Sclerosing Polycystic Adenosis of Tongue

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Abstract

Sclerosing polycystic adenosis (SPA) is a rare benign lesion of the salivary glands which appears histologically similar to sclerosing adenosis and fibrocystic disease of the mammary gland. To date, 67 cases of SPA have been reported in the literature, with the lesion arising in the minor salivary glands in only 9. The present report describes the 10th case of SPA. The patient was a 39-year-old Brazilian man who presented with an asymptomatic nodule on the ventral surface of the tongue. Based on a clinical diagnosis of benign salivary gland neoplasm, an excisional biopsy of the lesion was performed. Histopathological examination showed lobular proliferation of ductal and acinar elements surrounded by a fibrosclerotic stroma. Many of the ductal structures exhibited cystic dilation and were surrounded by periductal fibrosis, which is consistent with SPA findings. No recurrence of the disease was observed after a 5-year follow-up. A literature review is also discussed, focusing on both the etiology of SPA and the treatment options available.

Key words: Minor salivary glands — Neoplasia — Polycystic adenosis — Dysplasia
**Introduction**

Sclerosing polycystic adenosis (SPA) is a rare lesion of the salivary glands which histologically resembles sclerosing adenosis and fibrocystic disease of the mammary gland\(^3,6,8,10–12,14\). It has been described in a wide age-range of patients, encompassing from childhood to the eighth decade of life (9 to 84 years), with a mean age of 42 years, and is slightly more prevalent in women than in men (ratio of 4 : 3)\(^3,6\). The major salivary glands are the most affected sites, that is, the parotid and submandibular glands\(^14\). Few studies have found SPA in the minor salivary glands (12.2%), so the presence of this lesion in these sites is considered very rare (Table 1)\(^6\).

Smith et al., in 1996, characterized the histological features of SPA as follows: lobular proliferation of aberrant ductal and acinus elements, cystic ducts with frequent apocrine-like and sebaceous-like cells, eosinophilic intracytoplasmic granules within some acinar-type cells, intraductal epithelial hyperplasia with occasional collagenous spherulosis, and dense fibrosis which is frequently nodular\(^10\).

The lesion can arise in both the major and minor glands, with the latter being less common. Thus far, over 67 cases have been reported, and most were associated with the parotid gland (80%)\(^5,6,10\).

Its pathogenesis is still uncertain, but a recent study by Skálová showed some clonal specimens. This moved the theory of a neoplastic origin to one of inflammatory-reactive origin, which was previously believed to be the case\(^12\).

Although less frequent, SPA of the minor salivary glands has been diagnosed in 10 patients, with involvement of the buccal mucosa observed in 4. There was no gender predilection, and the age of the patients ranged from 35 to 82 years. In all cases involving the minor glands, the lesions were unifocal\(^5,6,9,15\). Recurrence may occur, but it is more associated with incomplete excision of the lesion\(^5,6,8\).

When SPA affects the mucosa, the underlying submucosal takes on a whitish, brownish, or yellowish color. During palpation, the nodules are mobile and well circumscribed\(^3\). They may occasionally be multinodular, measuring from 0.3 to 7 cm in size\(^3\).

To our knowledge, this is the first study to report a case of SPA arising in the minor salivary glands of the tongue.

**Case Report**

A 39-year-old Caucasian man was referred to our department complaining of a 3-month history of a painless nodule measuring 10 mm in diameter on the ventral surface of the tongue (Fig. 1). The clinical differential diagnosis was benign salivary gland neoplasm and an excisional biopsy was performed.

| Table 1 Reported cases of sclerosing polycystic adenosis in minor salivary glands |
|---------------------------------|--------|--------|------------------|
| Author                         | Gender | Age   | Location       |
| Gnepp et al., 2006             | M      | 75    | Buccal mucosa  |
| Noonan et al., 2007            | F      | 48    | Buccal mucosa  |
| Meer and Altini, 2008          | M      | 80    | Floor of the mouth |
| Gurgel et al., 2010            | F      | 70    | Hard palate    |
| Su et al., 2013                | F      | 54    | Sinonasal tract |
| Park et al., 2013              | M      | 49    | Nasal septum   |
| Mokhtari et al., 2014          | M      | 60    | Retromolar pad area |
Histopathological examination showed a well-circumscribed nodule characterized by a subtle lobular proliferation of ductal and acinar elements surrounded by a dense, collagenous stroma (Fig. 2a, b). Many of the ductal structures exhibited cystic dilatation and were surrounded by periductal fibrosis (Fig. 2a). In focal areas, the lining of the ductal epithelium contained cells with abundant eosinophilic cytoplasm and exhibiting intraluminal snout-like projections (Fig. 2b), reminiscent of the decapitation secretion seen in apocrine glands. Focal ductal epithelial cells were characterized by ballooning degeneration, producing a histological appearance similar to that of sebaceous cells (Fig. 2b). Intracellular mucin was positive for periodic acid-Schiff stain (Fig. 2c and d).
A histological examination was consistent with SPA. The patient is free of the disease after 5 years of follow-up.

