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# Polycystic Disease of the Parotid Glands

Eugene Brown, Meredith August, Ben Z. Pilch, and Alfred Weber

**Summary:** A 31-year-old woman had bilateral swelling of the parotid glands at 4 months of pregnancy. MR imaging showed marked enlargement of the parotid glands with increased signal on images with long repetition times. A diagnosis of polycystic disease of the parotid gland was made after biopsy and histologic examination. The radiographic and histologic features of this rare disease are discussed.

**Index terms:** Salivary glands; Neck, cysts

Polycystic (dysgenetic) disease of the parotid glands is a rare disorder that has only recently been described in the literature. The disease is usually seen in female subjects and is bilateral in the majority of cases. In this report, we present a case of this unusual entity and review its radiographic features.

## Case Report

A 31-year-old woman was seen by her obstetrician at 16 weeks of pregnancy because of dramatic bilateral painless swelling of the parotid glands (Fig 1). Her medical history was significant for recurrent episodes of "swollen glands" as a child. During the previous episodes, the swelling had always regressed and was not as severe. A chest radiograph presentation was normal, and seventh-nerve function was intact. Physical examination revealed diffuse, homogeneous enlargement of the parotid glands with a soft doughy consistency to touch. There was no suggestion of a focal mass lesion. The flow from Stensen ducts was clear bilaterally and of normal quantity. No mouth dryness, eye problems, or arthralgias were present.

The lesions persisted over a 10-month period, during which time the patient gave birth to a healthy girl at term. Because of the persistent swelling, magnetic resonance (MR) and sialography were performed. MR (Fig 2) showed generalized enlargement of both parotid glands with involvement of the deep and superficial lobes. Precontrast T1-weighted images (600/15/1 [repetition time (TR)/echo time/excitations]) showed well-circumscribed glands with multiple small areas of decreased signal intensity produc-

ing a mildly inhomogeneous appearance. Long-TR images (2944/90/1) showed marked hyperintensity of the glands, and postcontrast short-TR images (600/15/2) showed diffuse contrast enhancement and poorly demonstrated small cystic areas. The sialogram showed marked stretching of the intraglandular ductal system with no evidence of sialectasis or ductal irregularity. Initially, a needle biopsy was attempted. It was inconclusive but showed evidence of chronic inflammation with some occasional areas of amyloid deposition. A biopsy of the lip performed at the same time disclosed no evidence of sarcoidosis or Sjögren disease and was otherwise normal. Approximately 2 months later, an open biopsy of the parotid gland was performed.

The biopsy (Fig 3) showed numerous small cysts lined by flattened to cuboidal epithelial cells. Occasional nuclei of the lining cells were enlarged, and some protruded into the lumen. Many cells were vacuolated, and numerous cells were sloughed into the cyst lumens. Residual normal-appearing acini could be seen scattered among the cysts. These features are characteristic of the histologic findings of polycystic disease of the parotid gland (1-3).

## Discussion

Polycystic (dysgenetic) disease of the parotid glands is a rare disorder that was described by Seifert et al (1) in 1981. Ten well-documented cases have been reported (2, 4). These cases have certain distinctive clinical features: (a) all but one of the reported cases occurred in female patients; (b) enlarged, nontender parotid swelling had been present for months or years; (c) involvement was bilateral; (d) recurrent parotid swelling often occurred during childhood; and (e) symptoms of this disorder sometimes were delayed until adulthood.

