The Journal of Laryngology & Otology
November 2000, Vol. **114,** pp. 883–885

Misplaced parotid glands: bilateral agenesis of parotid glands associated with bilateral accessory parotid tissue

David Goldenberg, M.D., Renee Flax-Goldenberg, M.D.*, Henry Zvi Joachims, M.D., Nathan Peled, M.D.*

Abstract
Agenesis of the parotid is a rare entity. It is usually unilateral but bilateral cases have been described as a rare cause of xerostomia. Accessory parotid tissue is also uncommon and is not normally associated with aplasia of the parotid gland. The embryological, histological, clinical and radiological findings of these two entities will be discussed in addition to the report of an obscure case of bilateral parotid agenesis together with bilateral accessory parotid tissue.

Key words: Parotid Gland; Abnormalities

Introduction

Salivary gland embyrology

The epithelium of salivary glands is derived from the ectoderm of the oral cavity. These cells then differentiate into the ductal system, the acini, and other component cells of the salivary gland unit. Differentiation begins during the fourth to sixth week of development. The parotid gland buds are the first of the salivary glands to appear at the sixth week on the inner cheek near the angle of the mouth. They then migrate back toward the ear.[1]

Although the parotid anlagen are the first to develop, they become encapsulated after the submandibular and sublingual glands. This delayed encapsulation is critical because the lymphatic system develops within the mesoderm after the submandibular and sublingual glands encapsulate but prior to encapsulation of the parotid gland. The minor salivary glands do not start to develop until later.

Radiology

Over the past two decades a change has occurred in the preferred imaging modalities used to investigate patients with diseases of the major salivary glands. The old approach relied on plain films and sialograms, whereas today emphasis is on computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound. Although there are no set rules in the literature as to preference of one modality over another, the general consensus of many radiologists is that inflammatory diseases are best imaged by CT whereas tumours are best evaluated by MR.

Although most CT studies of salivary gland disease use contrast there often is no additional clinically useful information obtained over a noncontrast study. However, with MR imaging, contrast may help distinguish a cystic mass from a solid tumour and thus its routine use may be more justified than with CT. When contrast is used with MR imaging fat suppression sequences are necessary to improve lesion conspicuity.

Contrast is also very useful when evaluating perineural tumour spread from salivary gland malignancies.[2] Most CT studies of the major salivary glands and the neck are performed using axial 3 mm contiguous scans. Spiral or helical scans can also be used and are especially advantageous in children and patients who may have difficulty remaining quiet or motionless during the examination.[3]

MRIs of the major salivary glands are usually performed with 3 mm-thick slices. Non-contrast T1-weighted and T2-weighted sequences are obtained and then post-contrast T1-weighted, fat-suppressed images are obtained. Fat suppression may be needed for fast-spin echo T2-weighted scans as well. Axial views are obtained for all sequences, and coronal and sagittal views may be added as needed.[4]

Overall salivary masses are probably visualized better on MRI than on CT but calcifications are identified better on CT.[3]

CT and MRI are equally capable of demonstrating the anatomical absence of major salivary gland tissue where the glandular beds are replaced by fat. These modalities are useful for proving the absence of major salivary glands especially when accessory parotid tissue is present and visualization of the duct opening can mislead the observer on clinical examination (Figures 1 and 2).[5]

^{99}Tc-pertechnetate scintiscanning is a complimentary examination showing functioning salivary gland tissue which is beyond the scope of CT or MRI. The radionucleotide is accumulated by salivary gland epithelium, maximum activity in the gland is attained 20 minutes after injection.[6]

Case report

A 10-year-old boy presented after his family noticed bilateral facial swelling. The duration of this swelling was uncertain.

From the Department of Otolaryngology – Head and Neck Surgery, Rambam Medical Center Technion Faculty of Medicine, Haifa, and the Department of Radiology*, Carmel Medical Center, Haifa, Israel.
Accepted for publication: 22 June 2000.

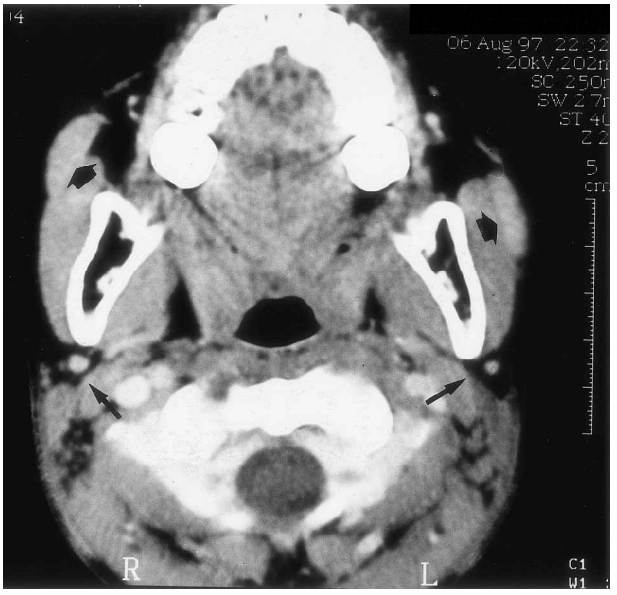

FIG. 1
Axial CT scan of the face shows agenesis of the parotid glands (arrows), and ipsilateral accessory parotid glands (thick arrows).

