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Case Report

Unilateral submandibular gland aplasia with ipsilateral parotid gland hypertrophy presenting as infra-auricular swelling

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ABSTRACT

Congenital absence of any one of the major salivary glands is usually asymptomatic per se but may come to the fore during evaluation of a relatively insignificant consequence it may lead to. We report a rare case of unilateral submandibular gland aplasia presenting with a long-standing ipsilateral infra-auricular facial swelling due to compensatory secondary hypertrophy of the parotid gland, which was mistaken as recurrent parotitis. This was evident on computed tomography as an absent right submandibular gland with smooth bulky ipsilateral parotid having no ductal dilatation or periglandular inflammation. An awareness of this etiology, though rare, would be helpful to both the radiologist and the referring physician as it would prevent an unnecessary biopsy of the enlarged parotid and mitigate the scare arising out of the clinical findings of an infra-auricular swelling.

Keywords: Submandibular, Hypoplasia, Parotidomegaly, Compensatory

INTRODUCTION

Oral saliva serves many purposes including mouth lubrication during chewing, preventing dryness and also serving microbicidal function. It is adequately produced by a set of 3 major and numerous minor salivary glands. No drastic manifestation occurs if there is an acquired hypo/malfunction of any one of these except for the local effects related to that particular gland e.g., pain, swelling, etc.[1-3] Congenital absence of any one of the major glands is usually asymptomatic per se but may come to the fore during evaluation of a relatively insignificant consequence it may lead to. It may, however, also present with features of xerostomia and its sequelae namely difficult in swallowing and dental caries. We report a rare case of unilateral submandibular gland aplasia presenting with a long-standing ipsilateral infra-auricular facial swelling due to compensatory secondary hypertrophy of the parotid gland, which was mistaken as recurrent parotitis. To the best of our knowledge, this is the first such reported case of ipsilateral submandibular gland aplasia detected during evaluation for suspicious parotidomegaly.

CASE REPORT

A 48-year-old female presented with complaint of right parotid swelling for last 8 months. There was no history of trauma, fever or significant local pain. The swelling was firm to touch, mildly tender, conforming to the contour of the underlying parotid gland with no overlying skin warmth, discoloration or sinus tract. A clinical suspicion of parotitis was entertained and she was referred for contrast enhanced computed tomography (CECT) of the neck. CECT revealed a diffusely bulky right parotid gland. There was no alteration of the glandular attenuation or any periglandular inflammation with maintained smooth outline of the parotid, which however seemed bulkier than its contralateral counterpart by about 30-40% (Figure 1). No ductal dilatation was seen. An incidental finding was non-visualization of right submandibular gland (Figure 1, 2). Few subcentimetric lymphnodes were seen in bilateral

level I locations, which however were not significant. No other significant finding was seen. Thereafter, the patient was questioned for any symptoms of dry mouth, dental caries or difficulty in swallowing, to which she responded in negative. Combining all the findings, it was deduced that the absence of right submandibular gland had probably caused compensatory hypertrophy of the ipsilateral parotid gland. On re-evaluation, no facial/mandibular/lacrimal/aural abnormality was seen on acquired CT scan. We, thus, were able to definitely rule out other causes of enlarged parotid namely nonmalignant (sialadenitis, infections, Sjögren syndrome, cysts and lymphadenopathy) and malignant disorders (lymphoma, adenoid cystic carcinoma and metastatic diseases). A biopsy was thus avoided.

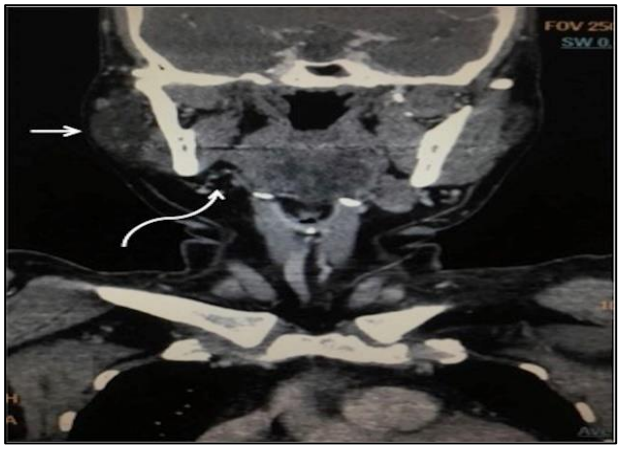

Figure 1: Coronal reformat of CECT of the neck depicts bulky right parotid gland (arrow) with smooth outline and normal attenuation. Note the absent right submandibular gland (curved arrow).

