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Polycystic Parotid Disease: A Rare Case Report

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Abstract Polycystic (dysgenetic) parotid disease is a benign cystic condition of parotid gland arising from distal ductal system, commonly involving the parotid gland. It is commonly seen in females and is seen mostly involving the parotid gland bilaterally. Clinically it manifests as a slowly progressive, non tender fluctuant swelling of parotid gland with no defects in the salivary gland function. Here we report a rare case of polycystic parotid disease diagnosed in an 83 year old male patient.

Keywords: *polycystic parotid disease, parotid cysts, benign parotid cysts, parotid gland, benign parotid disease*

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1. Introduction

Benign cysts of salivary glands are rare. They are seen commonly involving the parotid gland and most of them represent cystic component of neoplasms. Polycystic (dysgenetic) parotid disease is a benign cystic condition of parotid gland and accounts for 6% of all salivary gland enlargements [1] and only 14 cases of this disease have been reported in the literature so far. It is a developmental disorder arising from distal ductal system of parotid gland. It often manifests in childhood or early adulthood but can occur later in life. It is commonly seen in females and is seen mostly involving the parotid gland bilaterally. [2] Clinically it manifests as a slowly progressive, non tender fluctuant swelling of parotid gland with no defects in the salivary gland function [3].

Polycystic parotid disease has distinct histopathological features4 resembling that of polycystic condition affecting kidneys and pancreas and it has been found that there is no clinical association between these organs [1].

Hereby, we report a rare case of polycystic parotid disease, to our knowledge, only the third case reported in a male patient that was diagnosed in an 80 year old patient, wherein the age and sex of the patient did not correlate with other reported cases.

2. Case Report

An 83 year old male patient reported to us with a chief complaint of swelling below the right ear since 5 days. There was no history of trauma or toothache preceding the swelling. The swelling was associated with mild pain on digital manipulation since few days and had progressed in size to attain the present size. There was no history of paraesthesia, dryness of mouth, dryness in the eyes, difficulty in speaking, swallowing, breathing, difficulty in jaw movements and mouth opening. There was no history of other associated symptoms such as fever, loss of appetite, fatigue, joint pain or loss of weight. Patient was a known diabetic (controlled) and hypertensive (under medication) since 15 years. There was no history of any other systemic illness. There was no history of any deleterious habits like smoking or alcoholism. Patient's family history was non-contributory.

General physical examination revealed a well built and nourished patient with vital signs within satisfactory limits. The regional lymph nodes were not palpable.

Extra oral examination revealed a solitary diffuse swelling on the right side of the face in the parotid region measuring approximately about 5 x 3 cm in size extending superiorly from front of tragus of the ear to 3 cm below on the angle of the mandible inferiorly. There was no elevation of ear lobe. The skin over the swelling appeared normal. There was no local rise in temperature and the swelling was tender on palpation. The swelling was firm in consistency, non fluctuant, non - lobulated, non - pulsatile and fixed to the underlying structure.

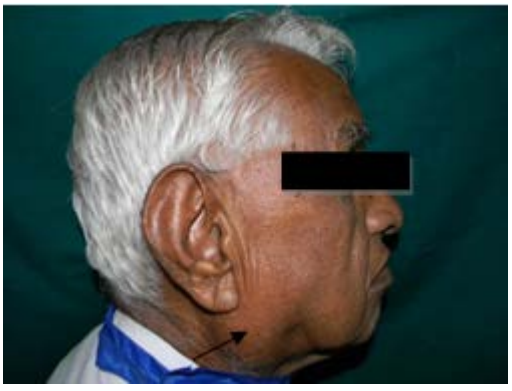

Figure 1. Swelling on the right side of parotid region

On intra-oral examination of hard and soft tissues no abnormalities were detected. Right and left Stenson's

ductal opening was patent, non-inflamed and copious salivary flow was noted.

Based on the history and clinical examination a provisional diagnosis of Warthin's tumor was made.

Clinical differential diagnosis included were bacterial sialadenitis, Pleomorphic adenoma, Sjogren's syndrome, HIV salivary gland disease, Benign lymphoepithelial cyst of parotid gland, Parotid cyst, Branchial cleft cyst, Mucoepidermoid carcinoma, Adenoid cystic carcinoma and Dermoid cyst

Keeping the above differential diagnosis in mind the patient was then subjected to radiological and routine hematological investigations. Orthopantamography showed no abnormalities. Patient was negative for HIV, Erythrocyte sedimentation rate was raised (48 mm/hr) and Random blood sugar and urine sugar were within normal limits.

