Sclerosing Polycystic Adenosis of Hard Palate: A Rare Entity in Salivary Glands

Abstract

Sclerosing polycystic adenosis (SPA) is a rare neoplasm of salivary glands, with a striking resemblance to the benign fibrocystic disease of the breast. The most common site of occurrence is the parotid gland. However, submandibular gland, minor salivary glands, and buccal mucosa may also be affected. It shows a slight female predilection and occurs over a wide age range. The exact nature of the disease is unknown but is postulated to be a neoplasm recently. The treatment is effective by localized surgical excision, and recurrence is rare. To the best of our knowledge, there is only one case report of SPA involving palate. Here, we describe an additional case report of SPA occurring in the palatal region of a 49-year-old male patient.

Keywords: *Adenosis, neoplasm, palate, sclerosing*

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Introduction

Sclerosing polycystic adenosis (SPA) is a newly reported, extremely uncommon, yet distinctive, reactive lesion of the major and minor salivary glands that closely resembles fibroadenosis of the breast. Smith *et al*., in 1996, described nine cases of major salivary glands as SPA, which were previously unreported and were histologically distinct.[1] Recently, SPA is included in the salivary gland tumors under subsection of "other epithelial lesion" in the 4th edition of the World Health Organization Classification of Head and Neck Tumors.[2]

SPA is an unique tumorous condition of salivary glands as it is composed of acinar and ductal components with variable cytomorphological characteristics including foamy, vacuolated, apocrine, mucus, clear, balloon squamous, columnar, and oncocyte-like cells. It has a varied combination of histological features, of which few are reminiscent of histopathological changes that occur in the sclerosing adenosis of the mammary gland.[3]

In general, SPA is reported most commonly in the major salivary glands, particularly in the parotid glands, and very few cases were reported in the minor salivary glands, with only one being reported in the palatal region.[4] Here, we have discussed about clinical, histological, and immunohistochemical features of SPA which will aid in differentiating this lesion from other lesions with similar characteristics.

Case Report

A 49-year-old male patient visited the outpatient department of our college with a chief complaint of an intraoral swelling on the left palatal region for 8 years. The swelling was initially smaller in size, asymptomatic, and has gradually progressed to the present size. The patient gave a history of teeth extraction in the upper left posterior region. His medical history was noncontributory.

On extraoral examination, no abnormality was detected, and lymph nodes were not palpable. Intraoral examination revealed missing teeth in relation to 24, 25, 26, and 27 and grossly decayed tooth in relation to 28. A solitary, roughly oval-shaped swelling of size 2 cm × 3 cm was seen on the left palatal region extending anterioposteriorly from the edentulous region of 25, 26, 27, and 28 tooth region and mediolaterally, 2 cm lateral to midline till the edentulous ridge [Figure 1a]. Overlying mucosa was normal. On palpation, it was nontender, firm in consistency, nonreducible, and noncompressible.

Clinical differential diagnosis of palatal abscess, benign salivary gland tumors, and benign connective tissue tumors was considered.

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Orthopantomograph was taken, but no significant changes were seen [Figure 1b]. Based on the clinical and radiological findings, a provisional diagnosis "pleomorphic adenoma" of palate was given.

Incisional biopsy was done, and microscopic examination of the H and E stained tissue section revealed a well-circumscribed lesion comprising acinar and ductal components. These acinar cells were hyperplastic containing fine to larger eosinophilic granules with focal areas showing oncocytic clear cell metaplastic changes. Numerous cystically dilated ducts were seen which were lined by flattened to cuboidal cells with focal areas showing mucus cell changes. The lumen of these cysts contained eosinophilic secretory material [Figure 2a and b]. These acinar and ductal components were embedded in a dense, sclerotic collagenous stroma with few areas showing lipocytic component. Intense chronic inflammatory cell infiltrate with lymphoid follicles was noticed [Figure 2c and d]. In addition, advanced histochemical staining such as periodic acid–Schiff (PAS) and immunohistochemical staining was done.

Special staining with PAS showed the presence of large acinar cells with coarse eosinophilic PAS-positive cytoplasmic granules [Figure 3a]. Immunohistochemical staining was performed for S-100 and cytokeratin. Myoepithelial cells, surrounding ductal elements, demonstrated immunoreactivity for S-100 [Figure 3b]. Acinar and ductal cells showed positivity for cytokeratin [Figure 3c]. Based on all these features, a confirmed diagnosis of SPA was made. Complete surgical excision of the lesion was done.

