

# Surgical Treatment of Rare Sclerosing Polycystic Adenosis of the Deep Parotid Gland

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**Summary:** We experienced a rare case of sclerosing polycystic adenosis (SPA) arising in a parotid gland. A 33-year-old man who underwent unspecified surgery for a lesion in the left parotid gland 23 years ago presented with a lesion on the same site. 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.)

**S**clerosing polycystic adenosis (SPA) is a rare salivary gland lesion that was first reported in 1966.<sup>1</sup> It is characterized by a benign, sporadic, and rarely multifocal lesion that mostly arises from a parotid gland.<sup>1</sup> 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.

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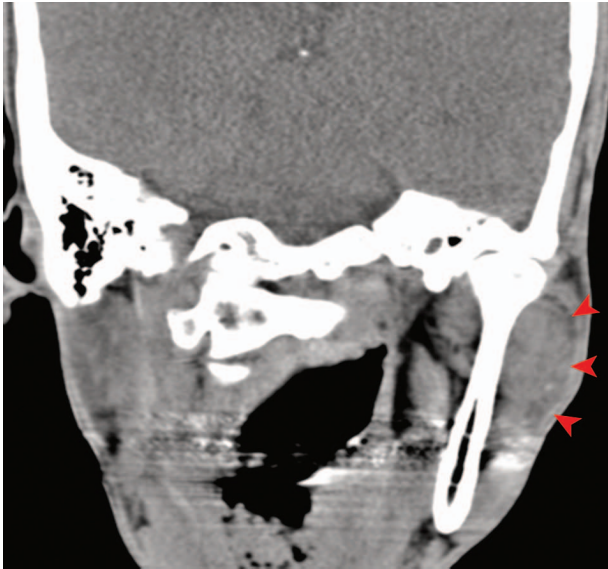
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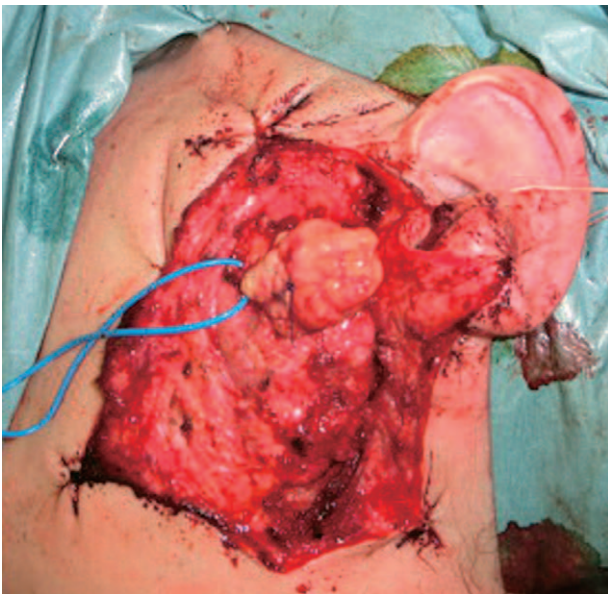
## CASE REPORT

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 showed an encapsulated heterogeneous mass in the left parotid gland (Fig. 1). The clinical and imaging findings suggested that he might have recurrence of pleomorphic adenoma. We planned a superficial parotidectomy to prevent recurrence and facial nerve injury. However, intraoperative findings using a nerve stimulator (TS-260, Keisei Medical Industrial Co, Ltd, Tokyo, Japan) showed that the mass was located just below the buccal branches of the facial nerve and was embedded deep in the parotid gland. Moreover, surrounding scarred tissues from the previous surgery made it difficult to identify the main trunk of the facial nerve. We identified buccal branches from the periphery that traced up to the main trunk. To preserve the facial nerve, we dissected it and resected the tumor by marginal excision. The tumor was well circumscribed (Fig. 2).

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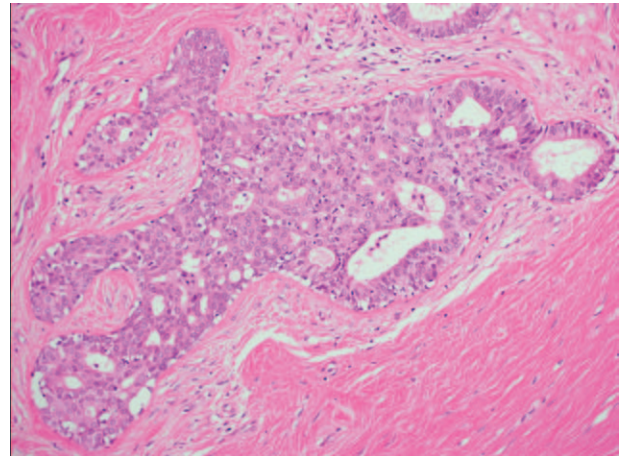


**Fig. 1.** Preoperative plain computed tomography image. Arrow heads indicate the presence of an encapsulated heterogeneous mass in the left parotid gland.



