Case Report

Sclerosing polycystic adenosis of minor salivary glands: Report of a rare case with diagnostic approach and review of literature

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Abstract

Sclerosing polycystic adenosis (SPA) is an uncommon entity occurring in the salivary glands, with majority of the cases reported in major salivary glands reminiscent of fibrocystic disease of the breast. SPA arising in minor salivary glands of the oral cavity constitutes an exceedingly rare phenomenon. Here, we report a case of SPA that presented as a solitary, submucosal mass on the left lower labial mucosa in a 19-year-old male. The pathology features and a clinicopathologic diagnostic approach highlighting key features are discussed here. Similar cases published in the English literature are reviewed.

Keywords: Mouth, salivary glands, sclerosing polycystic adenosis

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INTRODUCTION

Sclerosing polycystic adenosis (SPA) is a rare, reactive/neoplastic, inflammatory process of the major or minor salivary glands (MSG) reminiscent of sclerosing adenosis of breast.[1] Since its first description, only over sixty cases have been reported worldwide, with majority occurring in major salivary glands. Only about twelve cases have been reported in MSG. Here, we present a case of SPA of MSG in a 19-year-old male patient.

CASE REPORT

A 19-year-old male patient reported with a slow-growing painless swelling in the lower labial mucosa of 7 months' duration. There was no history of trauma at the site. The family history and medical history were unremarkable and noncontributory. There was no history of lip sucking or lip biting either. On examination, a well-defined solitary, round and soft-to-firm submucosal swelling approximately 1.5 cm × 1.5 cm in size was present on the left lower labial mucosa [Figure 1a]. The mass was homogenous in consistency, partly movable with no associated tenderness or ulceration. The overlying mucosa was of normal color. The lesion was excised under LA and sent to the department of oral pathology and microbiology for histopathological and immunohistochemical analysis. On gross examination, the specimen was smooth surfaced, was grayish white and was firm, measuring about 1.5 cm × 1 cm [Figure 1b]. The cut surface was firm and typically demonstrated a well-circumscribed, mass containing multiple, tiny cysts.

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Histopathologic examination revealed unencapsulated but well-delineated nodules characterized by a subtle lobular proliferation of ductal and acinar elements surrounded by a densely sclerotic, collagenous stroma. The most remarkable feature was the presence of variably sized collections of ducts, many of which were cystically dilated and contained variable amounts of intraluminal, eosinophilic material. The small-caliber ducts and dilated larger ducts were lined by flattened-to-cuboidal epithelium, with occasional mucin containing vacuolated and apocrine-like metaplasia, and were surrounded by abundant hypocellular hyalinized collagenous tissue with focal lymphocytic infiltrate. Some of the ducts exhibited hyperplasia [Figure 2a-e]. Normal salivary gland acini were frequently found between and inside of the lobules, with occasional acini demonstrating fine, barely discernible eosinophilic granules. Sections were also subjected to immunohistochemical staining by calponin (myoepithelial marker) [Figure 2f] and periodic acid–Schiff (PAS)-diastase staining as an adjunct to aid the diagnosis. Some acinar cells showed PAS positive-diastase-resistant eosinophilic granules within the cytoplasm. In addition, a continuous layer of calponin-positive myoepithelial cells was found in most ducts and serous acini. Based on these observations, a final confirmatory diagnosis of SPA was rendered.

DISCUSSION

Salivary gland lesions represent one of the most arduous and perplexing domains in the field of diagnostic pathology. These include various benign and malignant salivary gland neoplasms as well as a plethora of nonneoplastic lesions which often emerge as an enigma to even the most experienced pathologists. In general, the clinical similarity between neoplastic and nonneoplastic lesions, the morphological diversity and histologically overlapping features can make differentiation between these lesions difficult to ascertain.