Discussion

SPA is a rare, benign salivary gland lesion, recently described, sharing the anatomopathologic features of very frequent and well-described benign mammary lesions. SPA commonly occurs in the parotid gland (80.5%), submandibular gland (7.3%), and minor salivary glands (12.2%). Among the minor salivary glands, commonly involved sites are hard palate, floor of the mouth, and buccal mucosa.[4] It shows slight female predilection and occurs over a wide age spectrum of 9–84 years.[3] In the present case, swelling was present in the palatal region of a 49-year-old male patient involving the minor salivary glands, which is a rare site of occurrence. Previously, only one case of SPA was reported in the hard palate of 70-year-old male patient, and the present case becomes the second to be reported in the palatal region.

Pathogenesis of SPA is unknown. Initially, it was described as a reactive, postinflammatory process, but recent study has addressed a possible association with Epstein–Barr virus (EBV). Correlation of expression of EBV and Bcl-2 at the molecular level is demonstrated by direct upregulation of the latent membrane protein 1 or indirectly through suppression of Bax. EBV-encoded small RNAs upregulate translation of the Bcl-2 gene possibly through inactivation of protein kinase R. More specifically, EBV, as gamma herpes virus, contains a single viral Bcl-2 homolog which encodes a protein that functions in preventing apoptotic death of virus-infected cells.[5] This etiopathogenic pathway favors the neoplastic nature of the disease.

Typically, SPA of the major salivary glands are slow-growing, deep-seated, round, palpable masses, with or without associated pain and tenderness. SPA of the minor salivary glands is seen as asymptomatic, firm, smooth, freely movable submucosal nodules that are white, cream, or yellow in color. The mean tumor size ranges from 0.3 to 6 cm. SPA is often seen as a solitary lesion, but rare cases with multifocality have been documented.[3] In the present case, it is seen as a solitary lesion of size 2 cm × 3 cm involving the minor salivary glands.

Histologically, the lesion is typically well circumscribed, yet unencapsulated and consists of a proliferation of microcysts, ducts, and acinar structures in a densely sclerotic stroma. Irregularly defined lobules of abundant hyalinized collagen surrounded by variably sized cystically dilated ducts. Occasionally, closely packed small ductular elements and strangulated tubules reminiscent of sclerosing adenosis of the breast may be seen. The ducts are lined by flattened

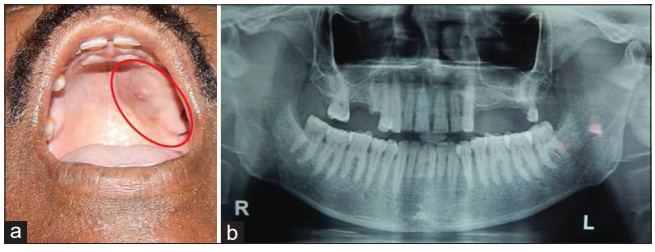

Figure 1: (a) Solitary lesion in the palate, (b) no significant changes seen in the orthopantomograph

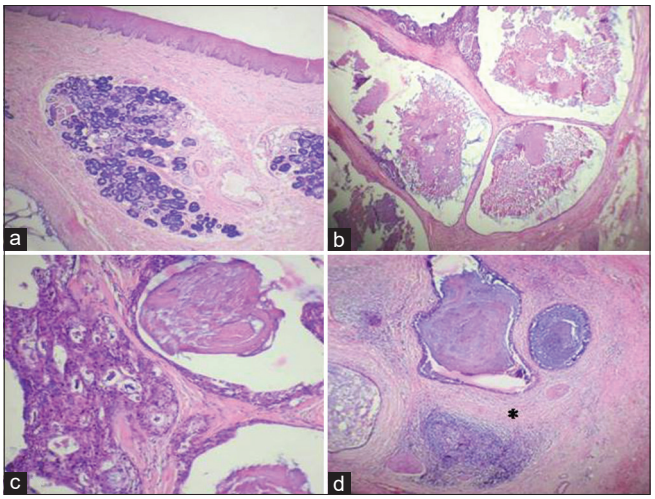

Figure 2: (a and b) Numerous cystically dilated ducts with focal areas showing mucus cell changes. The lumen of the cysts containing eosinophilic secretory material. (c) Dilated duct lined by flattened to cuboidal cells. (d) Acinar and ductal components embedded in a dense, sclerotic collagenous stroma with few areas showing lymphoid follicles* (H and E × 4)