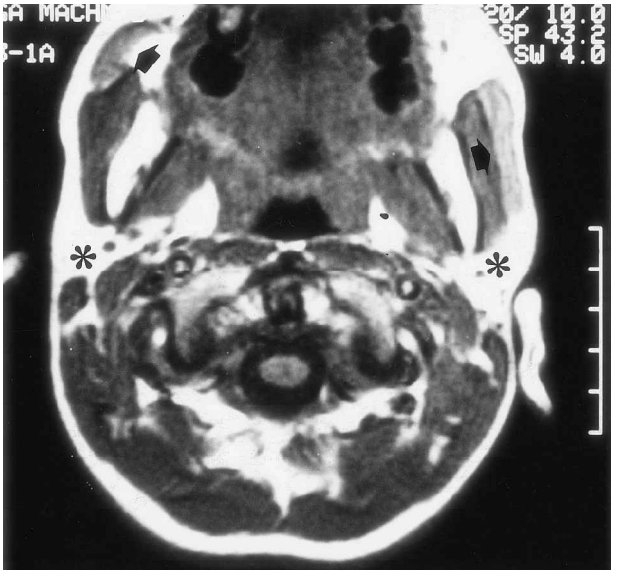

FIG. 2
MRI T1-weighted sequence of the face shows agenesis of the parotid gland (asterix) and ipsilateral accessory parotid tissue.

His medical and family history was normal and he was not being treated with any medication. There was no complaint of pain or xerostomia. Ophthalmological examination was normal with normal tear production. On physical examination there was diffuse non-tender swelling over the body of both sides of the mandible, more prominent on the right side. There were no mandibular, facial or auricular malformations and there were no enlarged cervical nodes or masses on palpation. Intra-oral examination revealed an adequate amount of saliva and the dental decay rate was not excessive. Stenson's papilla was small and anteriorly placed on both sides. The rest of the physical examination was normal.

A computerized tomography (CT) scan was performed (Figure 1) showing bilateral absence of the parotid glands, the glandular bed replaced with fat. In addition bilateral ectopic glandular tissue was noted anterior to the masseter muscles on both sides of the face. A magnetic resonance imaging (MRI) scan of the face was performed (Figure 2) to verify these findings. It revealed fat in the parotid beds and bilateral accessory glandular tissue anterior to the masseter muscles. Based on these findings a diagnosis of bilateral agenesis of the parotid glands with bilateral accessory parotid tissue was made. The parents of the child refused any further diagnostic examinations.

Discussion

Hypoplasia agenesis of salivary glands

Hypoplasia or absence of the major salivary glands with or without the absence of Stensen's duct has been documented and may be partial or total. It is not usually associated with accessory or ectopic salivary tissue. Familial absence of salivary glands was first described by Ramsey.[7] A familial incidence of parotid agenesis was noted in one family with seven patients spanning three generations. All of the individuals had xerostomia and both parotid glands and ducts were absent.[8] Whyte[6] and later Wiedemann[9] found this disorder to be a complex, pleotropic autosomal dominant disorder, more commonly found in males,[10] and may occur together with first and second branchial arch anomalies, mandibulofacial dystosis and hemifacial microsomia.[6] Other conditions associated with agenesis of major salivary glands include lacrimal apparatus malformations such as agenesis of the lacrimal glands and aplasia of the lacrimal punta.[11] This combination is now known as lacrimo-auriclo-dentodigital (LADD), or Levy-Hollister Syndrome. It is a true multiple congenital anomaly (MCA) syndrome comprising hypoplasia, aplasia or atresia of the lacrimal system, anomalies of the ears and hearing loss, hypoplasia, aplasia or atresia of the salivary system, dental anomalies and digital malformations including absent, hypoplastic or triphalangeal thumbs. It is also an autosomal dominant disorder.[9]

Clinical suspicion of gland agenesis should arise when the papillae of the salivary glands are absent and in cases of xerostomia in which more common causes have been excluded. Total parotid agenesis may lead to reduction in salivary flow and thus may lead to dental disease including caries and infection.[6] The submandibular, sublingual and minor salivary glands contribute most significantly to the total resting salivary flow. Therefore unilateral or partial parotid agenesis may not be noticed by the patient.[8]

In the case we report there was no xerostomia presumably because of the remaining normally functional salivary tissue and possibly functional ectopic tissue as well.

Accessory salivary tissue

The existence of salivary tissue in locations other than the major salivary glands (parotid, submaxillary and sublingual) or the minor salivary glands of the oral cavity, pharynx and upper airway is known as heterotropia.[12] Heterotropic or accessory salivary gland tissue has been reported in numerous sites throughout the body including the hypophysis, middle ear, external ear canal, mandible, thyroglossal duct, capsules of the thyroid and parathyroid glands, lymph nodes and in the upper and lower neck regions.[12,13] In the neck nearly all heterotropias have been described along the anterior border of the SCM muscle, particularly around the sterno-clavicular joint.[13] Accessory parotid tissue has been described as collections of parotid tissue, unilateral and separate from the main gland. The tissue typically lies anterior to the parotid gland along the masseter muscle cephalad to Stenson's duct, along the course of the buccal branch of the facial nerve.[14] The

https://doi.org/10.1258/0022215001904257 Published online by Cambridge University Press

accessory tissue is usually 3 cm or less in diameter. Although the accessory tissue is not continuous with the main parotid tissue it usually drains alongside the main parotid gland into Stenson's duct.

