CASE REPORT

CT and MR findings of bilateral submandibular gland aplasia associated with hypertrophied symmetrical sublingual glands herniated through mylohyoid defects

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There are very limited reports of aplasia of bilateral submandibular glands associated with bilateral sublingual gland hypertrophy. We report the case of a 13-year-old girl with bilateral submandibular gland aplasia associated with symmetrical sublingual gland hypertrophy that herniated from bilateral mylohyoid defects and presented as bilateral masses on CT and MRI images. Prolapsed bilateral sublingual gland hypertrophy should be considered in patients with bilateral submandibular masses to avoid unnecessary biopsy or surgery. We suggest that radiological evaluation of these cases should be performed before any intervention. *Dentomaxillofacial Radiology* (2012) **41**, 79–83. doi: 10.1259/dmfr/23245765

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Introduction

Major salivary gland aplasia is frequently seen in some congenital disorders including lacrimoauriculodentodigital (LADD) syndrome, mandibulofacial dysostosis and ectodermal dysplasia associated with aplasia or dysplasia of lacrimal or thyroid glands.^{1–3} However, isolated aplasia of major salivary glands is a very rare entity that is most commonly seen in parotid glands.⁴ Clinically, patients may be asymptomatic or may present with dryness of the mouth, have difficulty in chewing and swallowing and suffer from dental caries. Also, pseudomasses can be seen as a result of compensatory hypertrophy of the contralateral or other major salivary glands, or accessory salivary tissue.⁵

The muscular base of the oral cavity is formed by the mylohyoid muscle that forms a sling inferior to the tongue. It inserts on the slightly obliquely orientated mylohyoid line on the medial surface of the mandible, with the posterior aspect more cranial than the anterior aspect. The muscle is thickest posteriorly, and thins as it approaches the mental tubercle. Defects of this muscle are common; radiologically detectable deficiencies were

identified in three of four individuals in a previous study.⁶

To the best of our knowledge, there are very limited cases of isolated aplasia of bilateral submandibular glands associated with bilateral sublingual gland hypertrophies in the literature.^{7,8} In those reports, CT findings were reported in adult patients. Ahmed et al reported a case of non-syndromic bilateral submandibular gland aplasia with hypertrophied sublingual salivary tissue, the latter herniating through the mylohyoid boutonnière to present as a palpable mass on the left side with corresponding CT findings.⁷

In this report, we present CT and MRI findings of a 13-year-old girl with bilateral submandibular gland aplasia associated with symmetrical sublingual gland hypertrophy that herniated from bilateral mylohyoid defects presenting as bilateral masses.

Case report

A 13-year-old girl was referred for CT examination for bilateral submandibular masses pronounced with swallowing. She was otherwise healthy and first became aware of the masses 1 year previously. She had no history of head and neck cancer or surgery. No dental

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a **b**

Figure 1 Bilateral submandibular masses of the patient. No abnormality is seen at rest (a); however, masses appear following modified Valsalva movement (b)

a **b** **c** **d** **e** **f**

Figure 2 Consecutive contrast-enhanced transverse CT images from the submandibular region. There is fat but no submandibular glands in the expected location of glands (arrows). Bilateral hypertrophied salivary glands are seen (stars)

caries was noticed. Bilateral non-tender submandibular masses were noticed that became pronounced with modified Valsalva movement on physical examination (Figure 1). No sign of inflammation was observed. There was no adenopathy, and oral, pharyngeal and maxillofacial examination revealed no pathological finding.

Sonography was performed for initial radiological evaluation. Bilateral submandibular glands were not identified and symmetrical masses with glandular echogenicity were seen anterior to the expected locations of submandibular glands. No lymphadenopathy was detected. A routine CT of the soft tissues of the neck with intravenous contrast was performed. On the CT there were no pathological lymphadenopathy or masses. Submandibular glands could not be identified on transverse, multiplanar or maximum intensity projection (MIP) images (Figure 2). Bilateral enhancing hypertrophied sublingual salivary tissue accompanied by symmetrical mylohyoid defects was seen (Figure 3). Both sublingual glands protruded to the submandibular space through the defects. A non-contrast MR study was performed to

verify the diagnosis with oral consent of the patient and her parents. Transverse T_1 weighted and fat suppressed proton density images confirmed the CT findings (Figure 4). On both CT and MRI, hypertrophied sublingual glands were homogeneous and had distinct margins. There was no radiological sign of inflammation or tumour.

Discussion

Agenesis or aplasia of the major salivary glands is very rare and has been mainly described for parotid and submandibular glands, more commonly in the former. When present, it is usually associated with multiple other developmental head anomalies.¹⁻³ Unilateral absence of submandibular gland may be associated with hypertrophy of the contralateral counterpart or ipsilateral sublingual gland.⁵ In previous reports of bilateral submandibular gland aplasia, bilateral sublingual hypertrophy accompanied this congenital absence.^{7,8}

Figure 3 Maximum intensity projection (MIP) oblique transverse CT image (slab thickness: 17.6 mm) shows bilateral mylohyoid (M) and hypertrophied sublingual glands (lines)

Figure 4 Transverse T_1 weighted (a) and fat suppressed proton weighted MR images (b) demonstrate homogeneous-hyperintense and hypertrophied sublingual glands on each side (stars)

Pathogenesis of bilateral salivary gland aplasia is unknown; however, it most likely results owing to a disturbance during the foetal development.⁹ During the fourth to eighth weeks of foetal development, the major salivary glands form from oral ectoderm proliferation and outpouching. The submandibular glands develop after the parotid glands, and the sublingual and minor salivary glands develop last.¹⁰

Submandibular gland aplasia may be associated with severe caries in mandibular permanent incisor teeth as well as other signs of salivary hypofunction, such as dry mouth, disturbed oral sensation and oral infections.¹¹ Mathison et al reported bilateral submandibular gland aplasia in a patient with CT findings.⁸ Their 34-year-old female patient had accompanying nodular-sclerosing Hodgkin lymphoma but no sign of dysphagia, dry mouth or extensive dental caries. Ahmed et al also did not report those findings in their 62-year-old male patient.⁷ Similar to these reports, our patient did not suffer any problem related with salivary deficiency. We think that the common finding of their and our cases, namely bilateral sublingual gland hypertrophy, may compensate for any salivary hypofunction.

