeal space -rationale and technique for concomitant en bloc parapharyngeal dissection. J Oral Maxillofac Surg **60**:408-413.