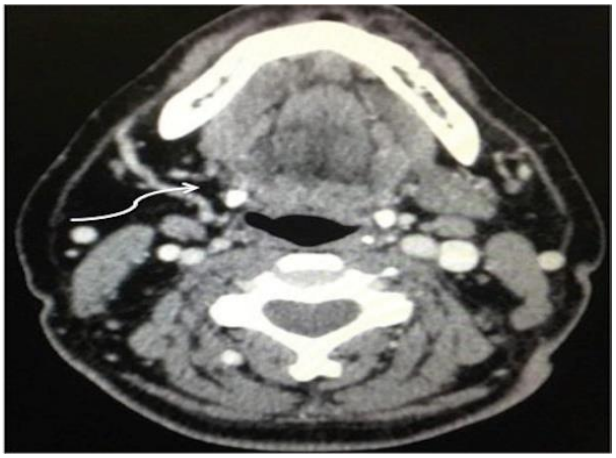

Figure 2: Axial CT of the neck reveals aplasia of the right submandibular gland (curved arrow).

DISCUSSION

During embryonic period, major salivary glands start developing around 6-8 weeks of gestation. All major salivary gland pairs are primarily derived from oral epithelium beginning as buds. It has been proposed that the parotid gland is ectodermal in origin, whereas the other two pairs of major salivary glands namely submandibular and sublingual glands are endodermal.[3] The primordial buds, which originate from the oral epithelium grow and branch into the adjoining mesenchymal tissue ultimately giving rise to the adult glandular ductal structure and acini and are associated with surrounding neurovascular development. Parotid is the first to be formed between the maxillary process and the mandibular arch. Subsequently the submandibular glands appear in the 6th week and complete their formation around 13-16 weeks arising from between the mandibular arch and tongue. The sublingual glands arise during the 9th week and form by early second trimester.[4]

'Aplasia', which is a Greek word, means congenital absence or defective development of tissue/organ. Isolated unilateral major salivary gland aplasia is a rare phenomenon with <100 case reports till date. The first case of a major salivary (submandibular) gland aplasia case was reported by Gruber in 1885.[4] The proposed mechanism is probably related to the disturbances of salivary organogenesis during fetal development of the first and second branchial arches. The exact etiology, though, still remains conjectural. It can be unilateral or bilateral, even involving more than one pair of glands. Unilateral hypoplasia is rarer still. It can be isolated or associated with other developmental anomalies related to the face such as hemifacial microsomia (craniofacial microsomia, first and second branchial arch syndrome, otomandibular dysostosis and lateral facial dysplasia), mandibulofacial dysostosis and lacrimo-auricular-dento-digital syndrome (mainly affects the eyes, ears, mouth, and hands).[5]

In previous case reports describing unilateral submandibular gland aplasia, few associated findings have been reported like sublingual gland hypertrophy (likely compensatory) and ectodermal dysplasis on imaging while symptom related findings include severe dental caries as well as other reduced salivary function such as xerostomia, oral infections and associated ectodermal dysplasia features.[3] The present case was unique as the submandibular gland aplasia was detected as an incidental finding during evaluation for parotidomegaly. It was, actually, the glandular aplasia, which had led on to compensatory hypertrophy of the parotid gland mimicking inflammation of the gland clinically. Till now, there have been reports of compensatory enlargement of either the contralateral submandibular gland or sublingual glands as compensation for reduced salivary production. The involvement of ipsilateral parotid gland has never been described. It is difficult to conclude the precise reason for involvement of the parotid and not the others but the mechanism seems definitely to make up for the reduced oral salivary production. Though we evaluated using a CT scan, there are other modalities to evaluate salivary glands namely USG, CT, MRI, sialography and scintigraphy (technetium T99m pertechnetate) studies.

CONCLUSION

We reiterate that, though very rare, suspicion of submandibular aplasia should be raised and the CT scan thoroughly studied during evaluation for parotidomegaly in a patient. This may be helpful to both the radiologist and the referring physician as it would prevent an unnecessary biopsy of the enlarged parotid and mitigate the scare arising out of the clinical findings of an infra-auriular swelling.

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