Ultrasound guided Fine needle aspiration cytology (FNAC) of bilateral parotid gland was done which revealed multiple hypoehoic cystic spaces of various sizes with largest size being in the lower pole of right side of parotid gland of measuring approximately 3.3x1.8 cm in size. Two small hypoechoic nodules within the right parotid gland measuring approximately 10 mm and 9 mm in size were also noted. There was no evidence of calculi or duct dilatation or abscess. FNAC of right parotid revealed turbid white material from the mass. On microscopic examination smear studied showed sheets of neutrophils, macrophages and degenerated cells with background showing necrotic material. There was no evidence of tuberculosis or malignancy in the smears studied.

Patient was further subjected to MRI for confirmation of this diagnosis. On MRI T1 and T2 weighted images showed multiple well defined mildly thick (3.0 mm) walled cystic lesion in the right parotid inferiorly at superficial and deep lobes junction measuring 2.6x2.2 cm. Similar multiple other cystic lesions measuring 5.0 mm to 1.5 cm were noted in the contra-lateral parotid gland.

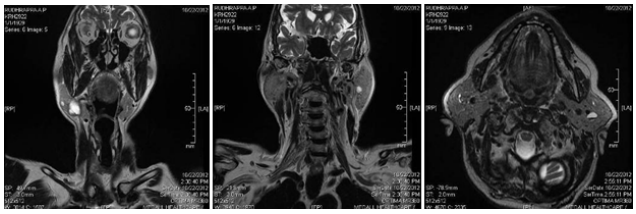

Figure 2. Post – contrast coronal and axial T1 & T2 (W) MRI images showing multiple cystic lesions with marked signal hyperintensity

There was no obvious nodularity/septation or radiating sinus/fistulous tract noted. Based on these advanced diagnostic imaging modalities a final diagnosis of Dysgenetic Polycystic parotid disease was made.

Antibiotics and analgesics were prescribed and patient recalled after one week. On follow up, swelling was no longer tender. Due to the presence of multiple cysts in the junction of superficial and deep lobe of parotid gland the patient was made cautious about the complications of surgery affecting the facial nerve and its recurrence, as spontaneous resolution has been reported. Patient refused surgery and hence symptomatic treatment was done and patient is under regular follow up.

3. Discussion

Polycystic parotid disease (PPD) is a rare benign condition of parotid gland which may have a hereditary background. Mihalyka [5] in 1962 first reported the clinical features of PPD, which was further confirmed histopathologically by Seifert and Donath [6] in 1981 and the condition was termed as bilateral dysgenetic polycystic parotid disease. [1] PPD mostly involves the parotid gland however only one case has been reported affecting the submandibular gland. [7] Siefert et al (1981) was the first who analysed this disorder among two unrelated patients out of 5,739 cases of salivary gland disease. [6] He considered it to be a developmental anomaly due to the absence of inflammation in the connective tissue and that the developmental abnormality of the intercalated duct system of the gland caused the cystic changes, particularly a disturbance of branching and canalization of the terminal salivary duct buds during the second stage of development of salivary gland that extends to the seventh embryonal month [8].

Brown et al. reported a case where the swelling had worsened at the time of pregnancy and regressed after parturition within 4-6 months. They speculated that the expression or exacerbation of the underlying condition could be due to hormonal changes [9].

Despite this disease being a rare occurrence, it has characteristic histologic appearance resembling that of a polycystic condition seen affecting the pancreas and kidneys.

Three benign cysts that commonly affect salivary glands are Salivary duct cyst, Sclerosing polycystic adenosis and Dysgenetic polycystic parotid disease of which the latter is the rarest [7].

This disease is believed to be inherited as an autosomal dominant pattern. Normally, sex-linked traits are seen in males. This disease is atypical in that the normal linkage appears reversed, in which several families affected were only females. [9] Armed Forced Institute of Pathology reported one case that occured in a male [10] and another reported by Karen A Eley et al. in an 8 year old boy. [1] Herein, we report a rare case in which an elderly male was affected. The condition is also known to be familial with one case having occurred in mother and daughter as reported by Brown et al. [9] Our patient had an insignificant family history.

