CASE REPORT

Sclerosing Polycystic Adenosis: A Rare and Unusual Lesion of the Salivary Gland

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Abstract

Background: Sclerosing polycystic adenosis (SPA) is a rare, benign, and reactive inflammatory lesion of salivary glands. The distinctive pseudoneoplastic appearance resembles those of the benign fibrocystic disease of the breast. Most of the cases have been reported in the major salivary glands especially in the parotid gland (80%) with a wide age range and equal sex predilection. Only sporadic cases have been reported in minor salivary glands of the nasal septum, buccal mucosa, hard palate, floor of the mouth, retromolar pad, and lacrimal gland. Varying degrees of ductal atypia and recurrences are also reported in this lesion.

Case presentation: A 65-year-old male patient is presented with a pinkish painless mass at the junction of the hard and soft palate since 12 years.

Management and prognosis: The patient underwent a local surgical excision and a histopathology revealed sclerosing polycystic adenosis. The prognosis was good. No recurrence was noted on 12 months follow-up.

Conclusion: Though SPA is a benign and reactive inflammatory lesion of the salivary gland with a low neoplastic potential, this rare case is reported to differentiate it from other more clinically sinister salivary gland pathologies.

Keywords: Hard palate, Minor salivary glands, Sclerosing polycystic adenosis.

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Introduction

Sclerosing polycystic adenosis (SPA)—also synonymous with sclerosing polycystic adenoma—is a newly added, benign and reactive inflammatory lesion of salivary glands that can simulate neoplasm both clinically and histologically with a striking resemblance to the benign fibrocystic disease of the breast.[1] In recent WHO 2017 classification, the lesion was for the first time included in the "other epithelial lesion" category.[2] Most of the reported cases occurred within the major salivary glands, especially in the parotid gland (80%), and only sporadic cases have been reported within the minor salivary glands.[3–5] This is an additional case of SPA, involving the minor salivary glands at the junction of hard and soft palate.

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Case Description

A 65-year-old male patient is presented with a painless mass in the hard palate for the past 12 years. On examination, a pale pink, solitary, smooth, and spherical swelling of approximately 1.5 × 1 cm size was noted at the junction of the hard and soft palate. On palpation, the lesion was firm in consistency, pedunculated, nontender, nonindurated, and non-ulcerated. The patient's medical history was noncontributory. The provisional diagnosis included was torus palatinus, fibroma, and benign salivary gland lesion or nerve tissue tumor. An excisional biopsy was performed under local anesthesia and submitted for a histopathological examination.

On a gross pathological examination, the lesion appeared dirty white, well-circumscribed with a smooth surface, measured about 1.4 × 1.2 × 0.9 cm, and had a firm and rubbery consistency.

On microscopic examination, under a low-power view, the lesion was well-circumscribed with a partial pseudocapsulation. The lesional tissue showed numerous acinar and ductal proliferations, and the ductal structures were irregular and variable sized and showed cystic changes surrounded by a sclerotic/hyalinized stroma (Fig. 1).

On a high power view, many of the ducts exhibited a cystic dilatation and were lined by flattened epithelial cells with occasional foamy and clear cells (Fig. 2A). Eosinophilic coagulum was noted within the dilated ducts (Fig. 2B). There was no dysplasia/cytological atypia in the given section. The lipocytic component was also seen within the stroma. A focal sparse collection of chronic inflammatory cells composed of lymphocytes and plasma cells were also noted in the stroma (Fig. 3A). Residual minor salivary glands were noted at the periphery of the lesion (Fig. 3B). A few brightly stained eosinophilic zymogen granules were seen in some of the tubuloacinar structures. On the basis of the histopahological examination, the final diagnosis of sclerosing polycystic adenosis was made.

