Sclerosing Polycystic Adenosis of the Sublingual Gland: a Case Report

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Abstract: *A case of sclerosing polycystic adenosis involving the sublingual gland in a 60-year-old Chinese female is presented.*
Key words: *sclerosing polycystic adenosis, sublingual gland*

Sclerosing polycystic adenosis (SPA) was first recognised as a distinct sialadenopathological entity by Smith et al[1] in 1996. The authors reported nine cases and believed that this lesion represented a pseudoneoplastic process similar to fibrocystic changes that occur in the breast. The clinical presentation may simulate a slow growing tumour involving the salivary glands. The lesion is unencapsulated, showing tubulo-acinar adenosis with dilated ducts, apocrine metaplasia, epithelial hyperplasia and cystic changes associated with fibrosis[2-7]. According to recent reports by Gnepp and co-workers, to date approximately 40 cases have been described in the literature[6,7]. The majority of the lesions involved the parotid gland, three involved the submandibular gland, and two arose within the oral cavity in minor salivary gland sites. Herein, a case of SPA involving the sublingual gland in a 60-year-old Chinese female is reported.

Case report

A 60-year-old Chinese female was referred for treatment of a slowly growing, painless mass in her left sublingual region. She had noticed the swelling over 2 months. Her medical history was unremarkable. Clinical examination revealed a well-defined, immobile and moderate tender mass in the sublingual region, with no ulceration of the surface mucosa or skin and no facial nerve paralysis. Needle aspiration revealed mucinous content. The clinical impression was, therefore, of a benign lesion, possibly ranula, a dermorid/epidermoid cyst, or sialolithiasis. The lesion mass and the sublingual gland were both removed surgically. The post-operative course was uneventful and there were no signs of recurrence 6 months after the surgery.

Pathological findings

At gross examination, the tumour nodule was well-delineated, firm in consistency, and measured $2 \times 1 \times 1$ cm. The cut surface appeared yellow-grey in colour with needle-tip-like cystic spaces. Microscopically, the lesion demonstrated abundant sclerotic collagenous tissue that contained cystically ectatic ducts, smaller ductules and acinar elements arranged in a vaguely lobular pattern (Fig 1a). The accentuated sclerosing collagenous tissue showed distinct hyalinisation, especially surrounding the cystic ducts (Fig 1b). Compared with the normal sublingual gland, there were more oval and elongated or distorted ducts in the lesion. Atrophy of the salivary parenchyma was prominent. The lining cells of the ectatic ducts were flattened to cuboidal, unilayered or occasionally multilayered, and showed no signs of atypia or dysplasia (Fig 1c). Some dilated ducts contained small papilla that projected into the lumens (Figs 1b and 1c). Focally, the

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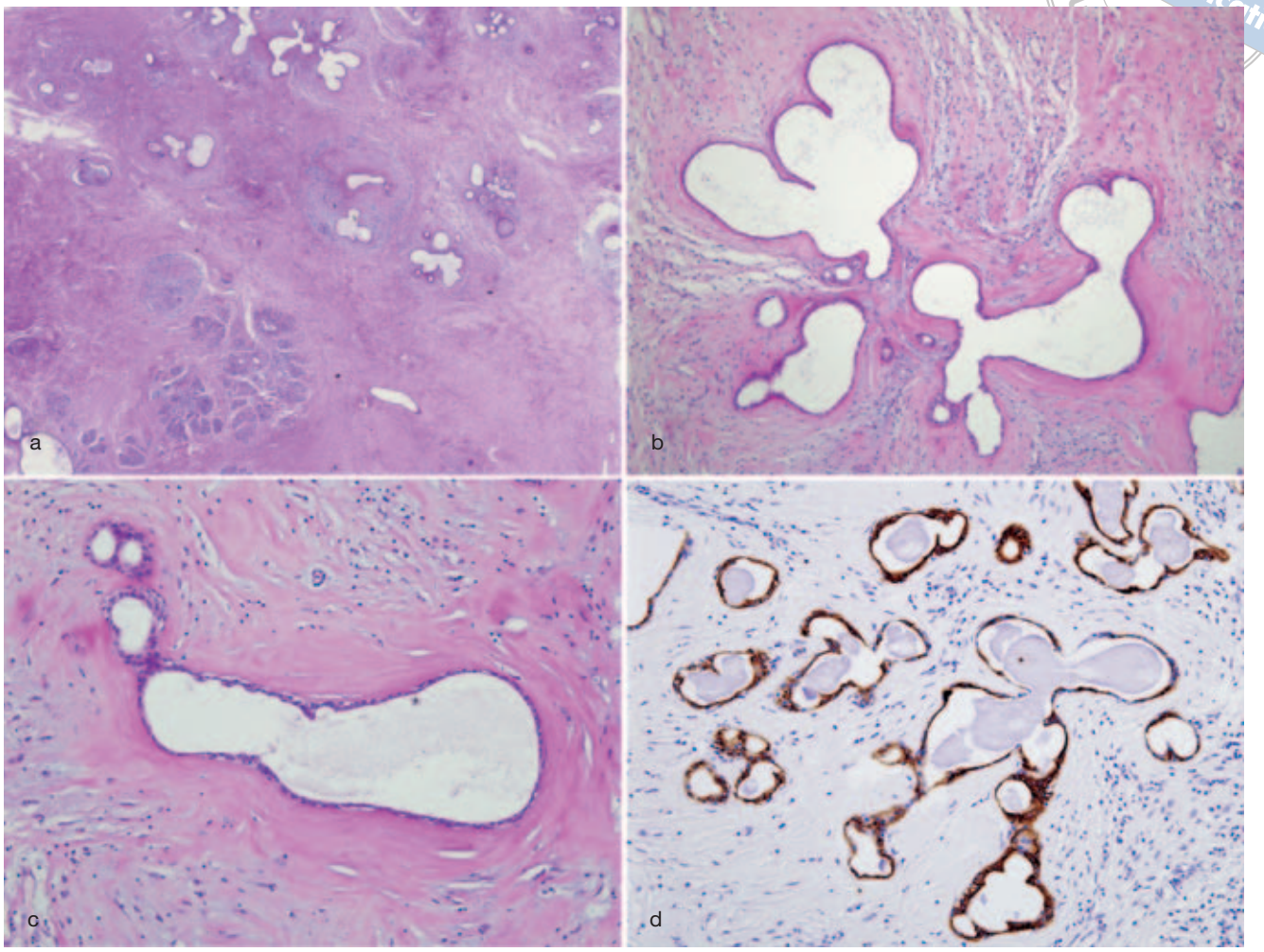

