

CASE REPORT

Dysgenetic polycystic disease of the parotid gland: Report of a case and review of the literature

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ABSTRACT

Dysgenetic polycystic disease (DPD) is a rare cystic lesion affecting mainly the parotid salivary glands with only 14 reported cases in English literature. DPD of parotids is more common in females, common in younger age group, mostly bilateral in occurrence and commonly affects the superficial lobe. Surgery is performed for symptomatic relief and cosmosis. We report a case of a unilateral DPD of the parotid gland with distinctive histopathology. Patient was treated by performing total parotidectomy with preservation of the facial nerve.

Key words: Dysgenetic, parotid, polycystic

INTRODUCTION

Dysgenetic polycystic disease (DPD) of salivary gland is a rare cystic condition^[1] with distinctive histopathology^[2] resembling that of polycystic condition affecting the kidneys and pancreas. Parotid glands are the most commonly affected amongst the salivary glands. This disease is thought to be a developmental disorder of the distal ductal system of the salivary gland^[2,3] and is common among women.

In this paper, we present a rare case of unilateral DPD of the parotid gland.

CASE-REPORT

A 21-year-old woman came to us with a chief complaint of progressively growing, non-painful swelling near the right ear since 2 years. She complained of a sudden increase in size and associated dull, aching and continuous pain. Her medical history was insignificant. Family history revealed that her mother and elder sister were operated on the lower jaw for multiple cystic lesions, reports indicated they had odontogenic keratocysts.

Extra-oral examination revealed diffuse swelling on the right side of the face [Figure 1a] measuring about 3 cm × 5 cm

extending from tragus of the ear to 2 cm below the angle of mandible superior-inferiorly and anterior-posteriorly it extended between posterior border of mandible and mastoid process. Elevation of the right ear lobe was also evident. Swelling was tender, firm in consistency and fluctuant; skin overlying the swelling was normal and was not fixed to underlying structures. Regional lymph nodes were not palpable. Facial nerve function was intact bilaterally. Salivary flow from the Stenson's duct was clear bilaterally. Provisional clinical diagnosis of a benign cystic neoplasm was made.

Computed tomography scan [Figure 2a] revealed an enlarged right parotid gland extending into the parapharyngeal region. There were multiple small cystic areas in the gland with one large cystic area in the deep lobe. The entire gland was well-circumscribed with no evidence of infiltration into the surrounding tissue giving the impression of a benign cystic lesion. The parotid on the left side was normal.

Considering the clinical, radiological presentation and presence of multiple cystic spaces in the gland, malignancy was excluded. As the lesion occupied most of the gland, consent was taken from the patient for total parotidectomy under general anesthesia.

Total parotidectomy with preservation of the facial nerve was performed by modified Blair's incision. Facial nerve was located by proximal identification technique. The branches of facial nerve were easily separated from the tissue as the lesion was cystic and encapsulated in nature. Superficial as well as deep lobes were removed in two portions and were sent for histopathological examination and the surgical defect was reconstructed with posterior belly of digastric flap. Post-operatively facial nerve function was normal with mild

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paralysis of marginal mandibular branch, which recovered after 1 month.

Excised specimen (part superficial to the facial nerve) measured 6.0 cm × 4.2 cm × 1.8 cm and was dark brown in color, cut surface showed adipose and gland tissue. The part deeper to the facial nerve measured 6.8 cm × 5.2 cm × 6.0 cm [Figure 2b], pale tan to dark brown in color and the cut surface revealed small cystic spaces containing mucin like material. Histopathological sections from the superficial part of the gland revealed serous acini and ducts with connective tissue septae separating the parenchyma. Histopathological sections from the gland deeper to the facial nerve revealed only multiple cystic spaces of varying diameter ranging from few millimeters to few centimeters in a loose, myxomatous and bland connective tissue stroma [Figure 3]. The cystic spaces were lined by a variety of epithelial cells-cuboidal, columnar and squamous cells [Figure 4]. They contained watery to pale eosinophilic material in them [Figures 5a and 6]. Entire parenchyma was replaced by the cystic spaces and connective tissue [Figure 7]. Immunohistochemical staining for pan cytokeratin (CK), epithelial membrane antigen, progesterone and estrogen receptors were performed and only panCK was positive suggestive of epithelial origin of

the disease [Figure 5b]. Thus, a final diagnosis of DPD of the parotid gland was arrived at. The patient was followed-up for 18 months with no evidence of recurrence [Figure 1b].

