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Agenesis of the unilateral parotid gland associated with pleomorphic adenoma of the contralateral parotid gland

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Abstract
Congenital absence of the parotid gland is extremely infrequent. We present here a case of unilateral parotid gland agenesis with pleomorphic adenoma of the contralateral parotid gland. Even though pleomorphic adenoma is the most common tumour of the parotid gland, to our knowledge this is the first case of these two conditions being seen together.

Key words: Parotid Gland; Abnormalities; Pleomorphic; Adenoma; Magnetic Resonance Imaging

Introduction

Congenital absence of the major salivary glands, particularly of the parotid gland, is an unusual disorder and only a few cases have been reported so far. Agenesis of the parotid gland may be unilateral or bilateral and it may occur alone or with agenesis of the other major salivary glands or with other congenital abnormalities of the head and neck.[1–7] Although pleomorphic adenoma is the most common benign tumour of the parotid gland in adults, it has not been reported in the sole parotid gland of a patient before. We present here a case of unilateral parotid gland agenesis with pleomorphic adenoma of the contralateral parotid gland.

Case report

A 35-year-old woman presented with a one-year history of a slowly enlarging, painless mass on the left parotid region. Furthermore, she had a history of intra-oral dryness in the right part of her mouth. Her medical history was otherwise unremarkable. On examination, there was a solid, mobile, non-tender mass in the left parotid tail with no evidence of facial weakness. There were no remarkable developmental anomalies of the head and neck. Intra-oral examination revealed the absence of the orifice of the right Stensen's duct. The saliva on the right side of the oral mucosa was inadequate, but the dental decay rate was not excessive. The results of an ophthalmological examination and rate of tear production were normal. Ultrasonographic examination of the left parotid gland showed a well-defined, hypoechoic, solid mass with a size of 17 × 18 × 33 mm, while the right parotid gland was not visualized. The patient was evaluated with magnetic resonance imaging (MRI) before and after intravenous contrast administration. A hyperintense, well-demarcated mass involving both deep and superficial lobes of the left parotid gland was seen on fat-saturated T2-weighted images (TR: 4000; TE 80) (Figure 1). On T1-weighted magnetic resonance images (TR: 600; TE: 15), the left parotid mass was seen to be hypointense when compared with the rest of the gland, and showed heterogeneous contrast enhancement after contrast agent injection. MRI did not identify the right parotid gland on the head and neck region. Salivary gland scintigraphy with tecnetium-99m pertechnetate showed a hypoactive region in the left parotid gland and no activity in the right parotid gland, with normal filling and emptying of the other salivary

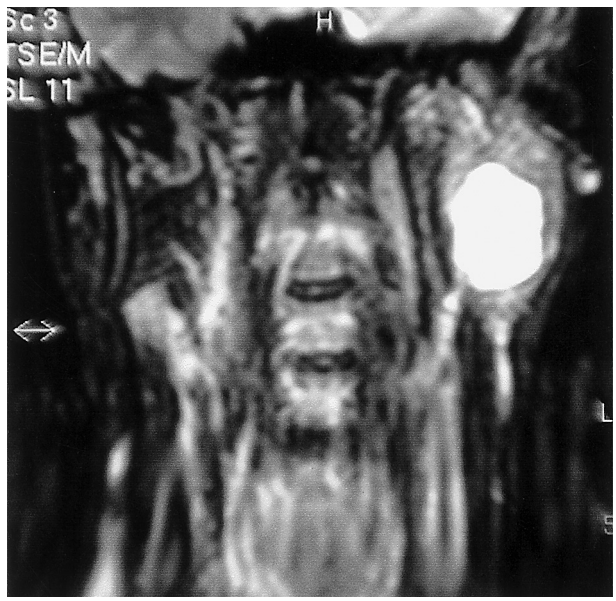

Fig. 1

A 35-year-old woman with left parotid mass. The fat-saturated turbo spin echo T2-weighted (TR: 4000, TE: 80) coronal image through the parotid gland demonstrates a well-defined hyperintense mass within the left parotid gland. The right parotid gland is not visualized either in the right parotid region or in the head and neck region.