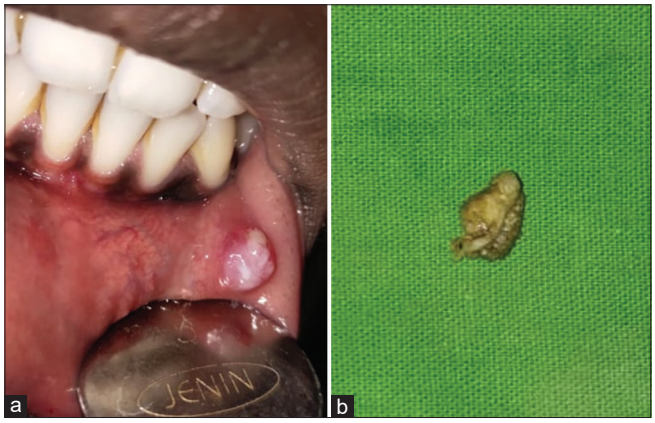

Figure 1: (a) A well-defined solitary, round, soft-to-firm submucosal swelling on the left lower labial mucosa. (b) Gross specimen was smooth surfaced, was grayish white and was firm

Westra *et al*. conducted a second review of histopathologic diagnoses made elsewhere and found that out of a total of 97 cases of salivary gland lesions, nine were re-diagnosed as a different lesion and in 8 of these cases it led to a modification of prior therapy.[2] This review presents a novel entity of SPA arising in salivary glands, which in addition to the aforementioned complexities is further marred by its unusual appearance and the enormous unfamiliarity among the pathologists to perceive it as a distinct lesion. Currently, this lesion is categorized as a nonneoplastic process in the latest WHO classification of salivary gland lesions. Only about sixty cases have been reported worldwide, mostly arising in the parotid gland, out of which only 12 have occurred in MSG of the oral cavity. This report and review describes only the second case of SPA arising in minor glands of the lower lip.

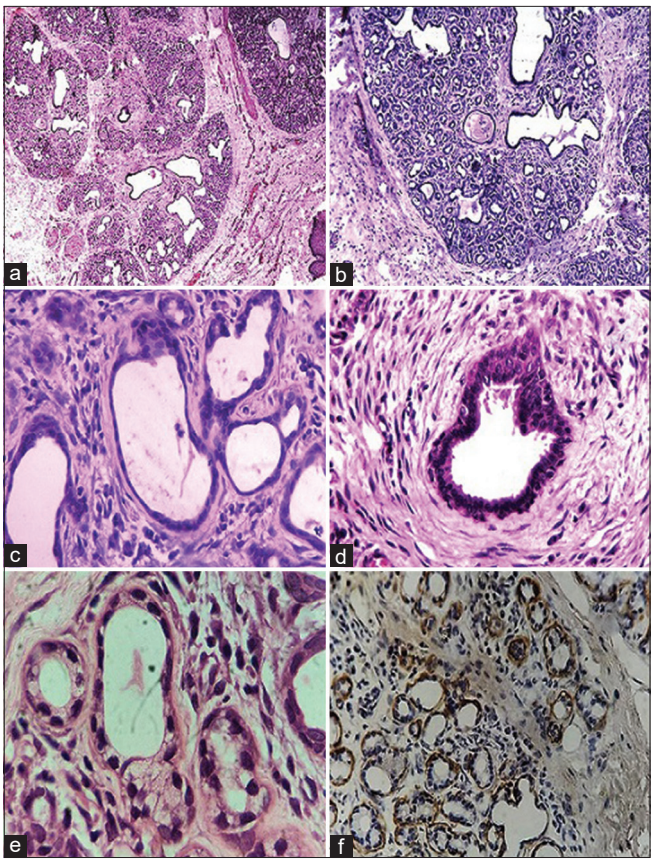

Figure 2: Histopathological image showing (a) lobular arrangement of variably sized microcysts in a densely sclerotic background (×40, H and E). (b) Cystically dilated ductal structures with mild chronic inflammatory infiltrate (×100, H and E), (c) dilated ducts lined by flattened-to-cuboidal cells with eosinophilic cytoplasm and few areas showing ductal hyperplasia (×400, H and E), (d and e) apocrine-like and mucinous metaplasia of duct (×400 H and E), (f) calponin stain demonstrating prominent peripheral staining of myoepithelial cell layer (×100, immunohistochemistry)

Table 1: Review of documented cases of sclerosing polycystic adenosis of the minor salivary glands