It is believed that the disease is inherited and might be sex linked (4). Normally, sex-linked traits are restricted to males. This disease is unusual in that the normal linkage appears reversed, with only females affected in several

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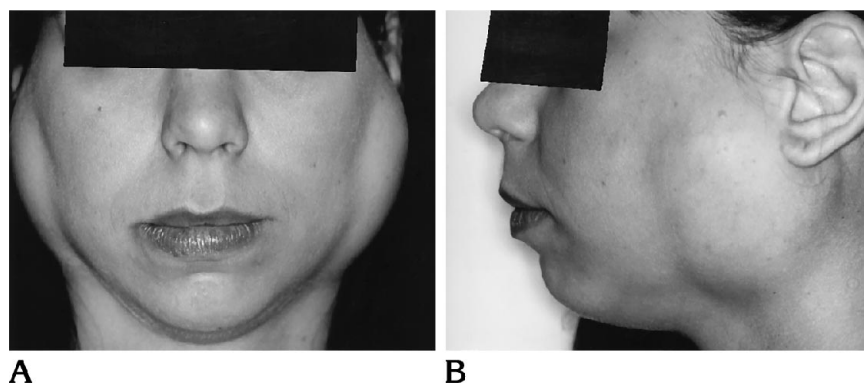


Fig 1. Photographs show marked enlargement of the parotid glands.

families. The condition is known to be familial, with one case having occurred in a mother and daughter. One of two cases reported by the Armed Forces Institute of Pathology did occur in a male subject (5). There is no known association between this entity and polycystic disease involving other organ systems, such as the kidneys or pancreas.

The pathologic findings in this condition are small epithelium lined cysts that replace most of the normal glandular parenchyma; only scattered normal acini are seen. Also, there are vacuolated cells and sloughing of cellular material into the cyst lumens. One puzzling feature of our case was the amyloidlike material (staining with Congo red) that was seen in the needle aspirate. Subsequently, an open biopsy showed this to represent eosinophilic spheruliths. These were seen in occasional ducts, and have been previously described (1-3). Interestingly, Dobson and Ellis (3) described such spheruliths to be congophilic and consis-

tent with amyloid, likely explaining the cytologic aspirate findings of amyloidlike material in the current case. The disorder is thought to arise from a developmental defect in the ductal system of the developing parotid gland (1).

The radiographic findings are consistent with the histologic appearance of this lesion. There is bilateral enlargement of the parotid glands with multiple cystic areas replacing the parenchyma. The MR findings in our case were distinctive. MR imaging showed markedly enlarged parotid glands with a mildly inhomogeneous appearance on short-TR images (600/15). There was uniform, generalized enhancement after injection of contrast material. On long-TR images (2944/90), there was striking, increased signal intensity throughout the entire substance of the glands, presumably reflecting the permeation of the cystic areas throughout the gland parenchyma and the relative paucity of unaffected acini (2). It has been suggested that the cytoplasm of the epi-



Fig 2. A, Axial short-TR image (600/15/1) shows mildly inhomogeneous signal in the parotid glands. B, Postcontrast coronal image (600/15/2) shows minimal diffuse enhancement. C, Axial long-TR image (2944/90/1) shows diffuse increased signal from the gland parenchyma.

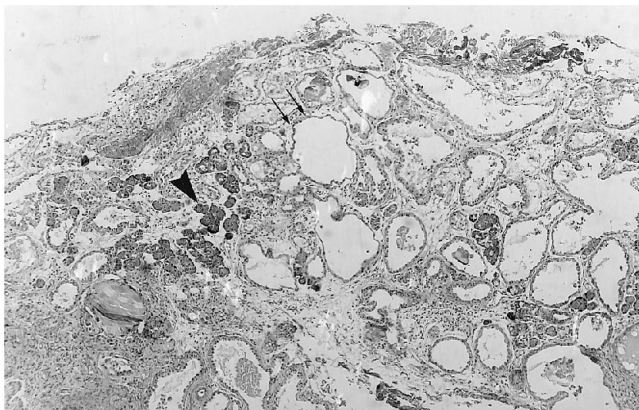
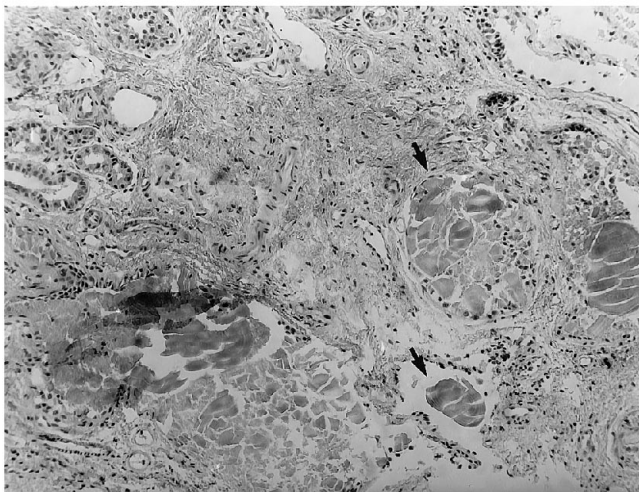
**A****B**