DISCUSSION

Spectrum of non-neoplastic diseases of the major salivary glands include acinar/ductal malformations, cysts, sialadenitis, sialolithiasis, sialadenosis, human immunodeficiency virus-associated lympho-epithelial cysts (LC), oncocytosis and sialometaplasia.

Three benign cysts commonly affect the parotid glands [Table 1] they are LC, salivary duct cyst (SDC) and polycystic or dysgenetic cyst.^[4] Polycystic or dysgenetic disease of the

Table 1: Comparison of clinical features of various parotid salivary gland cysts

Clinical Parameter	Salivary duct cyst	Sclerosing polycystic adenosis	Dysgenetic polycystic disease
Age predilection	Older 5 th decade	12-63 years	Young adults and in children
Sex predilection	None	None	Mostly in females
Location	Mostly parotid	Mostly parotid	Mostly parotid

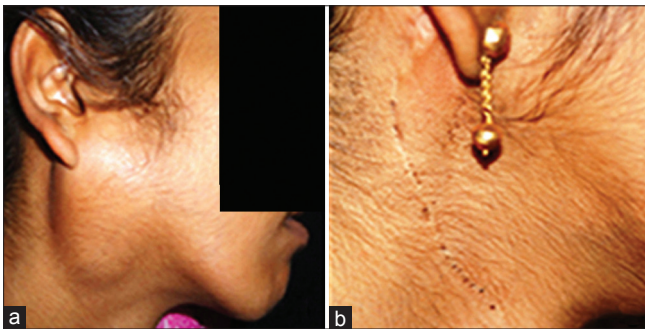


Figure 1: (a) Diffuse swelling on the right side of the face extending between the tragus and angle of mandible, (b) Patient at 18 months follow-up with no evidence of recurrence, shows minimal surgical scar

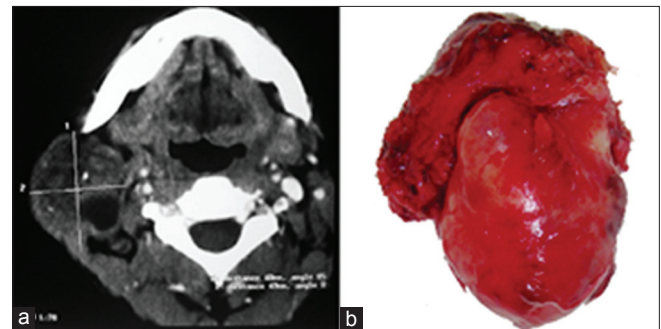


Figure 2: (a) Coronal section of computed tomography scan showing enlarged right parotid gland extending into the parapharyngeal region, (b) Excised specimen showing affected deep lobe of the parotid gland

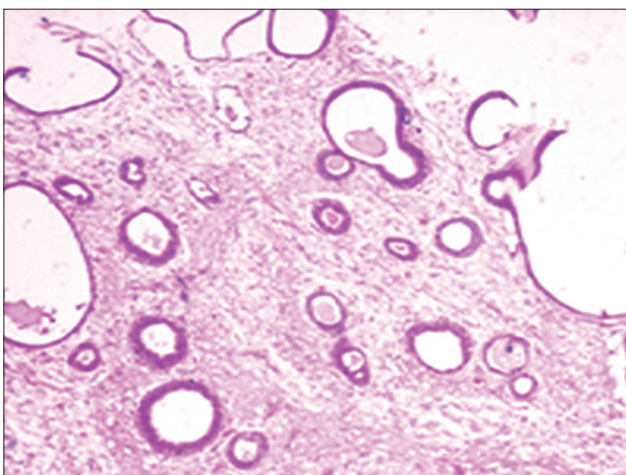


Figure 3: Photomicrograph showing multi-cystic spaces of various sizes lined by epithelium within a bland connective tissue stroma (H&E stain, ×100)