The precise embryogenesis of accessory SGT is unknown and most theories remain speculative. Accessory salivary gland tissue is thought to arise either from salivary inclusions within lymph nodes[15] or from anomalous downward migration of salivary tissue.[16]

Accessory parotid glands are found in approximately 20 per cent of the population.[17] Nemecek found accessory parotid tissue to be histologically identical to normal parotid gland tissue[18] while in a large study on cadavers Toh found 26 per cent of the accessory parotid tissue to be histologically different from the main parotid tissue, containing both mucus and serous acini (i.e. mixed acini) as opposed to the serous nature of the main parotid gland. Toh found that 50 per cent of accessory parotid glands are either round or triangular in shape with 100 per cent containing anastomosis of the zygomatic and buccal branches of the facial nerve buried within them.[19]

Accessory parotid tissue is susceptible to most salivary pathologies. Between one and seven per cent of all parotid neoplasms arise from accessory glands, 50 per cent of these are histologically malignant.[20] The surgical anatomy of accessory parotid tissue is important, since the failure to remove this tissue may be associated with recurrence of neoplastic lesions.[21]

References

1 Sperber GH. In: Derrick D, ed. *The Salivary Glands in Craniofacial Embryology*. London: Wright, 1989:188–91
2 Parker GD, Harnsberger HR. Clinical-radiologic issues in perineural tumor spread of malignant diseases of the extracranial head and neck. *Radiographics* 1991;**11**:383–99
3 Silvers AR, Som PM. Head and neck imaging – salivary glands. *Radiol Clin North Am* 1998;**36**:941–66
4 Chaudhuri R, Bingham JB, Crossman JE, Gleeson MJ. Magnetic resonance imaging of the parotid gland using the STIR sequence. *Clin Otolaryngol* 1992;**17**:211–7
5 Bhide VN. Agenesis of the parotid gland: Association with ipsilateral accessory parotid tissue. *Am J Radiol* 1998;**170**:1670–1
6 Whyte AM, Hayward MW. Agenesis of the salivary glands: a report of two cases. *Br J Radiol* 1989;**62**:1023–6
7 Ramsey WR. A case of hereditary congenital absence of the salivary glands. *Am J Dis Child* 1924;**28**:440
8 Weisenfeld D, Ferguson MM, Allan CJ, McMillan NC, Scully CM. Bilateral parotid gland aplasia. *Br J Oral Surg* 1983;**21**:175–8
9 Wiedemann HR. Salivary gland disorders and heredity. *Am J Med Genet* 1997;**68**:222–4
10 Smith NJD, Smith PB. Congenital absence of the major salivary glands. *Br Dental J* 1977;**142**:259–60
11 Gomez RS, Aguiar MJ, Ferreira AP, Castro WH. Congenital absence of parotid glands and lacrimal puncta. *J Clin Pediatr Dentistry* 1998;**22**:247–8
12 Batsakis JG. Heterotopic and accessory salivary tissues. *Ann Otol Rhinol Laryngol* 1986;**95**:434–5
13 Tay HL, Howitt MB. Heterotopic pleomorphic adenoma in the neck. *J Laryngol Otol* 1995;**109**:445–8
14 Toh H, Kodama J, Fukuda J, Rittman B, Mackenzie I. Incidence and histology of human accessory parotid glands. *Anat Rec* 1993;**236**:586–90
15 Youngs LA, Scofield HH. Heterotopic sailvary gland tissue in the lower neck. *Arch Pathol* 1967;**83**:550–6
16 Sternberg SS. *Histology for Pathologists*. New York: Raven Press, 1992
17 Frommer J. The human accessory parotid gland: Its incidence, nature, and significance. *Oral Surg Oral Med Oral Pathol* 1977;**43**:671–6
18 Nemecek JR, Marzek PA, Young VL. Diagnosis and treatment of accessory parotid gland masses. *Ann Plast Surg* 1994;**33**:75–9
19 Toh H, Hamada N, Ohmori T. Accessory parotid gland in man. *J Fukouka Dent Coll* 1984;**11**:38–43
20 Batsakis JG. Accessory parotid gland. *Ann Otol Rhinol Laryngol* 1988;**97**:434–5
21 Ellis GL, Auclair PL, Douglas RG. *Surgical Pathology of the Salivary Glands*. Philadelphia: W. B. Saunders Co., 1991

Address for correspondence:
Dr David Goldenberg,
Freud St 32,
Haifa,
Israel.

Fax: 972 4 8346781
E-mail: gdavid@tx.technion.ac.il

Dr D. Goldenberg takes responsibility for the integrity of the content of the paper.
Competing interests: None declared