The section was also subjected to a special staining with periodic acid-Schiff (diastase resistant) (PAS-D) and mucicarmine, which demonstrated the presence of an intracellular mucin. The

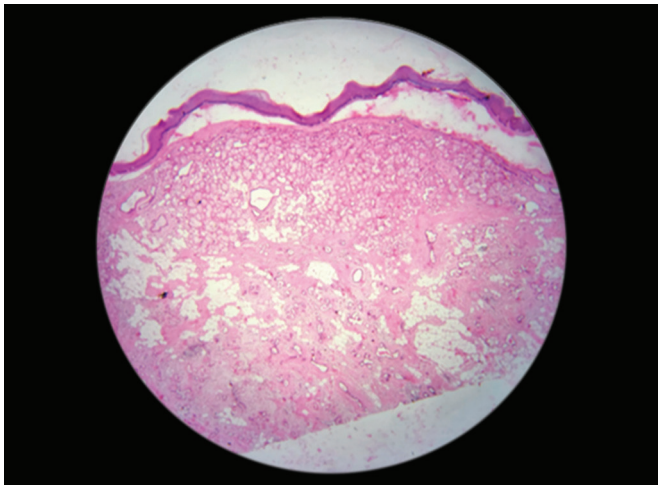
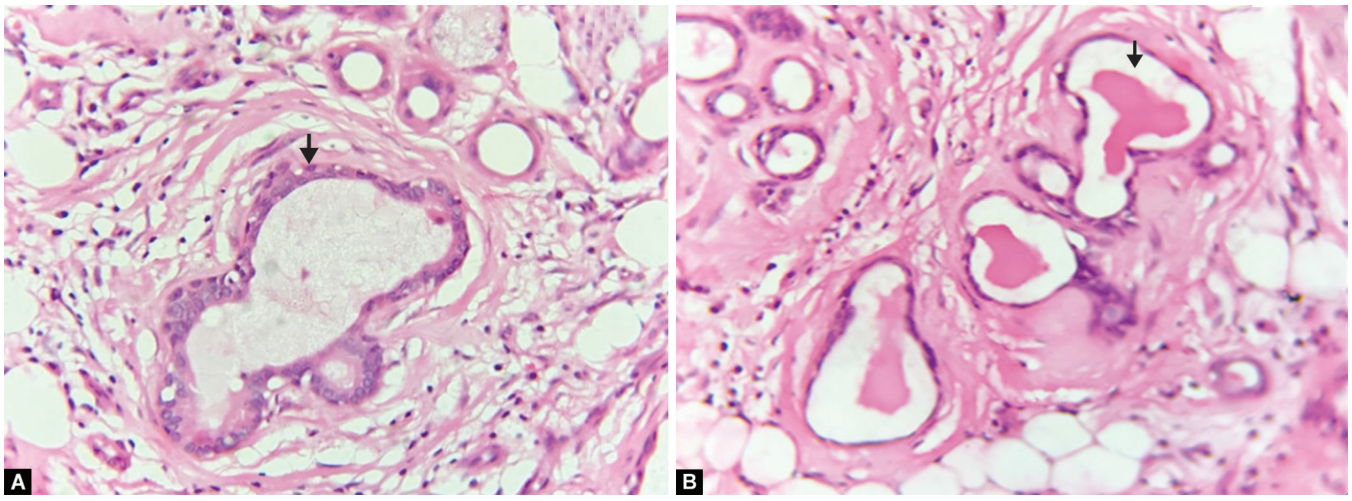


Fig. 1: A well-circumscribed partially encapsulated tissue shows numerous acinar and ductal proliferations and the ductal structures are variable sized, showing cystic changes surrounded by a sclerotic/hyalinized stroma (H and E stain-10×)