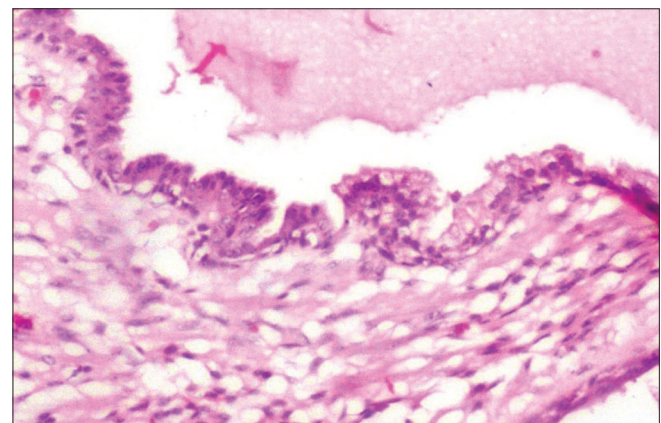


Figure 4: The epithelium resembles embryonic type of epithelium which lacks a clear basement membrane (H&E stain, x400)

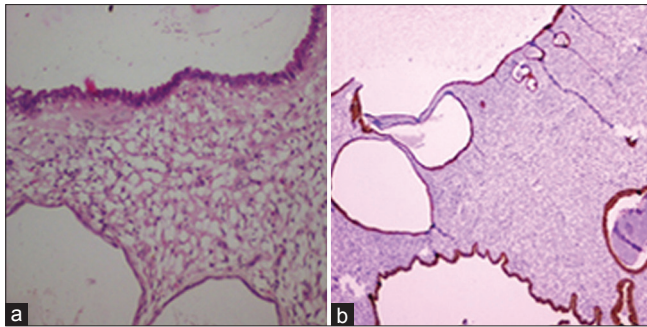


Figure 5: (a) Epithelial lining under high magnification (H&E stain, ×400) with bland connective tissue, (b) Immunohistochemical photomicrograph showing pan cytokeratin expression in the epithelial cells of the cystic spaces (IHC stain, ×100)

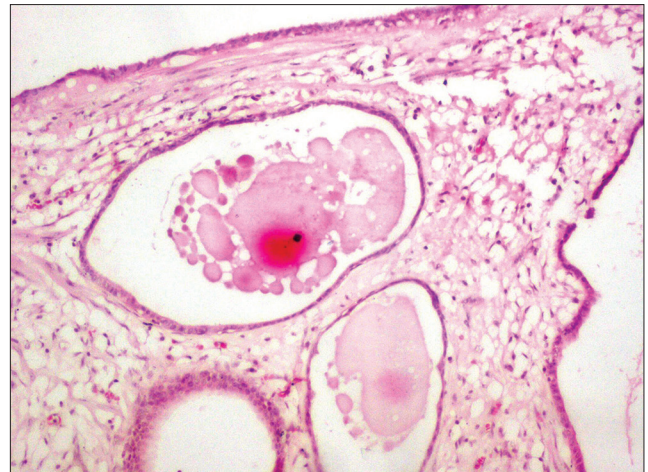


Figure 6: Section shows multiple cystic spaces lined by epithelium. The epithelium thickness varies and the cystic spaces show eosinophilic secretion within the lumen. The connective tissue is loose and edematous along with scanty inflammatory cells. Salivary gland parenchyma is absent (H&E stain, ×100)