Author	A/G	Location	Clinical presentation	Histopathology												Immunohistochemistry
				Well circumscribed	Lobules	Variably sized cystic spaces	Ductal epithelium				Stroma			Acini		
							Cells	Apocrine/ squamous/ mucous metaplasia	Hyperplasia	Atypia	Hylinized sclerotic	Periductal fibrosis	Inflammatory infiltrate	Presence	Eosinophilic granules	
Gnepp *et al.*	NA	Buccal mucosa	Mass	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA
Noonan *et al.*	48/ female	Maxillary left mucobuccal fold	Asymptomatic firm, freely movable nodule	+	+	+	Cuboidal	+	−	−	+	+	Sparse	+	+	−
Noonan *et al.*	80/ male	Floor of mouth	Asymptomatic firm, submucosal mass	+	+	+	Cuboidal	−	+ (cribriform)	−	+	+	Sparse	+	NA	−
Noonan *et al.*	70/ male	Hard palate	Asymptomatic pink, doughy-firm sessile mass	+	+	+	Flattened to cuboidal	+	−	−	+	+	Sparse	+	+	−
Meer and Altini *et al.*	35/ male	Buccal mucosa	Single, firm, nontender nodule	+	+	+	Flattened	+/−/−	−	−	+	+	Moderate	+	−	CK [AE1/AE3] + S100 + P63 +
Swelam *et al.* (3 cases)	−	2 in retromolar area, 1 in palate	Asymptomatic submucosal swellings	+	+	+	Flattened	+/−/−	+	−	+	+	Mild	+	−	+ for S100, Ki67 EBV, Bcl-2
Mokhtari *et al.*	60/ male	Retromolar pad	Swelling	+	+	+	Flattened to cuboidal	+/−/+	+ (cribriform, Roman bridge)	−	+	+	Sparse	+	−	Alpha SMA+ Ki 67 <1 %
Ogasawara *et al.*	59/ female	Buccal mucosa	Single, firm nontender 1.5 cm nodule	+	+	+	Flattened	+/−/−	+ (papillary, cribriform, Roman bridge)	+ (mild)	+	+	Sparse	−	−	+ for EMA, CK7, CK14 − for ER, PgR, erbB2/ Her2
Puranik *et al.*	70/ male	Lower lip	Solitary, soft-to-firm submucosal swelling	+	+	+	Flattened to cuboidal	+/+/+	−	−	+	+	Sparse	+	+	S100+ Calponin+
Mumtaz *et al.*	47/ female	Buccal mucosa	Asymptomatic, slow-growing lump	+	+	+	NA	−/−/−	−	−	−	−	−	−	−	−
Present case	19/ male	Lower lip	Solitary, soft-to-firm submucosal swelling	+	+	+	Flattened to cuboidal	+/−/+	+	−	+	+	Sparse	+	+	Calponin+ ER− PgR−

+: Positive, −: Negative, NA: Not applicable

Clinically, the lesion presents as a slow-growing, painless mass, simulating a wide array of benign and malignant neoplasms and other nonneoplastic lesions which also tend to occur in a similar fashion. Furthermore, SPA has diverse histological features. A review of SPA lesions arising in MSG was conducted including the present case, bringing the total count to 13 [3-9] [Table 1]. The histopathologic differential diagnosis consists of a variety of lesions such as polycystic (dysgenetic) disease presenting a diffusely honeycombed, lattice-like network of variably sized cysts with inspissated intracystic secretions replacing the normal parenchyma, with only small clusters of residual acini. Unlike SPA, fibrosis is not prominent and ductal and acinar proliferation is not seen. Sclerosing sialadenitis has prominent fibrosis with varying degrees of chronic inflammation; however, unlike SPA, the fibrosis does not form nodules, the salivary gland parenchyma is atrophic without ductal or acinar hyperplasia, and the cystic changes are usually minimal. The proliferation of ducts with surrounding basement membrane may simulate pleomorphic adenoma. Here, a lobulated growth pattern along with a definite mesenchymal component weighs against that diagnosis. Because acinic cell and ductal proliferation is frequently found in SPA, acinic cell carcinoma (ACC) can be a diagnostic pitfall. However, in SPA, the lobular architecture is typically maintained; the ducts are rimmed partially or completely by myoepithelial cells and the invasive,

