Sclerosing polycystic adenosis (SPA) is a rare salivary gland lesion that was first reported in 1966. It is characterized by a benign, sporadic, and rarely multifocal lesion that mostly arises from a parotid gland. Surgical tumor excision followed by immunohistochemical confirmation is essential to establish a definitive diagnosis. However, the standard surgical procedure for the treatment of SPA is not well established. To make plastic surgeons familiar with this disease, we describe a case of SPA and discuss the diagnostic and treatment protocols that were employed.

**CASE REPORT**

A 33-year-old man presented with a 3-year history of painless swelling of the left parotid gland. He had undergone surgery with an unknown procedure on the same site at 10 years of age. On physical examination, he had a palpable, painless, and elastic hard mass in a left preauricular area without facial nerve paralysis. Computed tomography scan revealed an encapsulated 3×2 cm lesion. Intraoperative findings showed that the tumor was embedded deep in the parotid gland. Marginal tumor excision was performed to preserve the facial nerve. Histopathological and immunohistochemical findings led to the final diagnosis of SPA. The surgery was not associated with any other complications. To date, 28 months after surgery, recurrence has not been observed. The treatment protocol of SPA has not yet been established. To make plastic surgeons familiar with this disease, we describe this case, which was successfully treated without any complications. (Plast Reconstr Surg Glob Open 2016;4:e645; doi: 10.1097/GOX.0000000000000614; Published online 17 March 2016.)
Histopathology revealed variably sized dilated cystic ducts in a background of dense fibrotic stroma. In other areas, there was proliferation of atypical ductal and acinar cells that formed a cribriform structure with no invasion (Fig. 3). Immunohistochemical analyses showed that the epithelial cells of acinar and ductal components were positive for broad spectrum cytokeratins (AE1/3) and epithelial membrane antigen. Staining for CD10, S100, and α-smooth muscle actin revealed a myoepithelial layer in the atypical ductal structure. Ki-67 was expressed in 10–15% of the epithelial cells. These immunohistochemical and histopathological findings led to a final diagnosis of SPA. It remains unclear whether this lesion was a recurrent or primary case. At present, 28 months after surgery, the lesion has not recurred.

DISCUSSION

SPA is very rare; since Smith et al1 reported the first 9 cases in 1966, approximately 66 cases have been reported.2–4 As a result, it remains challenging for surgeons and pathologists to correctly diagnose and determine the subsequent management of the lesion. The etiology of SPA also remains unknown.2,5

SPA lesions grow slowly and have been reported in patients ranging from 9 to 84 years of age.2,6 There is a slight predominance in females. The tumor is firm, rubbery, and well circumscribed. MRI reveals it to be a mass that has small cystic areas of relatively high intensity on T2-weighted images.7 However, imaging is not sufficient for diagnosing SPA; other tests are required. One may be a fine-needle aspiration, but it can be challenging to diagnose SPA because of their noncommittal features.5 Generally, the lesion must be resected and subjected to histological and immunohistochemical analysis to be diagnosed correctly.

The key histological features of SPA include lobular proliferation of ductal and acinar cells accompanied by cystically dilated ducts with apocrine and sebaceous metaplasia.2,5 The immunohistochemical analyses of SPA lesions are positive for cytokeratins (AE1/3), epithelial membrane antigen, S100 proteins, Gross Cystic Disease Fluid Protein
15 (GCDFP-15), and α-smooth muscle cell actin. Ki-67 labeling index is always low (1–2%). Although Ki-67 was expressed at relatively high levels, the other staining patterns were as described above.

Not only the diagnosis but also the treatment of SPA is based on surgical excision.8 However, a standard surgical procedure has not yet been established. Local recurrence has been estimated to occur in about 29% of cases.9 Table 1 shows the surgical approaches that were taken for primary SPA in the parotid gland in previous reports. Table 1 shows that the total recurrence rate was 18.2% (6/33 cases). The surgery-dependent recurrence rates were 44.4% (4/9 cases), 7.1% (1/14 cases), and 10% (1/10 cases) in patients treated by excision/enucleation, superficial parotidectomy, and total parotidectomy, respectively. Although the number of recurrent cases was insufficient for statistical analysis, the results suggested that removal of lesions with adequate margins of healthy tissue by superficial or total parotidectomy can result in a lower recurrence rate than that by excision or enucleation. In addition, tumors tend to recur after a period of >5 years in most reported cases.

Mackle et al17 recommended that primary and recurrent cases should be treated with superficial and total parotidectomy, respectively. However, total parotidectomy associates with complications such as facial nerve dysfunction, cosmetic deformities, and Frey syndrome.18 Additional postoperative radiation therapy and lymph node dissection are potential treatment options, especially for recurrent cases.