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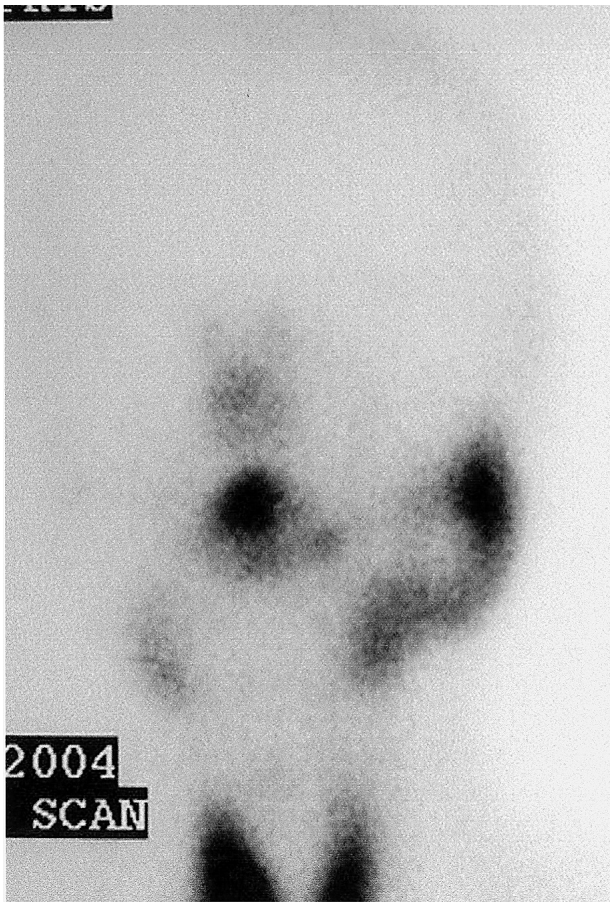

FIG. 2

Technetium-99m pertechnetate scintigraphy reveals a hypoactive area in the left parotid gland with the absence of right parotid activity, while other salivary glands have normal activity.

glands (Figure 2). No ectopic activity was detected. Fine-needle aspiration biopsy was performed on the left parotid gland mass and it was diagnosed as pleomorphic adenoma. The tumour was removed with an intact cuff of healthy tissue around it by a subtotal parotidectomy procedure. Histopathological examination of the specimen confirmed the fine-needle aspiration biopsy result.

- **Congenital absence of the parotid gland is a rare entity**
- **In this case parotid agenesis was associated with a contralateral parotid pleomorphic adenoma**
- **The clinical and radiological features are discussed**

Discussion

Congenital absence of the parotid glands is very rare. The most complete review of congenital salivary gland agenesis has been that of Salvinelli *et al*., who summarized the 28 cases known at that time.[1] Of those, only four cases were isolated as unilateral parotid gland agenesis. Actually, it is very difficult to ascertain the true incidence of parotid gland agenesis because it is usually asymptomatic.[1,2]

Agenesis of the parotid gland can be isolated or associated with other congenital abnormalities, including the absence of other major salivary glands, first and second branchial arch anomalies, and craniofacial deformities.[1–4] In addition, agenesis of the salivary glands may be combined with absent or defective lacrimal glands and/or ducts.[5] Salivary system agenesis may also be seen as a component of the lacrimoauriculodentodigital (LADD) or Levy-Hollister syndrome, which comprises lacrimal system developmental anomalies, ear anomalies and hearing loss, dental anomalies and digital malformations.[8] Our patient did not have any of these.

In the case of parotid gland agenesis xerostomia, excessive dental caries, periodontal diseases or oral candidiasis may occur due to the lack of saliva.[2,5] The patient may complain about gradually enlarging painless swelling in the contralateral parotid region due to the compensatory hypertrophy of the sole parotid gland.[2,6] The present case had xerostomia due to gland agenesis, but no related oral diseases. Despite most of the present salivary gland being involved in the pleomorphic adenoma, compensatory hypertrophy was not seen in the normal salivary gland tissue.

Although an absence of the orifice of the Stensen's duct can be taken as evidence of a developmental defect of the gland, it is usually overlooked by the clinician. Therefore, the diagnosis of parotid gland agenesis is based on imaging studies. Moreover, because it is usually asymptomatic, most of the time it is diagnosed incidentally while evaluating the other gland. Ultrasound imaging is usually the first choice in the evaluation of a parotid swelling, but it should be carried out not only on the affected side but also on the contralateral side in order to detect parotid gland agenesis. Computed tomography and MRI are also capable of showing agenesis of the salivary glands. Salivary gland scintigraphy with Tc-pertechnetate is a complementary**[Q2]** examination showing the absence of a functioning salivary gland, the presence of functioning salivary glands and, if present, heterotopic salivary tissue.

Pleomorphic adenoma is the most common salivary gland tumour, which constitutes about half of all tumours and 65 per cent of parotid gland tumours.[9] To our knowledge, this is the first case of pleomorphic adenoma in parotid gland associated with agenesis of contralateral parotid gland to be reported in the medical literature. The association of these two conditions possibly causes an increase in the severity of xerostomia after treatment of the pleomorphic adenoma. In order to avoid diminution in the production of saliva, we preferred a subtotal parotidectomy procedure saving as much of the normal parotid gland tissue as possible. The patient did not note any change in the degree of xerostomia after the surgery.

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