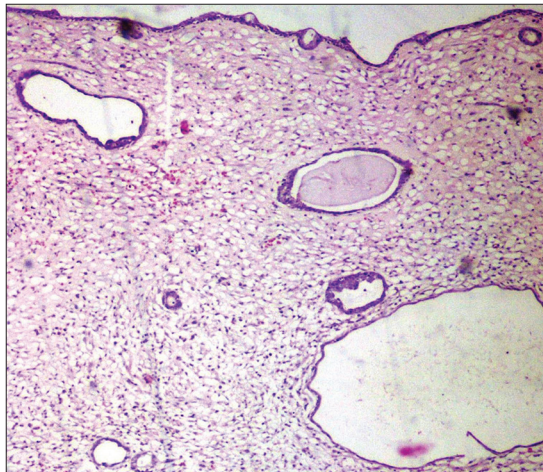


Figure 7: Section shows the entire salivary gland parenchyma replaced by cystic spaces of varying sizes and shapes and loose myxomatous connective tissue stroma which is slightly inflamed. The cystic spaces appear to be lined by single layered flat cells in some areas and hyperplastic in other. The ducts are enlarged and hyperplastic. There is light stained secretion in some cystic spaces (H&E stain, ×100)

parotid gland is the rarest of the non-neoplastic cystic lesions since only 14 cases have been reported in the literature.^[5] Among all the cases reported in the English literature this is the fourth case showing unilateral involvement.

DPD of parotid was first reported from an analysis of a large series of 5739 cases of salivary gland disorders.^[1] Because of absence of inflammation in the connective tissue, it is considered to be a developmental disorder arising from the distal duct system of the salivary glands, apparently limited to parotid gland and females with characteristic clinical presentation and distinctive; if not pathognomonic histopathological appearance.^[1-3] However, there are two reported cases of DPD affecting males^[5] and only one case affecting submandibular gland.^[6]

DPD of parotid resembles the DPD of other parenchymal organs such as pancreas and kidneys and is thought to result from an unknown common embryonic insult with an autosomal dominant pattern of inheritance in females.^[1] It presents as bilateral parotid swellings and overt clinical

signs are usually delayed even in adulthood.^[2] Pathogenesis of DPD has been related to disturbance in the ramification and canalization of the ductal system during the second stage of the development of salivary gland that extends to the end of the seventh embryonal month, hence it is thought to be a developmental malformation of the duct system.^[3]

Brown *et al.*, have reported a case where the swelling worsened with the onset of pregnancy and regressed within 4-6 months after parturition. They have speculated that hormonal changes might cause expression or exacerbation of the underlying condition.^[6]

In the present case, there was a unilateral fluctuating non-tender swelling in the right deep lobe of the parotid gland. The age and sex of the patient correlated with most of the other reported cases. In the absence of the superficial lobe or information about the location of surgery, it would have been difficult to even identify the tissue of origin as salivary gland because the entire deep lobe parenchyma was replaced by multiple cystic spaces interspersed with bland, loose myxomatous connective tissue stroma without any fibrosis. In the entire deep lobe, although the lobular pattern was appreciable the parenchyma was entirely replaced by epithelial lined cystic spaces of various sizes and configuration. Although residual acini were absent, a few ducts were seen in some places. The epithelium lining the cystic spaces ranged from flattened cells to cuboidal and columnar with no associated acute or chronic sialadenitis. Oncocytes were not found anywhere. The cystic spaces showed pale eosinophilic secretions in many places, but no microlith or spherolith formation was seen.

Because the cystic lesions in the parotid gland may represent an array of diverse entities with different biological

Table 2: Histopathological similarities among the three cystic lesions of salivary glands

Histopathological feature	Salivary duct cyst			Sclerosing polycystic adenosis	Dysgenetic polycystic disease
	MRC	ROC	MPC		
Cystic spaces in the parenchyma	Mostly unicystic but sometimes multicystic pattern also seen	Mostly unicystic but sometimes multicystic pattern also seen	Multicystic pattern	Multicystic spaces seen	Honey comb lattice work like multicystic spaces seen
Epithelium lining of the cystic spaces	Present	Present	Present	Present	Present

MRC: Mucous retention cyst, ROC: Reactive oncocytoid cyst, MPC: Mucopapillary cyst

Table 3: Histopathological differences among the three cystic lesions of salivary glands