Fig 3. A, Dysgenetic polycystic parotid gland has numerous small cysts (arrows) and interspersed normal acini (arrowhead) (hematoxylin-eosin,  $\times 50$ ).

B, Congophilic spheruliths (arrows) are in some of the cysts (hematoxylin-eosin,  $\times 125$ ).

thelial cells, which line the cystic cavities, are hydropic and filled with secretory material (4). This marked hyperintensity on long-TR images might provide a unique differential feature for distinguishing this entity from other lesions.

CT scans were obtained in two previously reported cases. In one case (4), the appearance was similar to the findings seen on precontrast T1-weighted MR, showing glandular enlargement with multiple cystic areas. In the other case, reported by Mandel and Kaymar (6), the CT features were atypical and involved only one parotid gland. These authors noted several stones in the involved gland and reported communication of the intraparotid ducts with the cysts. These findings have not, however, been reported by other authors. In fact, this case was

not bilateral, occurred in a child, and resolved spontaneously. All of these features are unusual, raising the possibility that this case might have been misdiagnosed as polycystic disease. Furthermore, no pathologic confirmation was obtained. The sialographic findings in our case were of the intraparotid ducts being stretched and deformed by multiple intraparenchymal cysts. Although one report suggested irregularity and fine beading of the ducts, we did not observe this finding. Despite the severity of involvement, inflammatory changes are not prominent in glands or ducts. Dobson and Ellis (3) noted that there was no clinical or histologic evidence of inflammation in their case.

It is interesting that the association with pregnancy has not been previously reported. In our case, the symptoms clearly worsened with the onset of pregnancy. The patient has reported that within 4 to 6 months after delivery the symptoms (swelling) regressed, and she has otherwise been in good health. We speculate that hormonal changes that occur during pregnancy might cause expression or exacerbation of the underlying condition. Although we would like to restudy the patient's parotid glands, the patient declined further biopsy or MR studies. She no longer has swelling and does not want to repeat the MR study because of claustrophobia. She is concerned, however, about the possibility of a recurrence, especially if she should have another child. Also, she is concerned about the possibility of passing this condition on to her daughters.

The differential diagnosis in cases of bilateral parotid swelling is limited. The major differential diagnosis would be sialodochiectasis or chronic sialectasis. In sialectasis, there is a history of recurrent episodes in the past, associated with eating, and stone disease. Viral infections such as mumps can produce parotitis; however, there should be constitutional symptoms and the clinical diagnosis can be established without radiologic studies. Granulomatous disease such as sarcoidosis often will show involvement of other organ systems, which can be demonstrated by biopsy or radiographs. Autoimmune diseases such as Sjögren disease might be considered. However, such patients might have decreased salivary flow and associated systemic findings. Finally a diffuse, infiltrative malignant condition, such as lymphoma or leukemia, might be considered but would be unusual without other clinical symptoms of disease.

Although this is a rare entity, the diagnosis should be considered if the following features are present: (a) a female patient with bilateral swelling of the parotid glands; (b) no past history of systemic disease such as Sjögren or sarcoidosis; and (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. We believe that together, the clinical and radiographic features are distinctive and should allow differentiation of this entity from other lesions. Because sialograms in this disease do not have a characteristic appearance, we do not believe they are helpful in making the diagnosis except to exclude other conditions.

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