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Case Report

Bilateral submandibular gland agenesis with bilateral parotid hypertrophy: a rare presentation

Rakhi Kumari*, Priyanko Chakraborty, Sidharth Pradhan, Purnima Joshi, Rajiv Kumar Jain

Department of Otorhinolaryngology, Institute of Medical sciences, Banaras Hindu University, Varanasi-221005, India

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***Correspondence:**
Dr. Rakhi Kumari,
E-mail: dr.rakhi.kumari@gmail.com

ABSTRACT

Recurrent laryngeal nerve injury and hypoparathyroidism are two of the much discussed major complications Bilateral submandibular gland aplasia associated with bilateral hypertrophy of parotid gland is one of the rarest case presentation. The enlargement of parotid gland is mostly due to a compensatory mechanism of submandibular aplasia. This case report describes a 45 year female presented with diffuse swelling over bilateral preauricular and infraauricular area. CECT of face confirmed diffuse bilateral parotid gland enlargement associated with absence of submandibular gland.

Keywords: Submandibular aplasia, Parotid hypertrophy, Cheek swelling

INTRODUCTION

Congenital absence of major salivary glands is a very rare disorder. They may be asymptomatic or may present with the complaints of dry mouth, difficulty in chewing and swallowing. Reduced saliva production decreases the protective effect of saliva within the oral cavity leading to increased incidence of dental caries. We present a unique case of bilateral submandibular gland aplasia associated with bilateral hypertrophied parotid glands, which has not been reported previously. Very few cases of bilateral submandibular gland aplasia, albiet with compensatory sublingual hypertrophy have been previously reported.[1]

CASE REPORT

A 45 year old female, presented in the OPD with chief complaint of bilateral cheek swelling for 3 years. On examination, there was diffuse swelling over bilateral pre-auricular and infra-auricular areas, which were soft in consistency, non tender and not fixed to skin (Figure 1).

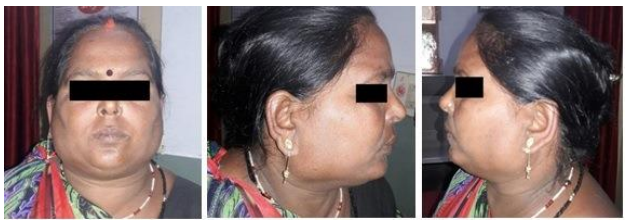

Figure 1: A, B and C shows front, right lateral and left lateral view of patient with swelling in parotid area.

There was no history of fever, pain, or facial asymmetry. The clinical impression was that of an enlarged parotid gland, and hence a fine needle aspiration was carried out. The cytological analysis confirmed it to be containing benign salivary gland cells. CECT of face revealed

diffuse enlargement of bilateral parotid glands without any mass lesion or inflammatory changes. Parotid ducts of both sides were of normal calibre without any evidence of stricture or stone. Submandibular glands were not visualised on either side (Figure 2 and 3). All other investigations were normal. As the patient had no any complain related to decreased salivation hence she was conservatively managed and kept under follow up.

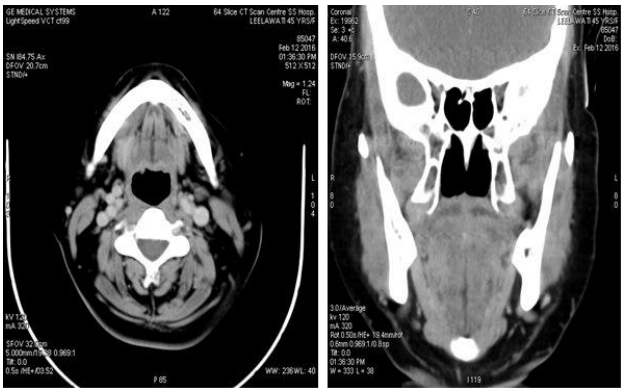

Figure 2: (A) Axial and (B) coronal CT scan showing absence of bilateral submandibular gland.