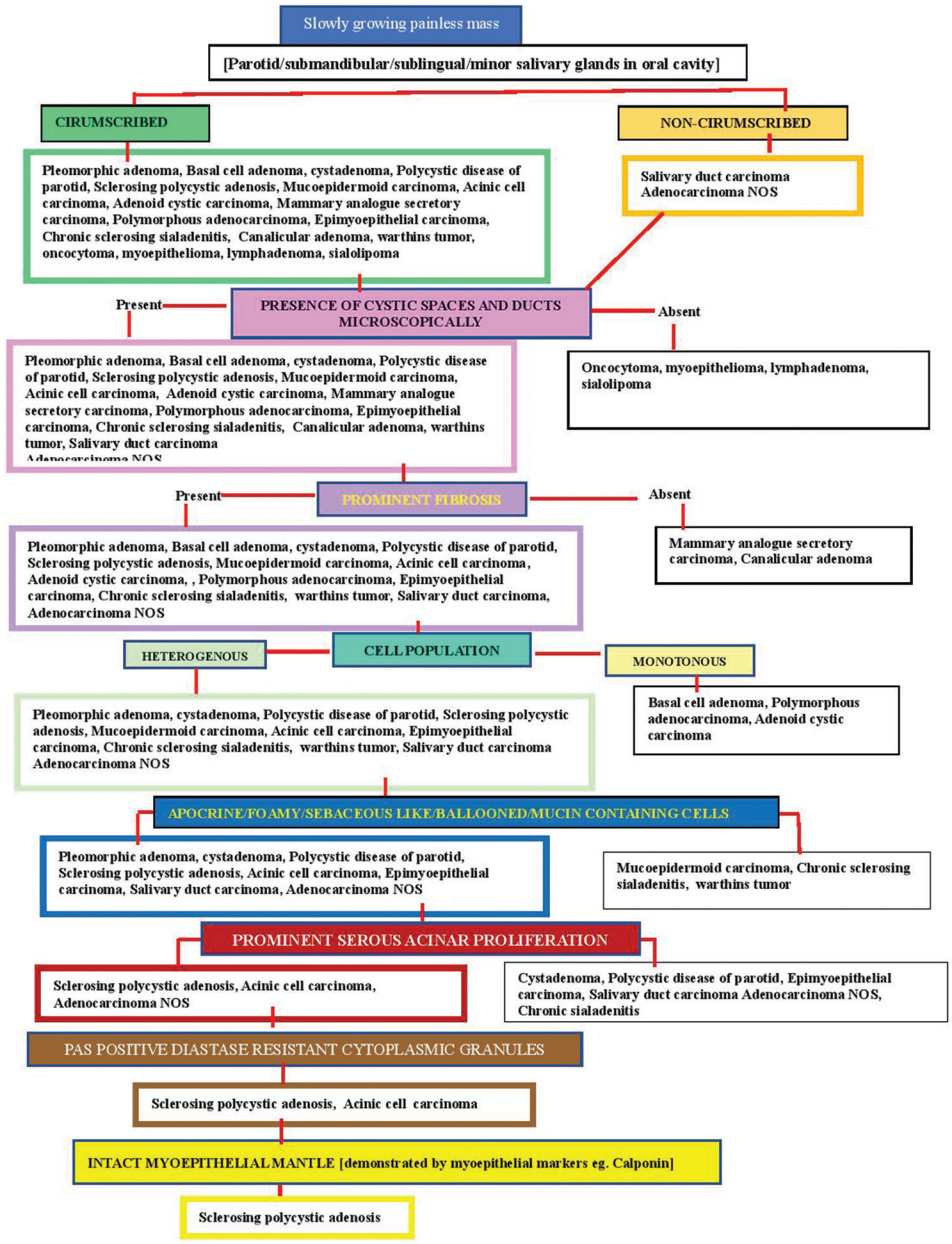

Figure 3: Clinicopathological diagnostic approach for sclerosing polycystic adenosis

destructive growth of a carcinoma is lacking. In ACC, the myoepithelial markers are negative. The focal papillary and prominent cystic components suggest considering a low-grade cystadenocarcinoma (currently classified under adenocarcinoma NOS in the WHO classification) in the differential diagnosis. However, the cystic pattern of the latter is typically more complex, with areas of invasive growth and a peripheral myoepithelial layer is absent.[4,10]

In effect, meticulous analysis of the previously reported cases elucidated few pivotal attributes of SPA that are fundamental to the diagnosis of this entity. To begin with, presence of circumscription; prominent dilated cystic spaces and a heterogeneous cell population that subsumes apocrine, foamy, sebaceous, squamous cell differentiation in a densely sclerotic stroma can yield significant clues to diagnose as well as curtail the multifarious lesions of salivary gland origin which may confound its recognition. In addition, identification of prominent zymogen like eosinophilic granules in serous acinar cells constitutes a vital feature of SPA that simulates ACC, although in SPA, the granules are much coarser and at times attain globular appearance. However, in our case, these granules were not very conspicuous on routine histopathological examination. PAS-diastase staining revealed mild positivity in focal areas. These findings are in agreement with the case of SPA in buccal mucosa reported by Ogasawara *et al*., showing no evidence of acinar-type cells with eosinophilic granules.[11] Nonetheless, careful exploration of acinar cells studded with dot-like eosinophilic granules should aid in the recognition of the lesion. Finally, the hallmark of SPA is the presence of an intact and continuous rim of myoepithelial cells surrounding cystic spaces and ducts as demonstrated by various myoepithelial markers such as calponin and alpha smooth muscle actin.