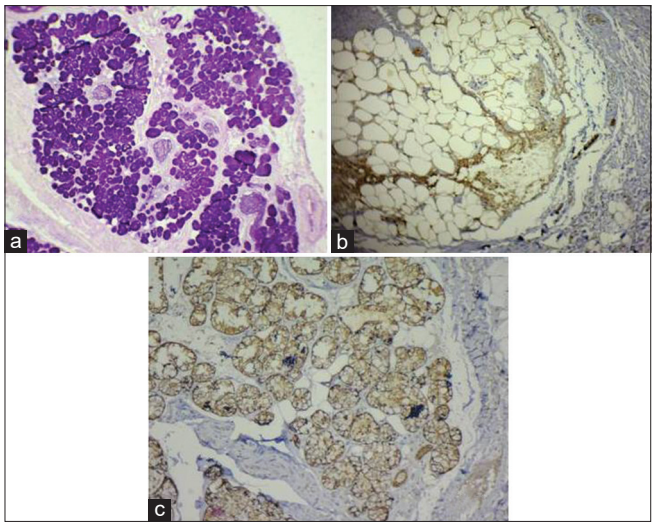

Figure 3: (a) periodic acid–Schiff staining showing the presence of large acinar cells with periodic acid–Schiff-positive granules (×4); (b) Immunoreactivity with S-100 confirmed the presence of myoepithelial cells, surrounding ductal elements (×4); (c) Immunoreactivity with cytokeratin showing positivity for acinar and ductal cells (×10)

to cuboidal epithelial cells, with some cells exhibiting apocrine-like metaplasia, manifested by well-defined apical snouting into the ductal lamina.[6] In few cases, mucous, sebaceous, squamous, foamy, vacuolated, and ballooned cells as well as brightly stained eosinophilic zymogen granules within some tubuloacinar structures are noted.[7]

In the current case, numerous cystically dilated ducts are seen, which are lined by flattened to cuboidal cells with focal areas showing mucus cell changes. The acinar and the ductal components are embedded in a dense, sclerotic collagenous stroma with few areas showing lipocytic component.

Histochemically, the hallmark of this tumor is the presence of large acinar cells with numerous coarse eosinophilic PAS-positive cytoplasmic granules, which is positive in the present case. Immunophenotypically, the lesion is biphasic in nature, demonstrating both epithelial and myoepithelial cells. The luminal cells show positive expression for AE1/AE3, CAM5.2, EMA, antimicrobial antibody, BRST-2,S-100 and the abluminal cells show positive expression for smooth muscle actin, S100 protein, p63. In the present case, immunopositivity was seen for S-100 and cytokeratin confirming the presence of myoepithelial cells and acinar elements of the lesion.[7,8]

Differential diagnosis of SPA includes pleomorphic adenoma, chronic sclerosing sialadenitis, and mucoepidermoid carcinoma. Lack of intraductal proliferation, apocrine, and sebaceous elements differentiates pleomorphic adenoma from SPA. The chronic sclerosing sialadenitis delineates from SPA by the presence of remarkable lymphoplasmacytic infiltration and the absence of ductal changes of hyperplasia. The malignancy can be differentiated based on their cytomorphologic features such as the presence of mucus-secreting cells, epidermoid type, and intermediate cells with the absence of ductal changes.[9]

Most cases of SPA are treated with localized surgical excision with clear margins. Prognosis of the lesion is favorable, and recurrence is rarely encountered.[3]

Conclusion

SPA is an uncommon, recently reported benign tumor of the salivary glands with not more than sixty cases reported till date. A very few cases are reported in the minor salivary glands. Till date, only one case of SPA has been reported in the palatal region with the present case being an addition to it. Therefore, clinicians and pathologists should be familiar with clinical, histological, and immunohistochemical features of SPA to avoid the misdiagnosis.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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