Sclerosing Polycystic Adenosis: A Rare Tumor of the Salivary Glands

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ABSTRACT

Case Presentation: A 74-year-old woman presented to the Head and Neck Surgery clinic with a 4-year history of a slowly growing, painful, left-sided neck mass in the tail of the parotid gland. Fine-needle aspiration suggested well-differentiated adenocarcinoma.

Discussion and Results: The patient underwent a superficial parotidectomy and super-selective neck dissection (level 2). Pathology revealed a tumor consistent with sclerosing polycystic adenosis.

Conclusion: Sclerosing polycystic adenosis is a rare inflammatory process that causes fibrocystic changes in the salivary gland. Apocrine-like metaplasia and epithelial atypia are common pathologic features. To our knowledge, a total of 51 cases have been described in the English-language literature.

INTRODUCTION

Sclerosing polycystic adenosis (SPA) is a rare, reactive, inflammatory lesion of the salivary glands resulting in fibrocystic changes and adenosis, similar to what occurs in the mammary glands.[1] Lesions present as slow-growing masses in salivary gland parenchyma. They are discrete, pale, and rubbery nodules. The tumors are not encapsulated but are well defined. Pathologically, they display dense sclerotic lobules and cystic change with hyalinized collagen separation. Apocrine-like metaplasia; epithelial atypia; and ductal, acinar hyperplasia are commonly observed. A distinguishing feature of this lesion is focal cystic spaces within the fibrotic stoma. Most observed cases occur in the parotid gland.[1-4] We report a typical case of SPA occurring in the parotid gland.

CASE PRESENTATION

A 74-year-old woman presented to the Head and Neck Surgery clinic at the Kaiser Permanente Medical Center in Oakland, CA, with a 4-year history of a growing, painful, left-sided neck mass. Two years before presentation, the patient underwent fine-needle aspiration with negative results for malignancy and did not pursue further workup. The mass persisted and continued to enlarge. The patient now had a firm 3.5-cm mass in the tail of the left parotid gland without overlying erythema. Fine-needle aspiration suggested a well-differentiated adenocarcinoma. Magnetic resonance imaging showed a well-defined, peripherally enhancing 3.5-cm lobe mass (Figures 1 and 2). Surgery was scheduled, and a superficial parotidectomy and a selective neck dissection (level 2) were performed. Final pathology revealed a 3.5-cm, well-circumscribed tumor consistent with SPA (Figures 3-5).

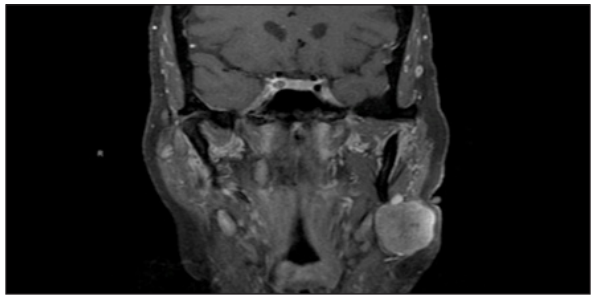

Figure 1. Magnetic resonance image, coronal cut. Coronal T1 image of tumor shows a left parotid mass.

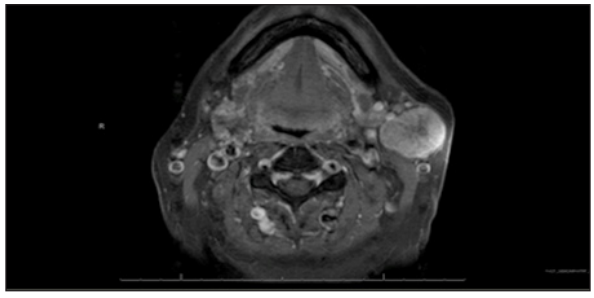

Figure 2. Magnetic resonance image, axial cut. Axial T1 image of tumor shows a well-defined, peripherally enhancing, 3.5-cm lobe mass.

DISCUSSION AND RESULTS

SPA is a rare, benign tumor of the salivary glands, which was first described in 1996.[5] About 80% of SPA cases present in the major salivary glands—specifically, the parotid gland.[1] However, cases have been observed in the minor salivary glands of the nasal septum,[2] buccal mucosa,[3] hard palate, floor of the mouth, and retromolar pad.[4] SPA has also been reported in the lacrimal gland.[3] SPA is equally common in men and women, and reported cases have a wide age of distribution.[1]

Usually, parotid SPA comprises deep-seated, slow-growing, round, palpable masses. Pain and tenderness may be present. The masses are multinodular, with cysts 1 mm to 2 mm in diameter. SPA may be multifocal.[1-8]

Histologically, SPA is characterized by acinar cells with robust eosinophilic structures similar to zymogen granules. Ductal epithelial atypia is common, and epithelial cells exhibit various cells of apocrine, foamy, vacuolated, and mucinous nature.[1-4] The lobular architecture usually includes atypical nests of myoepithelial cells.[1-4] However, infiltrative carcinoma growth does not occur.[6]