As such, the onus of clinching the accurate diagnosis rests upon careful observation of various histopathological features. This article makes a modest attempt to delineate a clinico-histopathological diagnostic approach for differentiating SPA from its close mimics[12] [Figure 3].

Treatment should consist of complete surgical excision with good surgical margins. Recurrences, sometimes multiple, have been reported quite frequently (19% as summarized by Gnepp).[4] In conclusion, SPA is a rare benign salivary gland lesion whose differential diagnosis includes a nonneoplastic as well as a variety of benign and malignant salivary gland tumors, particularly those with cystic and oncocytic features. In the major glands, SPA with focal epithelial dysplasia ranging from mild atypia to low-grade ductal carcinoma *in situ* has been reported.[13] Thus, familiarity with this entity is very important because many of its features overlap with those of other more commonly encountered and clinically significant salivary gland neoplasms.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Gnepp DR. Salivary gland tumor "wishes" to add to the next WHO Tumor Classification: Sclerosing polycystic adenosis, mammary analogue secretory carcinoma, cribriform adenocarcinoma of the tongue and other sites, and mucinous variant of myoepithelioma. Head Neck Pathol 2014;8:42-9.
2. Westra WH, Kronz JD, Eisele DW. The impact of second opinion surgical pathology on the practice of head and neck surgery: A decade experience at a large referral hospital. Head Neck 2002;24:684-93.
3. Swelam WM. The pathogenic role of Epstein-Barr virus (EBV) in sclerosing polycystic adenosis. Pathol Res Pract 2010;206:565-71.
4. Gnepp DR, Wang LJ, Brandwein-Gensler M, Slootweg P, Gill M, Hille J. Sclerosing polycystic adenosis of the salivary gland: A report of 16 cases. Am J Surg Pathol 2006;30:154-64.
5. Mokhtari S, Atarbashi Moghadam S, Mirafsharieh A. Sclerosing polycystic adenosis of the retromolar pad area: A case report. Case Rep Pathol 2014;2014:982432.
6. Noonan VL, Kalmar JR, Allen CM, Gallagher GT, Kabani S. Sclerosing polycystic adenosis of minor salivary glands: Report of three cases and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;104:516-20.
7. Meer S, Altini M. Sclerosing polycystic adenosis of the buccal mucosa. Head Neck Pathol 2008;2:31-5.
8. Puranik RS, Shree VB, Puranik SR, Anigol PS. Sclerosing polycystic adenosis of lower lip: A new and rare salivary gland entity. J Oral Maxillofac Pathol 2018;22:263-5.
9. Mumtaz S, Ali A, Singh M. Sclerosing polycystic adenosis of the oral cavity. Br J Oral Maxillofac Surg 2018;56:753-4.
10. Gnepp DR, Henley JD, Simpson RH, Eveson J. Salivary and lacrimal glands. In: Gnepp DR, editor. Diagnostic Surgical Pathology of the Head and Neck. 2nd ed. Philadelphia, PA: Saunders Elsevier Inc.; 2009. p. 432-34.
11. Ogasawara T, Kurosaka M, Jodai H, Kikuchi K, Ide F, Kusama K. Sclerosing polycystic adenosis with intraluminal crystalloids of the buccal mucosa: A case report and review of the literature. J Oral Maxillofac Surg Med Pathol 2015;27:580-7.
12. Chan JK, Cheuk W. Tumors of the salivary glands. In: Fletcher CD, editor. Diagnostic Histopathology of Tumors. 4th ed. China: Elsevier Health Sciences; 2007.
13. Skálová A, Michal M, Simpson RH, Stárek I, Prádná J, Pfaltz M. Sclerosing polycystic adenosis of parotid gland with dysplasia and ductal carcinoma in situ. Report of three cases with immunohistochemical and ultrastructural examination. Virchows Arch 2002;440:29-35.