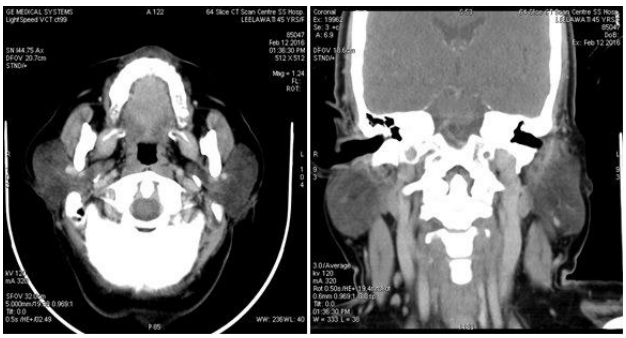

Figure 3: (A) Coronal and (B) axial CT scan of patient showing bilateral parotid hypertrophy.

DISCUSSION

Major salivary glands develop from the ectoderm. The salivary glands arise as buds from the in growth of epithelial lining of the oral cavity into the underlying mesenchyme. These primordial buds grow and extend into the underlying mesenchymal tissue and undergo extensive branching to form the ductal system of the glands. These glands are divided into lobules by the surrounding mesenchyme. Parotid glands appear during the fourth week, submandibular glands during the sixth week, and the sublingual glands during the ninth week of gestation.[2]

Major salivary gland aplasia may be an isolated finding or seen in association with Lacrimo Auriculo Dento Digital (LADD) syndrome, mandibulo-facial dysostosis and ectodermal dysplasia associated with aplasia or dysplasia of lacrimal gland.[3-5] LADD syndrome, is characterized by hypoplasia, aplasia or atresia of the lacrimal system, deafness, aural malformations, and dental and digital anomalies.[6]

The exact incidence of major salivary gland agenesis is not known due to the asymptomatic nature of most of the cases.[7] Isolated agenesis of parotid gland is more common than that of submandibular gland.[8] Isolated submandibular gland agenesis is very rare with only a few cases reported till date.

Patients with major salivary gland agenesis may remain asymptomatic or may present with complaints of dryness in mouth, difficulty in swallowing and chewing, and dental caries. Patients may present with complaint of mass in neck due to compensatory hypertrophy of other major salivary glands.[9] Most of the cases of congenital absence of major salivary glands are associated with other anomalies. Very few cases of isolated bilateral submandibular gland agenesis have been reported in association with bilateral sublingual gland hypertrophy.[1]

CONCLUSION

The above mentioned patient had no complaints related to decrease in salivation and the case is unique due to the presentation of isolated non-syndromic agenesis of bilateral submandibular glands in association with bilateral parotid gland hypertrophy. The enlargement of the parotid gland is most likely due to the compensatory hypertrophy.

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REFERENCES

1. Haktanır A. CT and MR findings of bilateral submandibular gland aplasia associated with hypertrophied symmetrical sublingual glands herniated through mylohyoid defects. Dentomaxillofacial Radiology. 2012;41:79–83.
2. Khaladkar SM, Ayush M, Chaudhary K, Thakkar DK. Isolated unilateral Non -Syndromic non-familial hypoplastic Parotid gland: Case Report. International J Healthcare and Biomed Res. 2014;2(3):88-93.
3. 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;70:1307–14.
4. 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;34:102–5.
5. Singh P, Warnakulasuriya S. Aplasia of submandibular salivary glands associated with ectodermal dysplasia. J Oral Pathol Med. 2004;33:634–6.

6. Hollister DW, Klein SH, De Jager HJ, Lachman RS, Rimoin DL. The lacrimo-auriculo-dento-digital syndrome. J Pediatr 1973;83(3):438–44.
7. Hodgson TA, Shah R, Porter R. The investigation of major salivary gland agenesis: A case report. J Oral pathology; Pedeatric Dentistry. 2001;23:131-4.
8. Gangidi SR, Cunliffe D. Hyperpalstic sublingual gland in association with a congenitally missing ipsilateral submandibiular gland. Journal of Surgical Case Reports. 2011;9(5);1-3.
9. Srinivasan A, Moyer JS, Mukherji SK. Unilateral submandibular gland aplasia associated with ipsilateral sublingual gland hypertrophy. AJNR Am J Neuroradiol. 2006;27:2214–6.

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