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CASE REPORT

Adenoid cystic carcinoma associated with salivary duct cyst in the sublingual gland

Yuzo Ogawa[1], Mitsunobu Kishino[1], Mitsuhiro Nakazawa[2], Soichi Iwai[2], Yoshiyuki Ohta[2], Takeshi Ishida[3], Yoshiaki Yura[2], Naokuni Ijuhin[1]

Departments of [1]Oral Pathology and [2]Oral and Maxillofacial Surgery II, Osaka University Graduate School of Dentistry and [3]Clinical Laboratory, Osaka University Dental Hospital, Osaka, Japan

We described an extremely rare case of adenoid cystic carcinoma associated with salivary duct cyst in the sublingual gland of a 40-year-old Japanese woman. The tumor was growing from the cyst wall and almost occluded the cyst lumen. The epithelium lining the cyst lumen contained both keratin 19-positive cells and α-smooth muscle actin-positive cells, indicating the cyst being derived from the acinus/intercalated duct of the sublingual gland. Therefore, our case has presented for the first time a direct evidence that adenoid cystic carcinoma arises from acinus/intercalated duct.
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A 40-year-old Japanese woman was referred to the Osaka University Dental Hospital in December 2002, for examination of swelling in the left sublingual region that was first noticed by a dentist about 1 month prior to referral. It was a 34 mm × 14 mm, non-tender and mobile mass with elastic consistency (Fig. 1A). Saliva appeared to flow out normally from the left submandibular gland. Computed tomography, magnetic resonance imaging, and gallium scintigraphy suggested the mass to be a benign sublingual gland tumor, most probably pleomorphic adenoma. The excised mass was well encapsulated. On cut section, it was a large cyst without recognizable fluid content. Instead, the cystic space was almost filled with firm, gray-white tumors protruding from the wall (Fig. 1B). Histologically, the cyst wall was a dense fibrous connective tissue with a very mild chronic inflammatory cell infiltrate. The luminal surface was lined by thin, flattened, squamous or cuboidal epithelium (Fig. 2A,B). Adenoid cystic carcinoma had been developing from the epithelium (Fig. 2B,C), and consequently, the intact epithelium was seen only occasionally. A small piece of sublingual gland tissue was attached to the cyst wall. Although some ducts and lobules maintained normal morphology, other ducts were dilated and lobules exhibited a sign of chronic inflammation and mesenchymal atrophy (Fig. 2D). Immunohistochemistry revealed that the cyst epithelium contained α-smooth muscle actin (α-SMA) and keratin 19 (K19) (Fig. 3A,B). Adenoid cystic carcinoma cells sometimes expressed either of these proteins. In the sublingual gland, α-SMA was seen in the myoepithelial cells (MECs) around the acini and intercalated ducts, whereas K19 was found in the serous acinar and duct cells. Expression of these proteins by the carcinoma cells and salivary gland cells has been reported previously (1). As small tumor nests were observed outside the cyst wall in some sections, the patient underwent the second surgery. The tumor was not found in the resected tissue, including the rest of the sublingual gland that showed inflammation and atrophy of some lobules.

Comments

The present case is an extremely rare case and sheds light on the histogenesis of adenoid cystic carcinoma. Both adenoid cystic carcinoma and salivary duct cyst are rare in the sublingual gland. Adenoid cystic carcinoma, a relatively common salivary gland malignancy, most frequently occurs in the parotid, submandibular, and palatal minor glands. By reviewing 312 adenoid cystic carcinomas in major salivary glands, Tomich (2) showed that only seven cases (1.2%) occurred in the sublingual gland. The majority of salivary duct cyst involves the parotid gland (85%) and the submandibular gland (10%). The remainder occurs in various other salivary gland sites (3). Among 166 cases of salivary duct cysts in major salivary glands, only 1 case involved sublingual gland (4).

Salivary duct cyst is thought to occur secondary to duct obstruction and thus may be a form of retention cyst (3, 4). The fluid content of salivary duct cyst, however, is different from that of mucous retention cyst not only qualitatively but also quantitatively. The content varies in amount among salivary duct cysts (3). There was no recognizable fluid

