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*Case Report*

## **Non-Syndromic Congenital Bilateral Submandibular Gland Aplasia and Bilateral Parotid Gland Hypoplasia in Child Patient: Report of a Rare Case**

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### **ABSTRACT**

Congenital absence of one or more salivary gland(s) is a rare disorder. Such patient may present first to a dental surgeon with oral complaints of xerostomia, burning sensation, difficulty in eating and extensive dental demineralization resulting in rampant caries. This paper reports a case of non-syndromic non-familial congenital bilateral submandibular gland aplasia and bilateral parotid gland hypoplasia in a young male patient. Clinical presentation, diagnostic work-up and management of such patients are also discussed.

**Keywords:** xerostomia, agenesis, salivary gland, dental caries, aplasia, hypoplasia

### **INTRODUCTION**

The term aplasia and hypoplasia of salivary gland means total or partial absence of the gland respectively. Major salivary glands develop from the ectodermal lining of the stomodeum during 6<sup>th</sup> to 8<sup>th</sup> week of intrauterine life. Although the etiology of salivary gland aplasia or hypoplasia is not exactly known, it is thought to be due to defect during its fetal development.<sup>[1]</sup> It may be unilateral or bilateral and involve one or more of the salivary glands. Congenital absence of salivary gland(s) is a rare disorder. It can be associated with other ectodermal developmental abnormality and presents as part of a syndrome such as lacrimo-auriculo-dento-digital (LAAD)

syndrome, hemifacial microstomia, mandibulofacial-dysostosis (Treacher-Collins syndrome), and ectodermal dysplasia.<sup>[2,3]</sup> This paper reports a case of non-syndromic non-familial congenital bilateral submandibular gland aplasia and bilateral parotid gland hypoplasia in child patient.

### **CASE REPORT**

A 12 year old male patient was referred to our clinic with chief complaint of burning sensation in the mouth and difficulty to chew and swallow solid food. The complaint was present since childhood. The child could not take solid food and had to intermittently shift to liquid diet. His

medical and family history was unremarkable. The general health and intellectual development was normal. Otological and ophthalmology evaluation revealed no abnormality. Patient's skin appeared normal, with no abnormality of hair growth or sweating disorder. On extraoral examination, face was bilaterally symmetrical. On palpation submandibular gland bulk could not be appreciated. The lips of the patient were dry with crackling and angular chelitis (Figure 1a). Intra-oral examination showed dry oral mucosa with depapillated fissured tongue (figure 1b). The orifices of wharton's duct could not be visualized. The patient had mixed dentition with multiple carious teeth. sialometry showed flow rate of 0.05 ml/min. A differential diagnosis of salivary gland hypofunction, gland hypoplasia and agenesis was made. Ultrasonography (using 5-9 MHz linear probe) revealed bilateral small size parotid glands, showing coarse echopattern. No evidence of dilation of intra-glandular ductal system or calculi was seen. The submandibular glands were absent bilaterally. Based on the clinical and radiological findings diagnosis of bilateral parotid hypoplasia and bilateral submandibular gland agenesis was established.