**Discussion**

A rare condition, SPA was first described by Smith *et al.* in 1996\(^{14}\). Currently, only 67 cases of SPA in the oral cavity have been reported\(^ {5} \). When considering reports involving the minor salivary glands, however, this paper is only the 10th to describe this condition, and the first to report SPA arising in the tongue.

According to the WHO 2017 classification, although this pathology belongs to the chapter on the salivary glands, it is categorized as “other epithelial lesions” due to similarities with fibrocystic changes and sclerosing breast adenosis\(^ {11} \). With regard to its etiology, some authors consider SPA to represent a pseudotumoral and inflammatory sclerosing process\(^ {3} \). On the other hand, other studies have demonstrated a monoclonal origin for this lesion, thus corroborating a possible neoplastic origin\(^ {2,12} \). In addition, because it resembles a fibrocystic lesion, breast adenosis, and different degrees of dysplasia, some authors consider SPA to be a low-grade malignant neoplasm\(^ {13} \).

In the minor salivary glands, SPA was reported in the buccal mucosa, muco-gingival sulcus, hard palate, floor of the mouth, and retromolar area\(^ {3,4,6,8} \). No study to date, however, has described this condition in association with the minor salivary glands of the tongue, as is done in the present case.

Although the growth pattern of the nodules in such cases is slow and painless, some patients have reported pain as a change in sensitivity, usually manifesting as a tingling sensation\(^ {2} \).

The standard histological findings of SPA include lobular proliferation of ductal and acinar cells accompanied by ductal cystic dilation associated with apocrine and sebaceous metaplasia. It may also present ductal epithelial hyperplasia and atypia that may range from mild to severe dysplasia/carcinoma \textit{in situ}\(^ {1,5} \). An important characteristic of these lesions occurs due to the presence of acini filled with dispersed zymogen and reddish granules\(^ {15} \). Immunohistochemistry may be positive for cytokeratin, epithelial membrane antigen, S100 proteins, cystic disease protein fluid, or smooth muscle alpha cells. The KI-67 index score is generally low (1 to 2%)\(^ {1,5} \). In cases of suspected malignancy, microscopic examination of SPA lesions reveals a lobular pattern of growth and an absence of associated tissue invasion. Immunohistochemistry should be positive for peripheral myoepithelial cells as a diagnosis of malignancy can be disregarded\(^ {1,5} \).

Surgical excision is the treatment of choice for SPA. There is no consensus, however, on the exact type of surgical procedure best employed, that is, excision or resection. Local recurrence can occur in approximately 11 to 29% of cases, with a lower incidence in cases where the associated glands have been partially removed. In other words, recurrence is more strongly associated with inadequate removal or multifocality of SPA. In addition, in cases of relapse, studies have shown that the tumor can recur even after 5 years postoperatively\(^ {1,5,6} \).

Malignant transformation of SPA has been described in just one case report, in which the development of ductal carcinoma from a parotid gland was observed some 33 years after the initial diagnosis\(^ {1} \). A few authors have also reported malignancy and evolution towards low-grade carcinoma in approximately 2 to 3% of cases\(^ {1,3} \). This incidence may be considered under-diagnosed, as follow-up data are poorly reported in the literature, however\(^ {3} \).

Sclerosing polycystic adenosis in the minor salivary glands is extremely rare. Effective treatment is based on conservative surgical excision, with the literature reporting excellent prognosis and no recurrences rates.
Compliance with Ethical Standards

1. Conflict of interest
The authors declare no conflict of interest.

2. Funding
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3. Ethical approval
All the procedures herein reported were in accordance with the ethical standards of the institutional and/or national research committee involved and the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

4. Informed consent
Informed consent was obtained from all individual participants included in the study.

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