In the present case, because superficial parotidectomy had apparently already been performed during childhood, our choices were marginal excision or total parotidectomy. We decided that the choice of surgical procedure should depend mainly on the functional prognosis rather than on the risk of recurrence. Surgeons should consider the risks of complications and recurrence that are associated with each surgical approach and explain these to the patient so that the patient can consent in an informed manner to one of the approaches. A recent report showing that carcinoma can recur again for the third time at an SPA lesion in the parotid gland 33 years after initial diagnosis also suggests that careful long follow-up is needed.9

Table 1. Surgical Treatments for Sclerosing Polycystic Adenosis in Previous Reports

| Year | Author | Age | Sex | Preoperative Diagnosis | Primary Surgery | Follow-up Time | Recurrence |
|------|--------|-----|-----|------------------------|-----------------|----------------|------------|
| 1996 | Smith et al.1 | 29 | M | PA | Excision | Lost | Unknown |
| 1996 | Smith et al.1 | 39 | M | Adenoma | Excision | Lost | Unknown |
| 1996 | Smith et al.1 | 32 | F | PA | SP | Lost | Unknown |
| 1996 | Smith et al.1 | 63 | M | Mixed tumor | SP | 2 y 3 mo | No |
| 1996 | Smith et al.1 | 16 | F | Benign salivary tumor | SP | 1 y 5 mo | No |
| 1996 | Smith et al.1 | 17 | M | Neoplastic lesion | SP | 7 mo | No |
| 1996 | Smith et al.1 | 16 | F | PA | SP | 6 y | Yes |
| 1996 | Smith et al.1 | 31 | F | Benign tumor | Excision | 16 mo | No |
| 1996 | Smith et al.1 | 12 | F | Benign tumor | Excision | 9 y | Yes |
| 1996 | Smith et al.1 | 31 | F | Malignant tumor | SP | 3 y | No |
| 2002 | Skálová et al.10 | 20 | M | Reactive inflammation | Parotidectomy | 27 mo | No |
| 2003 | Imamura et al.11 | 9 | F | Sialadenitis | Enucleation | 10 mo | Yes |
| 2004 | Mackle et al.17 | 45 | F | PA | SP | 3 y | No |
| 2004 | Mackle et al.17 | 46 | F | Unknown | SP | 5 y 2 mo | No |
| 2004 | Mackle et al.17 | 44 | F | PA | SP | 8 y | No |
| 2004 | Mackle et al.17 | 42 | F | Mixed tumor | Excision | 4 y 1 mo | No |
| 2004 | Mackle et al.17 | 32 | M | Unknown | Parotidectomy | 14 y | No |
| 2004 | Mackle et al.17 | 43 | F | Unknown | Excision | 5 y | No |
| 2004 | Mackle et al.17 | 51 | F | Neoplasm | Parotidectomy | 6 y 7 mo | No |
| 2004 | Mackle et al.17 | 47 | F | Unknown | Parotidectomy | 2 y 6 mo | No |
| 2004 | Mackle et al.17 | 49 | M | Unknown | Parotidectomy | 40 y | No |
| 2004 | Mackle et al.17 | 9 | F | Unknown | SP | 2 y 7 mo | No |
| 2004 | Mackle et al.17 | 35 | M | Unknown | Enucleation | 15 y | No |
| 2004 | Mackle et al.17 | 67 | F | Low-grade carcinoma | Parotidectomy | 2 y 10 mo | No |
| 2004 | Mackle et al.17 | 55 | M | Unknown | Parotidectomy | Unknown | No |
| 2009 | Fulciniti et al.13 | 57 | M | Sebaceous adenoma | SP | 5 mo | No |
| 2009 | Gupta et al.14 | 52 | M | Unknown | SP | 1 y | No |
| 2010 | Petriotti et al.15 | 68 | M | PA | Subtotal parotidectomy | 1 y | No |
| 2011 | Petersson et al.16 | 45 | F | Acinic cell carcinoma | SP | Unknown | No |
| 2014 | Manojlović et al.4 | 9 | F | PA | Parotidectomy | 10 y | Yes |
| 2014 | Canas Marques and Félix9 | 33 | F | Benign tumor | SP | 4 y | No |
| 2014 | Canas Marques and Félix9 | 23 | M | Unknown | Excision | 13 y | Yes |

PA, pleomorphic adenoma; SP, superficial parotidectomy.
CONCLUSIONS

We experienced a case of SPA, which is a very rare condition. The tumor was successfully removed from the deep parotid gland without complications. Twenty-eight months after surgery, recurrence has not been observed. Histological and immunohistochemical analyses of the surgically resected specimen confirmed the diagnosis. We hope this case will alert surgeons on the possibility of SPA when a tumor is detected in the salivary gland. To choose a surgical procedure, we also recommend that surgeons should consider the risks of complications and recurrence that associate with each surgical approach.

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