Sialography is the imaging technique of choice for delineating ductal anatomy and for identifying and localizing sialolithiasis. Sialography performed during active infection would further irritate and potentially rupture the already inflamed gland. Patent salivary ductal opening with copious salivary flow was noted bilaterally. [11] Hence sialography was not performed. As such Sialography in this disease do not have a characteristic appearance.

Neoplasia with cystic changes was considered in the differential diagnosis. Warthin's tumor is a benign neoplasm most commonly associated with cystic components. Pleomorphic adenoma, Mucoepidermoid carcinoma and adenoid cystic carcinoma are also associated with a cyst formation. [6] They usually manifest superficially in front of the ear, but may be in the parotid substance or well away from the gland, and may be cystic, filled with turbid brown or mucoid fluid or part

may be solid. Cysts derived from parotid epithelium may be entrapped within the lymph node occcurring as lymphoepithelial cyst which is uncommon in parotid gland. In recent years literature shows increased parotid lymphoepithelial cysts have been reported in HIV patients. In our present case patient was negative for HIV and on FNAC no neoplastic cells were noted hence the diagnosis of neoplasia with cystic changes was ruled out.

Typical presentation of PPD is that of a non tender mass although it might be symptomatic if infected. Clinically it is characterized by recurrent, painless swelling of the involved gland(s) which are not found associated with any other abnormality. In the present case the swelling might have been present since childhood or early adulthood which was unnoticed by the patient as it was asymptomatic. Our patient gave no history of recurrence [9].

Although DPPD is a rare entity, the diagnosis should be made if the following features are present: (a) A female patient with bilateral swelling of the parotid glands (b) no past history of any systemic disease such as Sjogren or sarcoidosis (c) MR findings of multiple, small areas of decreased signal within markedly enlarged parotid glands on precontrast short-TR images, with diffuse marked hyperintensity on long-TR images. This condition must be easily differentiated from other parotid gland enlargements as it is believed to be distinctive in its clinical and radiological features [9].

This was a rare case of DPPD affecting a male patient with characteristic features of PPD on MRI affecting both the parotid glands with secondarily infected unilateral parotid gland with no past history of any systemic disease.

The radiological features in the present case on ultrasound and MRI were characteristic of polycystic parotid disease with the rare occurrence in elderly male patient.

Microscopically according to the previous studies the whole of the gland may be occupied by cysts of varying size, which distend the lobules preserving the normal architecture of the parotid gland. Between the cysts, there are normal interlobular septa, residual serous acini, and excretory ducts. The cysts are lined by a single layer of flattened, cuboidal or columnar epithelial cells. In the case reported by Dobson and Ellis (1987), the columnar cells had more abundant eosinophilic cytoplasm with rounded luminal cell borders than intercalated duct lining cells. Stainable lipid in large amounts may be present in the epithelial cells. The cyst-lining cells may sometimes show pseudopapillary appearance. Occasionally, both striated ducts and distended acini can be seen opening into the cysts, while the ducts in remaining intact acini may be dilated. Inflammatory changes may be present if there is secondary infection. They are strongly eosinophilic and PAS-positive. They also stain with Congo red and show the apple-green birefringence characteristic of amyloid [9,12].

Although biopsy in DPPD was confirmatory our case showed characteristic radiological features on ultrasound and MRI which fulfilled the diagnostic criteria for DPPD as stated by Brown et al. In the present case, biopsy was not performed because the cysts were present at the junction of superficial and deep lobe of parotid gland thus would compromise the surrounding vital structures.

As the lesion is benign in nature, lobectomy or superficial parotidectomy is the treatment of choice. [7] However, enlargement of residual cystic tissue in the remaining lobes or further development of new lesions have not been reported. Our patient was 80 year old. Considering his age and location of multiple cysts bilaterally patient was instructed about the post surgical complications. Usually the surgery is performed for cosmetic purpose but our patient refused surgery and wanted the symptoms to be relieved. Observation in this case represents the therapeutic option as reported by Brown et al [9].

As not many cases of this nature have been reported in the literature long term follow up is required to monitor for the involvement of other salivary glands and to rule out recurrence.

In conclusion we report this rare case of DPPD which posed a challenge in clinical diagnosis and was confirmed by advanced imaging modality which could be misdiagnosed as a neoplastic salivary gland tumor.

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