**Fig. 2.** Intraoperative photograph. The excised lesion was encapsulated, elastic, firm, and light yellow. It was 3×2 cm in size and had a nodular appearance. It was surrounded by scar tissue. The blue vessel tape on the left wrapped the buccal branch.

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 anti-



**Fig. 3.** Intraductal proliferation of atypical epithelial cells in the background of dense sclerotic stroma (hematoxylin and eosin, 200×).

gen. Staining for CD10, S100, and  $\alpha$ -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 al<sup>1</sup> reported the first 9 cases in 1966, approximately 66 cases have been reported.<sup>2–4</sup> 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.<sup>2,5</sup>

SPA lesions grow slowly and have been reported in patients ranging from 9 to 84 years of age.<sup>2,6</sup> 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.<sup>7</sup> 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.<sup>5</sup> 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.<sup>2,5</sup> 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  $\alpha$ -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.<sup>8</sup> However, a standard surgical procedure has not yet been established. Local recurrence has been estimated to occur in about 29% of cases.<sup>9</sup> 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 al<sup>17</sup> 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.<sup>18</sup> 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.<sup>9</sup>

**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. <sup>1</sup>	26	M	PA	Excision	Lost	Unknown		
		39	M	Adenoma	Excision	Lost	Unknown		
		32	F	PA	SP	Lost	Unknown		
		63	M	Mixed tumor	SP	2 y 3 mo	No		
		16	F	Benign salivary tumor	SP	1 y 5 mo	No		
		17	M	Neoplastic lesion	SP	7 mo	No		
		16	F	PA	SP	6 y	Yes		
		31	F	Benign tumor	Excision	16 mo	No		
		12	F	Benign tumor	Excision	9 y	Yes		
		2002	Skálová et al. <sup>10</sup>	31	F	Malignant tumor	SP	3 y	No
		2003	Imamura et al. <sup>11</sup>	20	M	Reactive inflammation	Parotidectomy	27 mo	No
		2004	Mackle et al. <sup>17</sup>	9	F	Sialadenitis	Enucleation	10 mo	Yes
		2005	Bharadwaj et al. <sup>12</sup>	45	F	PA	SP	3 y	No
46	F			Unknown	SP	5 y 2 mo	No		
2006	Gnepp et al. <sup>8</sup>	44	F	PA	SP	8 y	No		
		42	M	Mixed tumor	Excision	4 y 1 mo	No		
		32	M	Unknown	Parotidectomy	14 y	No		
		43	F	Unknown	Excision	5 y	No		
		51	F	Neoplasm	Parotidectomy	6 y 7 mo	No		
		47	F	Unknown	Parotidectomy	2 y 6 mo	No		
		unknown	M	Unknown	Parotidectomy	40 y	No		
		49	M	Unknown	SP	2 y 7 mo	No		
		9	F	Unknown	Enucleation	15 y	Yes		
		35	M	Unknown	Parotidectomy	2 y 10 mo	No		
2009	Fulciniti et al. <sup>13</sup>	67	F	Low-grade carcinoma	Parotidectomy	Unknown	No		
		55	M	Unknown	Parotidectomy	4 y 6 mo	No		
2009	Gupta et al. <sup>14</sup>	57	M	Sebaceous adenoma	SP	5 mo	No		
2009	Gupta et al. <sup>14</sup>	52	M	Unknown	SP	1 y	No		
2010	Perottino et al. <sup>15</sup>	68	M	PA	Subtotal parotidectomy	1 y	No		
2011	Petersson et al. <sup>16</sup>	45	F	Acinic cell carcinoma	SP	Unknown	No		
2014	Manojlović et al. <sup>4</sup>	9	F	PA	Parotidectomy	10 y	Yes		
		33	F	Benign tumor	SP	4 y	No		
2014	Canas Marques and Félix <sup>9</sup>	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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