Fig 1 (a) SPA shows abundant sclerotic collagenous tissue that contains cystically ectatic ducts, smaller ductules and acinar elements arranged in a vaguely lobular pattern (haematoxylin-eosin [HE] stain, original magnification ×25). (b) The sclerotic stroma shows prominent hyalinisation, especially surrounding the cystic ducts. Areas of the ductal lining form papillary injections intruding into the lumens (HE stain, original magnification ×100). (c) The dilated ducts are lined by a unilayer of flattened epithelial cells showing no signs of atypia or dysplasia (HE stain, original magnification ×200). (d) Ductal epithelial cells are strongly positive for cytokeratin 19 (immunohistochemical stain, original magnification ×200).

cystically ectatic ducts were lined by some vacuolated cells. The stroma was hypocellular with focal lymphocytic infiltrates. Cytokeratin 19 (CK19) was recognised in ductal epithelial cells of the lesion (Fig 1d), which were also weakly positive for CK10 and 13. Smooth muscle antigen and S-100 protein were detected in flattened myoepithelial cells that surrounded ductal and acinar structures. Epithelial membrane antigen was positive in the luminal surface of ductal and acinar epithelium, but carcinoembryonic antigen was negative in the lesion areas.

Discussion

The histopathological features of the present case were similar to the cases reported previously, with the exception that present were relatively fewer cystically dilated ductal structures and a more prominent fibrosis with distinct periductal hyalinisation in the lesion. Other features such as apocrine-like metaplasia, hyperplastic intraluminal epithelium arranged in a cribriform pattern and ductal epithelial atypia that have been occasionally reported in SPA were not found in the present case. Compared with the parotid and the submandibular glands, the sublingual gland has an inconspicuous ductal system[8]. The intercalated ducts are short and difficult to recognise. Overall, the intralobular ducts are fewer in number in the sublingual gland[9]. These differences in normal structures among the major salivary glands may partly explain the slight variation in morphology of the present case. Immunostaining of smooth muscle actin and S-100

protein were found in the peripheral myoepithelial cells surrounding almost every ductal and acinar structure. The presence of a myoepithelial layer confirms that the hyperplastic ductal cells were within intact ducts. A strong reactivity for CK19 and a weak staining for CK10 and 13 in the cells lining the cystic ducts suggested that the epithelium of the dilated ducts was mostly a single layer.

The differential diagnosis of SPA should include polycystic disease, sclerosing sialadenitis and salivary gland tumours. Polycystic disease of the salivary glands displays a diffusely honeycombed, lattice-like network of cysts with inspissated intracystic secretions replacing the normal parenchyma, but unlike SPA, fibrosis is not prominent. Sclerosing sialadenitis has prominent fibrosis with varying degrees of chronic inflammation, but the fibrosis is not nodular and the proliferative epithelial component of SPA is not evident. The acinar cell and ductal proliferation of SPA may suggest the possibility of an acinic cell carcinoma, cystadenocarcinoma, and mucoepidermoid carcinoma. The consistent absence of metastasis, necrosis, infiltration, perineural or intravascular growth, and minimal mitotic activity or nuclear atypia suggest there is no malignancy. Some isolated areas of myxoid stroma in SPA may be suggestive of pleomorphic adenoma, but the lobular structure of SPA is typically maintained. The cystically dilated ducts, xanthomatous degeneration and nodular scar-like fibrosis in SPA are rarely seen in pleomorphic adenoma.

The pathogenesis of SPA is uncertain. Most histological features, such as ductal ectasia, sclerotic fibrosis, intraluminal epithelial proliferation, acinic cell hyperplasia, and metaplastic changes of the epithelium, point to a reactive post-inflammatory process. However, ductal hyperplasia and histological changes ranging from mild to severe dysplasia or carcinoma *in situ* have been documented in some SPAs[2,4,6]. To date, slightly less than 30% of reported SPAs have recurred, although no metastases or fatalities have been reported. Skálová et al[7] recently assayed six female cases of SPA using the polymorphism of the human androgen receptor locus as a marker and demonstrated the clonal nature of SPA. Therefore, some authors believe that SPA is a neoplasm, not just a reactive process[4,7].

In conclusion, SPA is a rare lesion involving the salivary glands as described recently in the literature. Although the clinical and histological features of the present case appear to indicate a benign lesion, the reported high recurrence rate, together with the presence of dysplasia and/or carcinoma *in situ* suggests that the possible low-grade malignant potential of SPA has yet to be fully characterised.

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