Correspondence: Yuzo Ogawa, DDS, PhD, Department of Oral Pathology, Osaka University Graduate School of Dentistry, 1-8 Yamadaoka, Suita, Osaka 565-0871, Japan. Tel.: +81 6 6879 2892/3. Fax: +81 6 6879 2895. E-mail: ogawa@dent.osaka-u.ac.jp
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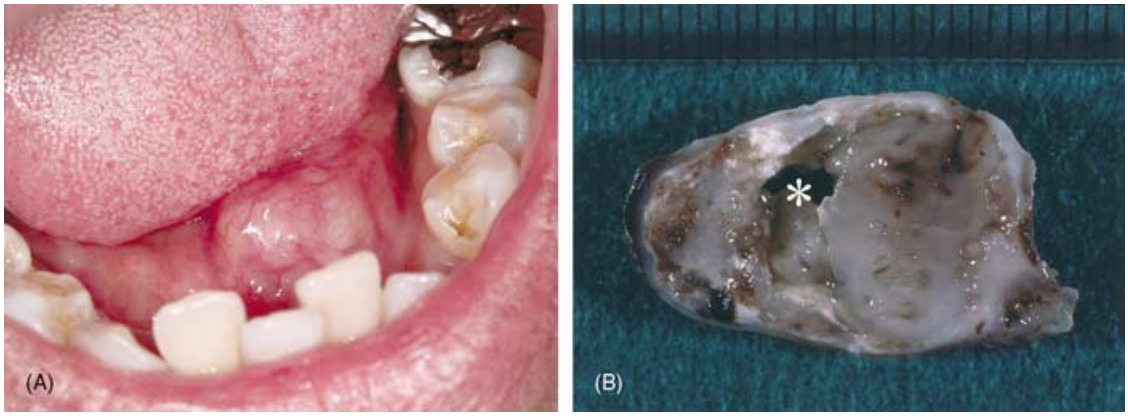

Figure 1 Sublingual mass without ulceration but with slight vascular markings (A) and cross-section of excised mass after fixation (B). An asterisk in B indicates cyst lumen. Each graduation of the ruler in B represents 1 mm.