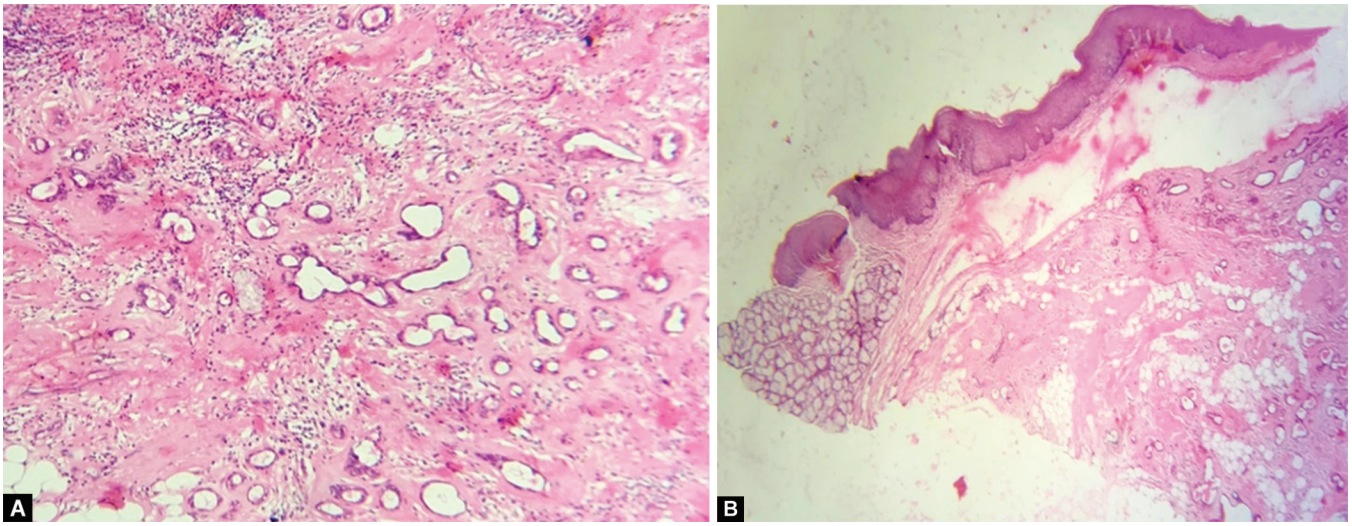
eosionophilic granules also showed positivity for PAS staining. For confirmation, immunohistochemistry was performed in the presence of adequate controls, where lining cells of the ductal and tubulo-acinar elements showed positivity for cytokeratin (AE1/AE3) with the preservation of the normal lobular architecture (Fig. 4). Both the ductal cells and the spindled myoepithelial cells, which surrounds the ductal and acinar structures, showed positivity for the S-100 protein (Fig. 5). Myoepithelial cells present at the periphery of the acini and duct showed positivity for α-SMA (Fig. 6).The proliferative index (Ki-67) was very low (<2%) in the acinar and ductal components.

Discussion

Sclerosing polycystic adenosis was first described by Smith et al. in 1996. Since then, till date not more than 60 cases of SPA have been reported in the available literature. The exact nature of this lesion is unknown, but this closely resembles benign cystic fibroadenosis tumors of the breast with features such as distinctive pseudo-neoplastic, reactive, and inflammatory process.



Figs. 2A and B: (A) Dilated ducts lined by flattened epithelial cells with occasional foamy and clear cells; (B) Eosinophilic coagulum within the dilated ducts (B) (H and E stain-40×)



Figs 3A and B: Stroma shows a focal sparse collection of chronic inflammatory cells (A) and residual minor salivary glands at the periphery of the lesion (B). ((A) H and E stain 40×; (B) H and E stain-10×)

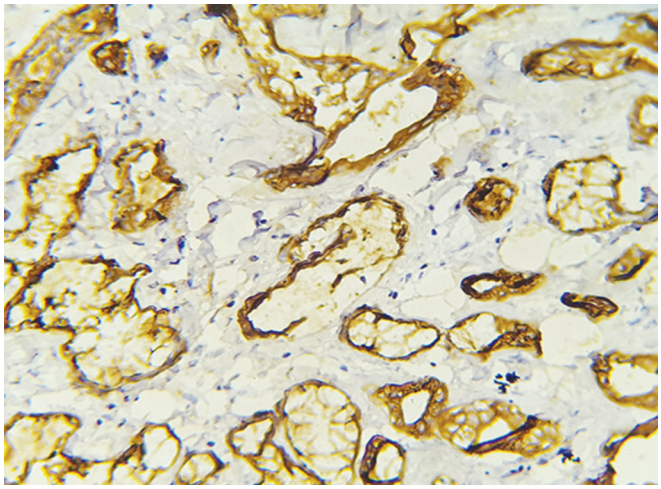


Fig. 4: Ductal and tubulo-acinar lining cells showed positivity for cytokeratin (AE1/AE3) with the preservation of the normal lobular architecture (IHC-40×)