White et al reported the deficiencies in the mylohyoid muscle in 77% of individuals who underwent CT scanning.⁶ They showed that the defects may contain fat, blood vessels, salivary tissue or all three characteristics. Accessory salivary tissue was identified in 37% of individuals who underwent scanning in their report. Hypertrophied sublingual glands may locate in the mylohyoid defects and herniate to the submandibular

space. However, this extremely rare entity was reported only once in the literature by Ahmed et al.⁷ They reported a case of non-syndromic bilateral submandibular gland aplasia with hypertrophied sublingual salivary tissue, the latter herniating through the mylohyoid boutonnière to present as a palpable mass on the left side with corresponding CT findings. We present a paediatric patient who had bilateral palpable and visible masses with modified Valsalva movement with CT and MRI findings. As far as we know, there are no previously reported cases of bilateral submandibular gland aplasia in a paediatric patient. Fat-suppressed proton density transverse MR images clearly showed the hypertrophied sublingual glands (Figure 4). Also, transverse CT and T_1 weighted MR images demonstrated the bilateral mylohyoid defects (Figure 3).

Salivary gland aplasia can be diagnosed with a variety of imaging techniques, which include CT, MRI, sonography, sialography or nuclear medicine (technetium $^{99}\text{Tc}^{\text{m}}$ -pertechnetate) studies.⁵ Among these methods, sonography is cheap, non-invasive and widely available, and thus very important. As in our case, many radiologists may prefer MRI and CT for the exact documentation of this abnormality. However, an elaborate sonographic examination by an experienced physician may be adequate.

Inflammations—in particular, viral sialadenitis, tumours and dermoid cysts—should be included in the differential diagnosis of sublingual gland enlargement. Of these, bilateral enlargement is most likely to be seen in viral sialadenitis. In such cases, clinical and imaging findings would be helpful for correct diagnosis.

Conclusion

Symmetrical submandibular gland aplasia associated with sublingual gland hypertrophy should be kept

in mind in patients with bilateral submandibular masses to avoid unnecessary biopsy or surgical interventions. CT or MRI should be performed before any intervention.

References

1. Inan UU, Yilmaz MD, Demir Y, Degirmenci B, Ermis SS, Ozturk F. Characteristics of lacrimo-auriculo-dento-digital (LADD) syndrome: Case report of a family and literature review. *Int J Pediatr Otorhinolaryngol* 2006; **70**: 1307–1314.
2. Haktanir A, Degirmenci B, Acar M, Albayrak R, Yucel A. CT findings of head and neck anomalies in lacrimo-auriculo-dento-digital (LADD) syndrome. *Dentomaxillofac Radiol* 2005; **34**: 102–105.
3. Singh P, Warnakulasuriya S. Aplasia of submandibular salivary glands associated with ectodermal dysplasia. *J Oral Pathol Med* 2004; **33**: 634–636.
4. Wiesenfeld D, Ferguson MM, Allan CJ, McMillan NC, Scully CM. Bilateral parotid gland aplasia. *Br J Oral Surg* 1983; **21**: 175–178.
5. Srinivasan A, Moyer JS, Mukherji SK. Unilateral submandibular gland aplasia associated with ipsilateral sublingual gland hypertrophy. *AJNR Am J Neuroradiol* 2006; **27**: 2214–2216.
6. White DK, Davidson HC, Harnsberger HR, Haller J, Kamya A. Accessory salivary tissue in the mylohyoid boutonniere: A clinical and radiologic pseudolesion of the oral cavity. *AJNR Am J Neuroradiol* 2001; **22**: 406–412.
7. Ahmed M, Strauss M, Kassaie A, Shotelersuk V, DeGuzman R. Bilateral submandibular gland aplasia with clinico-radiological mass due to prolapsing sublingual salivary tissue through mylohyoid boutonniere: A case report and review. *Dentomaxillofac Radiol* 2009; **38**: 121–124.
8. Mathison CC, Hudgins PA. Bilateral submandibular gland aplasia with hypertrophy of sublingual glands. *Otolaryngol Head Neck Surg* 2008; **138**: 119–120.
9. Matsuda C, Matsui Y, Ohno K, Michi K. Salivary gland aplasia with cleft lip and palate: A case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1999; **87**: 594–599.
10. Strong BE, Johns ME. Anatomy and physiology of the salivary glands. In: Bailey B, Newlands SD, editors. *Head and neck surgery-otolaryngology*. Philadelphia: Lippincott, Williams and Wilkins, 2006, pp 517–521.
11. Fracaro MS, Linnett VM, Hallett KB, Savage NW. Submandibular gland aplasia and progressive dental caries: A case report. *Aust Dent J* 2002; **47**: 347–350.