**Table I:** Clinical presentation of major salivary gland aplasia/hypoplasia

<ul style="list-style-type: none"> <li>Dry mouth (xerostomia)</li> <li>Burning sensation with inability to take spicy food</li> <li>Area of erythematous changes over oral mucosa</li> <li>Dry scaly lips</li> <li>Angular chelitis</li> <li>Depalliated / fissured tongue</li> <li>Absence of salivary duct opening(s)</li> </ul>	<ul style="list-style-type: none"> <li>Opportunistic oral fungal infection e.g. candidiasis</li> <li>Extensive dental caries with loss of tooth structure</li> <li>Reduced salivary flow rate (on sialometry)</li> <li>Dry skin, lack of sweating, hair loss, dryness and burning of eye when present as part of syndrome like ectodermal dysplasia.</li> </ul>
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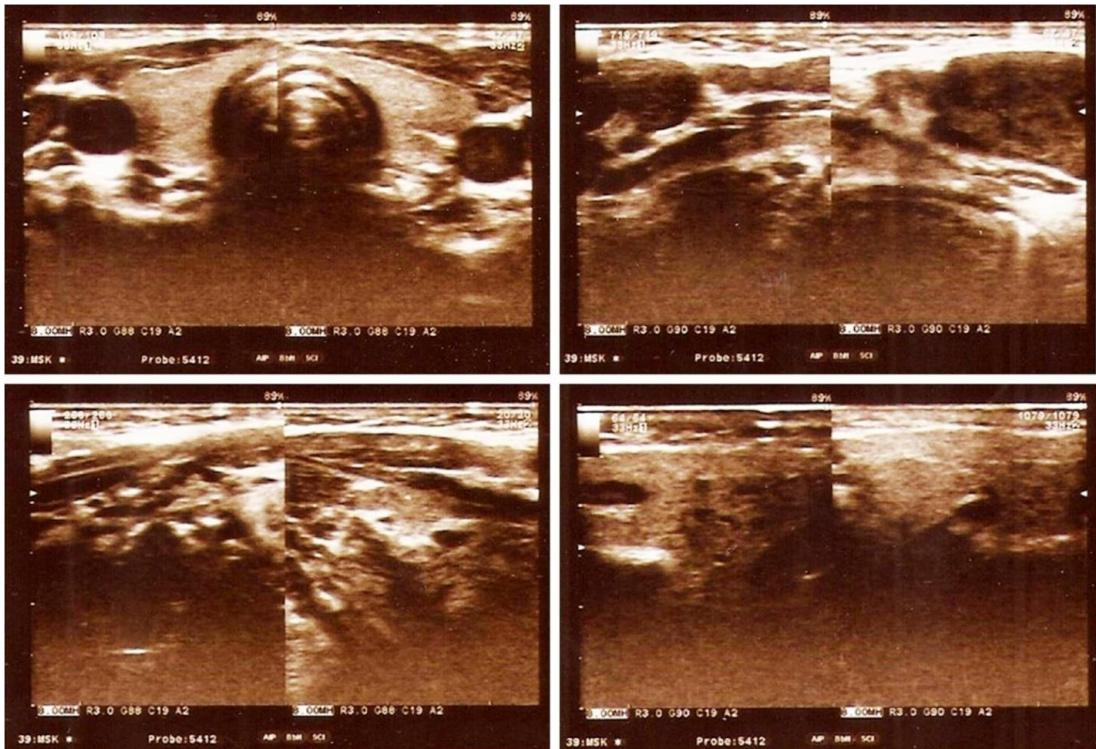
The management was symptomatic and included oral prophylaxis with counseling for oral hygiene maintenance. The decayed teeth were restored and topical fluoride application was done. Daily fluoride mouth rinses were advised. The patient was referred for dietary counseling to avoid cariogenic food. He was advised to take frequent sips of water during meal. Saliva substitute and sugar less gum was prescribed. The patient was put on regular dental checkups.



**Figure 1:** Dryness and crusting of lips with angular chelitis



**Figure 2:** Bald fissured tongue



**Figure 3:** Ultrasonogram showing absence of bilateral submandibular glands and hypoplasia of bilateral parotid glands.

## DISCUSSION

Aplasia of major salivary gland is extremely rare condition with an incidence of 1 in 5000 births.<sup>[3]</sup> It was first described by Gruber, in 1885.<sup>[4]</sup> Since then, 42 cases of salivary gland aplasia have been documented in English literature.<sup>[3]</sup> According to previous reports males appear to be more commonly affected than females (male: female ratio = 2:1).<sup>[5]</sup> Severity of its presentation depends on extent of involvement, which can be unilateral or bilateral, partial or total. Being a developmental disorder it usually manifests as xerostomia with associated sequel in child patient (Table 1). Therefore, pedodontists are commonly the first medical professionals they seek for consultation. Our case presented as burning sensation of

mouth with difficulty in taking solid diet in a young male patient.

The condition should be differentiated from other disorders causing xerostomia, like Sjögren's syndrome, drug-induced xerostomia, and ectodermal disorders (LAAD syndrome & ectodermal dysplasia).<sup>[3]</sup> Imaging of the salivary glands is essential for the final diagnosis of this condition. Ultrasonography, contrast enhanced computed tomography, magnetic resonance imaging, radionuclide imaging using technetium have been used to study the extent of involvement. In the present case ultrasonography revealed absence of bilateral submandibular glands with hypoplasia of bilateral parotid glands.

Management of patient with salivary gland aplasia is mainly symptomatic and is directed towards management of oral health

and maintenance of oral hygiene. Dental management includes oral prophylaxis, removal of non-restorable teeth and restoration of carious teeth. Further dental decay can be prevented by topical fluoride application regimen, fluoridated mouthwash and toothpaste. Diet and oral hygiene counseling is given to avoid cariogenic diet and prevent further dental decay. Residual salivary function can be improved by prescribing salivary stimulant/sialogogue (e.g. pilocarpine) and sugar-less chewing gum. Salivary substitute can be prescribed to moisten and lubricate the oral mucosa. Patients who develop secondary infection like candidiasis require antifungal therapy. The present case was managed by oral hygiene and diet maintenance. Restoration and topical fluoride application was done. Patient was advised frequent sips of water during meal. Long term follow-up was scheduled for evaluation of oral hygiene maintenance and prevention of dental decay. Salivary gland disorders may not be routinely seen by a dental surgeon. However patients with congenital salivary

gland aplasia/hypoplasia primarily presents with dental and oral manifestations of the disease. A dental surgeon needs to be aware of presentation, diagnostic work-up and oral care to be provided for this rare disorder.

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