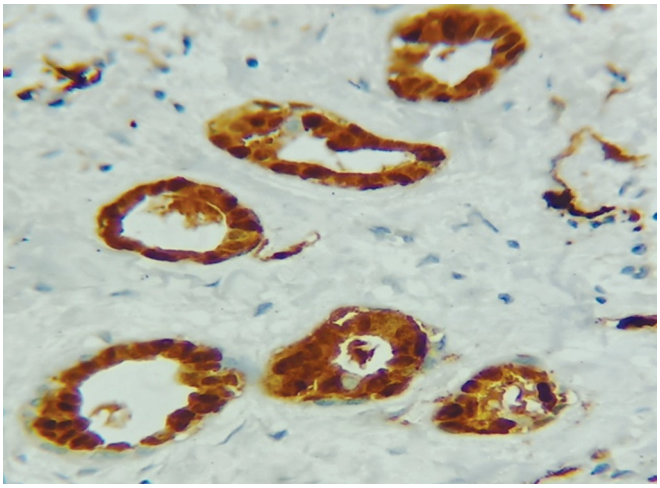


Fig. 5: Both the ductal and myoepithelial cells showed positivity for the S-100 protein (IHC-40×)

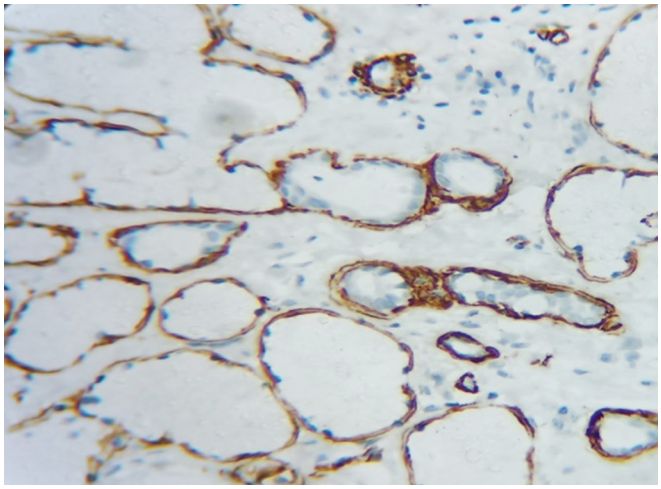


Fig. 6: Myoepithelial cells around the acini and ducts showed positivity for the α-SMA (IHC-40×)

On the basis of the available clinical features of the reported cases, the lesion is most commonly seen in the parotid gland (80%) followed by a rare involvement of the submandibular gland and more recently, involvement of the minor salivary glands of the nasal septum, muco-buccal fold, hard palate, floor of mouth, buccal mucosa, retromolar pad, ventral surface of the tongue, and lacrimal gland. The age distribution is wide (ranging from 9 years to 84 years) and most of the patients are in the fourth and fifth decades of life. The lesion is equally seen in both males and females with a slight female predilection noted in lesions occurring in the minor salivary gland.[1,3,4] In the reported cases, the size ranges from 0.3 to 7 cm. The lesion always occurs as an uni-focal lesion, whereas multi-focal lesions have also been reported in some of the studies. In the present case, the lesion was seen in a 60-year-old male at the junction of the hard and the soft palate.

Clinically, the lesion presents as a slow-growing asymptomatic, firm, smooth, and sub-mucosal nodule of varying size in the minor salivary glands.[3–5] The present case shares similar features.

Macroscopically, the lesion appears as well-circumscribed, cream/white/yellow in color, with smooth surface, measuring approximately of 1–2 mm in diameter with a firm and rubbery consistency. Our case also shows similar findings.