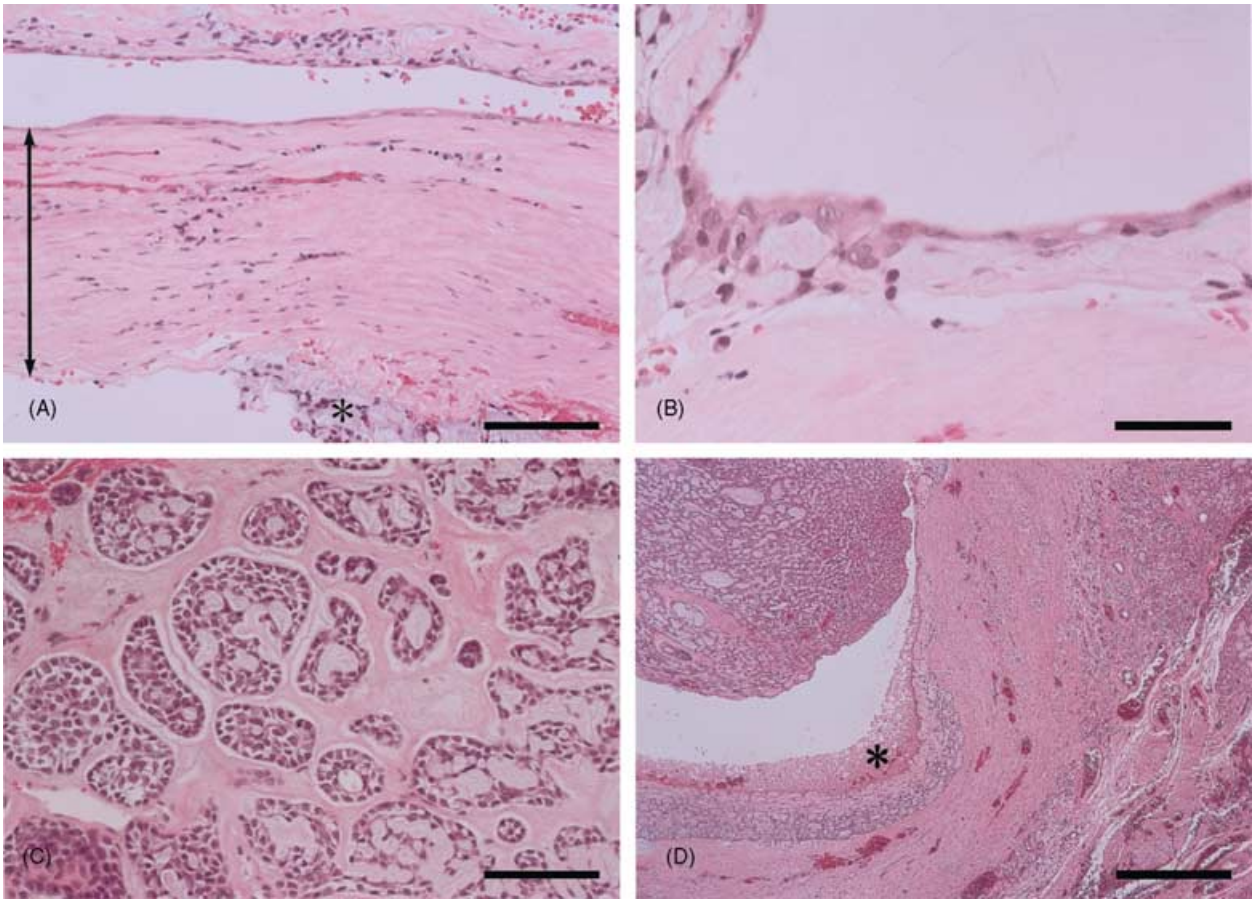

Figure 2 Hematoxylin and eosin staining. Cyst wall with normal epithelium (arrow in A), adenoid cystic carcinoma (C) developing from cyst epithelium (from right to left in B), and sublingual gland attached to cyst wall (right in D). Some lobules of the sublingual gland show chronic inflammation with parenchymal atrophy (D). Note a small nest of the tumor outside the cyst wall (asterisk in A) and a small amount of proteinaceous material attached to the luminal surface of the cyst (asterisk in D). Bars = 100 μm (A,C), 50 μm (B), and 500 μm (D).

content in the present cyst. Although the specific cause of duct obstruction is often not recognized, some cysts have been associated with benign and malignant tumors, post-operative or post-inflammatory strictures, calculi, and mucous plugs (3, 4). Although we cannot point out the exact cause(s) of the cyst formation, adenoid cystic carcinoma might have significantly contributed in the present case. The tumor that occurred in the acinus/intercalated duct (see below) might have slowly grown into the lumen and obstructed the salivary flow, resulting in the cystic dilatation. Slow growth is one of the characteristics of adenoid cystic carcinoma.

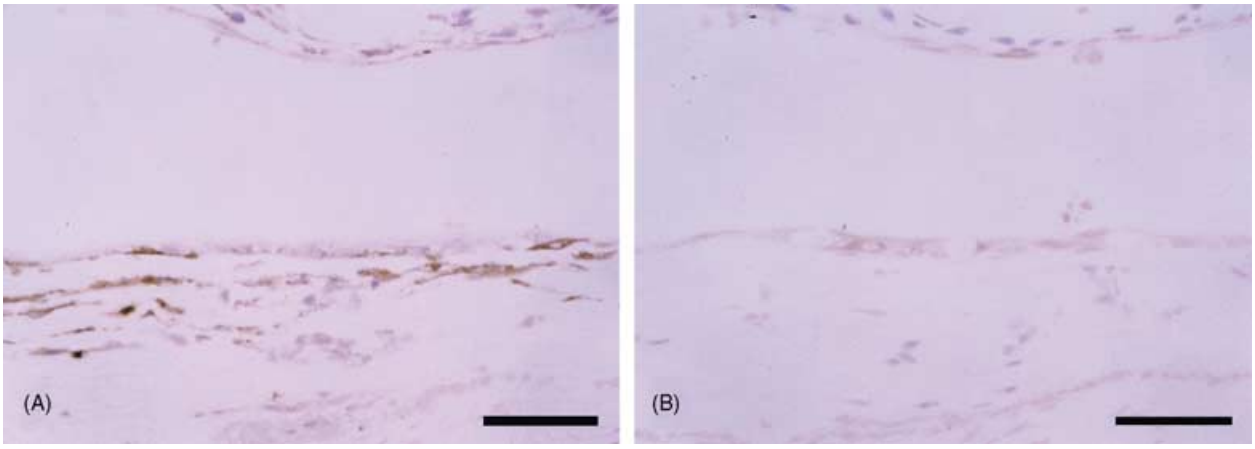

Figure 3 Immunohistochemistry for α-SMA (A) and K19 (B). Hematoxylin counterstaining. (A,B) and Fig. 2(A) are semiserial sections. Cyst epithelia with and without adenoid cystic carcinoma (upper wall and lower wall, respectively) express both α-SMA and K19. Myofibroblasts in the cyst wall are also positive for α-SMA (A). Bars = 50 μm.

To date, there are four documented tumors associated with salivary gland cysts: one basal cell adenoma associated with a lymphoepithelial cyst in the parotid gland; and three mucoepidermoid carcinomas associated with two mucous retention cysts in minor salivary glands and a salivary duct cyst in the parotid gland (5). Therefore, the present case is the first for adenoid cystic carcinoma associated with salivary duct cyst. Although adenoid cystic carcinoma cells are remarkably uniform in size, shape, and staining qualities, they show a differentiation toward either the luminal cell or the MEC in normal salivary gland (2, 6). Therefore, it is not unrealistic to assume that the tumor is derived from the acinus/intercalated duct. The present case has presented for the first time a direct evidence that adenoid cystic carcinoma arises from the acinus/intercalated duct. The epithelium of the cyst contained both K19-positive cells and α-SMA-positive cells. In the normal glands, only the acinus and the intercalated duct contain both of these cells.

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