Histopathological feature	Salivary duct cyst			Sclerosing polycystic adenosis	Dysgenetic polycystic disease
	MRC	ROC	MPC		
Acinar tissue	Present	Present	Present	Hyperplastic acinar elements distributed in the fibrous tissue present with eosinophilic PAS positive granules in the cytoplasm	Very minimal or totally absent in the affected lobule
Stroma	Normal	Normal with mild sclerosis	Normal	Hyalinized/sclerosed	Loose edematous/myxomatous
Cystic space	Uni/multicystic	Uni/multicystic	Multicystic	Multicystic	Multicystic spaces (lattice like/honeycomb pattern)
Type of epithelium	Cuboidal, columnar, non-keratinized stratified squamous with mucous metaplasia	Columnar often pseudo-stratified	Cuboidal columnar and focal areas of squamous epithelium with or without mucous metaplasia	Apocrine like metaplasia and ductal hyperplasia	Embryonal type of epithelium with lack of clear basement membrane Columnar/cuboidal or flattened and attenuated cells or even almost devoid of epithelium in some areas
Presence of oncocytes	Oncocytes absent	Oncocytes must	Absent	Absent	Absent
Inflammation	Minimal	Mild	Unknown	Of variable intensity	Absent (bland stroma)
Immunohistochemistry					
Estrogen positive receptors	Negative	Negative	Negative	Positive for both the markers	Negative
Progesterone positive receptors				EMA is positive	EMA is focally positive in our case

PAS: Periodic acid schiff, MRC: Mucous retention cyst, ROC: Reactive oncocytoid cyst, MPC: Mucopapillary cyst, EMA: Epithelial membrane antigen

behavior, a number of cystic lesions were considered in the differential diagnosis and was narrowed down to two possible lesions namely SDC and sclerosing polycystic adenosis [Tables 2 and 3]. We took multiple samples of the specimen again, so as to include all the representative areas to identify, if any mucoepidermoid or adenoid cystic carcinoma like areas, which are known to cause obstruction of the ducts resulting in SDC were present.^[7,8] Carcinomatous tissue was not seen in the sections; hence, a diagnosis of SDC associated with salivary gland carcinoma was ruled out. Furthermore, the histopathology did not resemble any type of SDC as described by Eversole such as presence of even minimal inflammation, oncocytes, mucous metaplasia and papillary projections which made SDC a less likely diagnosis.^[9] Absence of fibrosis, hyperplastic ductal and acinar epithelial elements did not favor sclerosing polycystic adenosis,^[10] which was excluded based on immunohistochemical markers as well. Thus, a

final diagnosis of DPD of the parotid gland was arrived at based on extensive involvement and replacement of salivary parenchyma with maintenance of lobular architecture and distinctive features of this disease as described in previously reported cases.^[1,2]

Since the present case resembles an autosomal dominant pattern with both mother and sister having been operated for multiple odontogenic keratocysts, the genetic predisposition of this condition cannot be ruled out as OKC's are also known to have a genetic predisposition. The same genetic abnormality, which was expressed in the mother and sister as multiple OKC's could have expressed as DPD in the present case. This reinforces the usage of the term "dysgenetic" for this condition. Both OKC and DPD are developmental cysts in nature while the former is odontogenic in origin, whereas the latter is salivary gland origin.^[9]

There are many theories, which have been put forward regarding the etiology of this condition such as genetic predisposition, retention of secretion leading to cyst formation and developmental malformation of salivary gland ductal system.^[5]

In all the previously reported cases of DPD with familial occurrences the cystic lesions were restricted to only salivary glands while in the present case instead of salivary gland genetic abnormality, the condition might have expressed as multiple OKC of the lower jaw in mother and sister.

In general, lobectomy or superficial parotidectomy is the treatment of choice as the lesion is benign in nature. However, further development of new lesions or enlargement of residual cystic tissue in the remaining lobes have not been reported.^[3] In our case, most of the gland was involved; hence total parotidectomy with the preservation of the facial nerve was performed, not only for cosmetic purpose but also to relieve symptoms.

Long-term follow-up is required to rule out recurrence and to screen for the involvement of other salivary glands since very few cases of this nature have been reported in the literature.

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