The lesion exhibits a great variation in the microscopic feature. The lesion is well circumscribed and shows a partial pseudo-encapsulation. The lesional tissue shows a proliferation of microcysts, ducts, and acinar structures in a densely sclerotic stroma with the preservation of normal and acinar structures. Most of the ducts are dilated and lined by flat/cuboidal epithelial cells with some cells exhibiting foamy, vacuolated, apocrine, mucous, clear, squamous, columnar, and oncocytic metaplasia. The hallmark of the tumor is the presence of large acinar cells with numerous coarse eosinophilic cytoplasmic granules that are PAS positive with or without digestion.[1,3–5] Varying degrees of ductal atypia/dysplasia and features of a ductal carcinoma *in situ* (DCIS) and an invasive carcinoma arising from SPA in which the infiltrative foci mimicking an intralesional invasive adenocarcinoma have also been reported in some cases.[6–8] However in the present case, only acinar, ductal, and microcystic proliferations, and foamy or clear cell metaplasia were noted without any dysplasia.

Immunohistochemically, the ductal cells are highly positive for cytokeratin (AE1/AE3, CAM5.2), epithelial membrane antigen (EMA), and anti-microbial antibody. Both ductal and myoepithelial cells are positivie for the S-100 protein. The acinar cells are strongly positive for GCDFP-15. Calponin, p63, alpha smooth muscle actin (α-SMA), and glial fibrillary acidic protein (GFAP) confirm the presence of myoepithelial cells surrounding the acinar and ductal structures.[1,3–5] The present case shows a strong expression of cytokeratin (AE1/AE2) in ductal cells, S-100 protein in ductal as well as peripheral myoepithelial cells and α-SMA in the myoepithelial cells with a very low expression of Ki67 <2%.

Though SPA is a benign and reactive inflammatory lesion of the salivary gland, this should be differentiated from other reactive, benign and malignant salivary gland lesions. Following lesions are considered as the differential diagnosis for the SPA.

- Chronic sclerosing sialadenitis/Kuttner's tumor: it is a rare and reactive lesion of submandibular gland histopathologically similar to the SPA. But the occurrence of salivary parenchymal atrophy and heavy lympho-plasmacytic infiltrates can be used to differentiate this lesion from the SPA.[1]

- Polycystic dysgenetic disease: it is an uncommon developmental condition mostly seen in the parotid gland with female predilection and usually occurs as a bilateral lesion. In that, the parenchyma is replaced by a honeycombed/lattice-like cysts of varying sizes and shapes with apocrine metaplastic changes. But the absence of an intraluminal proliferation and varying clinical presentation helps differentiate this lesion from the SPA.[1,5]
- Cystadenoma/cystadenocarcinoma: the presence of a papillary/mucinous growth pattern and an infiltrative behavior can be used to differentiate this lesion from the SPA.[1,5]
- Pleomorphic adenoma: it is the most common neoplasm of the salivary gland with the proliferation of a predominantly myoepithelial component within the background of the fibromyxoid/chondromyxoid stroma. A lack of an intraductal proliferation and metaplastic changes can be used to differentiate this lesion from the SPA.[8]
- The absence of ductal atypia and invasive growth is used to rule out malignanices such as the mucoepidermoid carcinoma, salivary duct carcinoma, and acinic cell carcinoma.[8]
- Clear cell odontogenic carcinoma is also included in the differential diagnosis, but the absence of odontogenic islands with significant atypia and infiltrative behavior are used to differentiate this lesion from the SPA[2]

For all the reported cases, a local surgical excision with or without a subtotal parotidectomy was the treatment of choice offering an excellent survival rate.[1,3–5] Recurrence was reported in one-third of the cases.[6–8] The nature of multifocality, inadequate surgical excision, and focal atypical hyperplasia/DCIS leads to high chances of recurrence. Malignant transformation (carcinoma arising from SPA) is very low and there is no evidence of metastasis reported till now. The present case was followed up for 12 months and no recurrence was reported till now.

The recently satisfied monoclonal nature of the SPA by the HUMARA assay and the reported cases of cytological atypia/DCIS concludes that the SPA is more likely to be a neoplasm than just a reactive process.

Conclusion

Sclerosing polycystic adenosis (SPA) is a rare, benign, and reactive inflammatory/neoplastic lesion of the salivary gland. Varying degrees of the ductal atypia and recurrence are also reported in some cases. Hence, this case of SPA of the hard palate is reported to differentiate this lesion from other more clinically sinister salivary gland pathologies to avoid a misdiagnosis.

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