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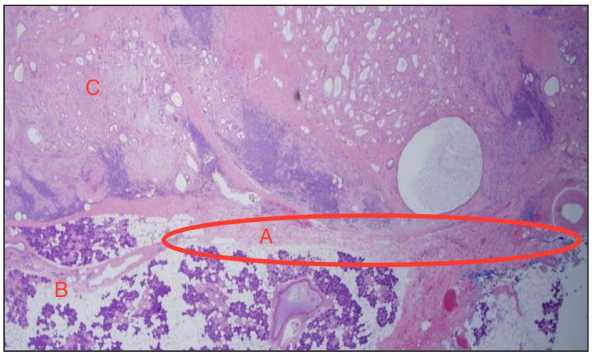

Figure 3. Histology sample of SPA at low power (200x magnification). Histology revealed a smooth border between normal salivary gland tissue and SPA. A section of smooth border is circled at A. The tissue surrounding B is normal salivary gland tissue. The tissue surrounding C is SPA.

SPA = sclerosing polycystic adenosis.

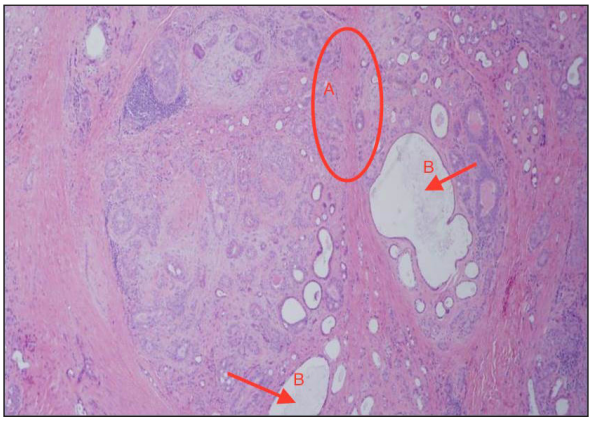

Figure 4. Histology sample of SPA at high power (400 x magnification). Histology shows lobular architecture of SPA and cysts and lobules of various sizes. A small section of architecture is circled at A; similar patterns in the figure are also examples of the lobular architecture. Cysts are denoted by arrows at B.

SPA = sclerosing polycystic adenosis.

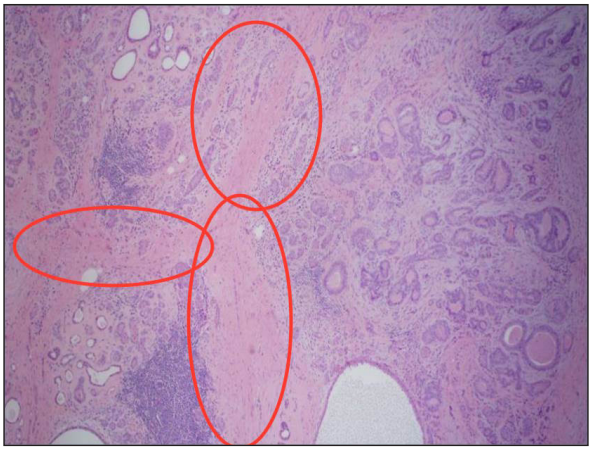

Figure 5. Histology sample of SPA at high power (400 x magnification). Collagen separation is present and circled. Sclerosing is visible, evidenced by collagen deposition.

SPA = sclerosing polycystic adenosis.

SPA is considered benign. However, one case of ductal carcinoma in situ has been reported.[6] Most cases of SPA are treated with localized surgical resection with clear margins.[8] Recurrence has been reported in up to one-third of cases. Recurrence generally occurs because of inadequate surgical resection and because of the multifocality of the SPA.[8] We found no cases of death or of metastasis attributed to SPA in a MEDLINE literature search.

There is a high chance of misdiagnosis because of the rarity of the disease and because clinicians and pathologists may be unfamiliar with it; for example, the present case was initially diagnosed as a well-differentiated adenocarcinoma.[2] Differential diagnosis included pleomorphic adenoma; benign polycystic disease; sclerosing sialadenitis; and malignant glandular neoplasias, such as mucoepidermoid carcinoma, acinic cell carcinoma, adenocarcinoma NOS (not otherwise specified), and salivary duct carcinoma.[2]

Our case was a 3.5-cm mass in the tail of the parotid gland. Presentation in the parotid gland and histologic findings of cysts and lobular architecture with collagen separation are characteristic of SPA.

CONCLUSION

SPA is a rare inflammatory process that causes fibrocystic changes in the salivary gland. Apocrine-like metaplasia and epithelial atypia are common features. To our knowledge, only 51 cases have been described in the English literature.[9] ❖

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

Acknowledgment

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