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Major salivary gland agenesis in a young child: consequences for oral health

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Summary. *Background*. Salivary gland agenesis is a rare condition. It can be associated with some equally rare syndromes so that diagnosis can be delayed.

Case Report. The authors describe a case report. A 3-year-old girl, presented with dry mouth, carious teeth, recurrent herpes labialis and tonsillitis to Newcastle Dental Hospital. This case highlights the diagnostic and therapeutic challenges posed by 'salivary gland agenesis'.

Conclusion. Primary and secondary paedicitric healthcare professionals should be aware of the possibility of salivary gland agenesis in the setting of the 'non drooling baby'. Early detection of 'salivary gland agenesis' would do much to prevent the deleterious oral affects which follow the absence of salivary protection in the oral cavity.

Introduction

Salivary gland agenesis or aplasia is when the salivary glands fail to form. The problem may occur in association with other congenital defects such as lacrimo auriculo dento digital syndrome [1,2], hemifacial microstomia, and ectodermal dysplasia [3]. Diminished saliva flow (xerostomia), sore lips, and dysphagia are frequently the presenting features. The results of prolonged xerostomia can have a major impact on oral health including rampant caries, accelerated periodontal disease, candidiasis, and ascending sialadenitis [4]. The following case report illustrates how complications associated with reduced saliva flow can produce a clinical challenge.

Case report

A 3-year-old girl was referred by her dentist to Newcastle Dental Hospital. Clinical problems included dry mouth, multiple carious teeth, recurrent tonsillitis, and herpes labialis. The mother intimated the child had lip crusting on waking and an absence of drooling as a baby.

The child had attended the dentist since age 18 months and had been treated for what appeared to be nursing bottle caries, as a dietary history revealed the regular inclusion of fruit juice. The situation had been reported to the family doctor and health visitor but no action had been forthcoming although swallowing was becoming increasingly difficult.

A panoramic radiograph demonstrated gross caries (Fig. 1), and on examination xerostomia, and a thick saliva residue were noted. A preventive regimen including oral hygiene, dietary advice, and daily fluoride supplements was encouraged. Symptoms of dry mouth were initially managed with an artificial saliva spray. This was not well-tolerated and replaced with Biotene® products – which comprise a toothpaste, mouthwash, and saliva replacement gel (Anglian, UK). Biotene in conjunction with sipping water regularly helped the dysphagia. As a result of the caries, 14 primary teeth were extracted under general anaesthesia (GA).

Radionuclide imaging, using technetium (Tc 99m) sodium pertechnetate was undertaken and showed no gland function (Fig. 2a,b). Ultrasound examination confirmed the findings of total absence of normal salivary tissue (Fig. 3).

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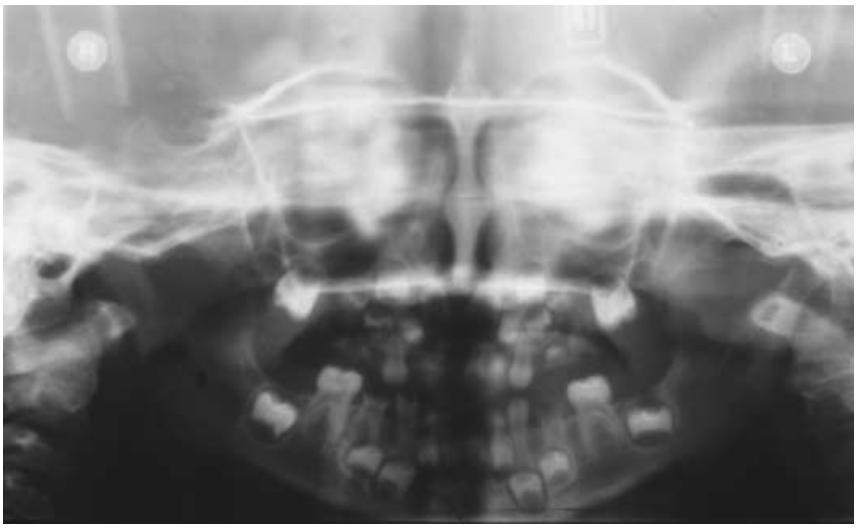


Fig. 1. Dental panoramic radiograph showing gross caries.

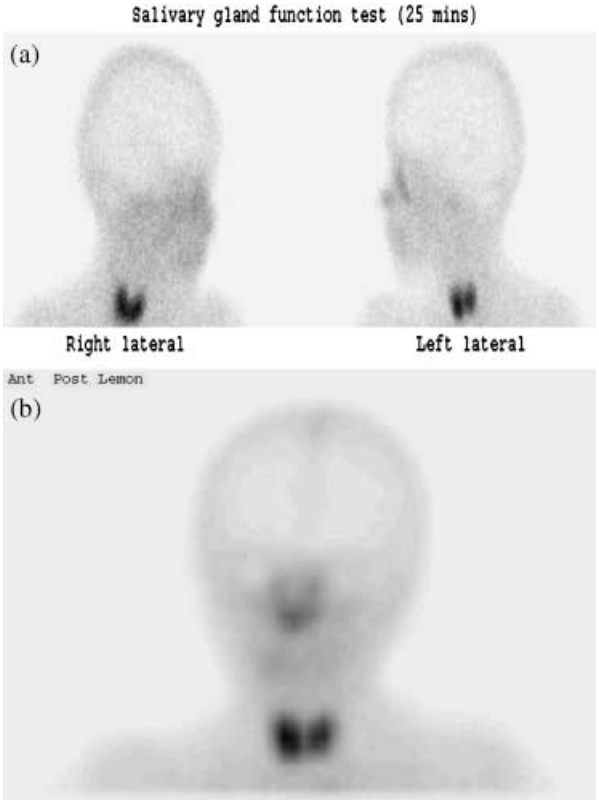


Fig. 2. (a,b) Tc 99m scintiscan showing absence of normal salivary tissue. 2a—Lateral view; 2b—Anterior view.

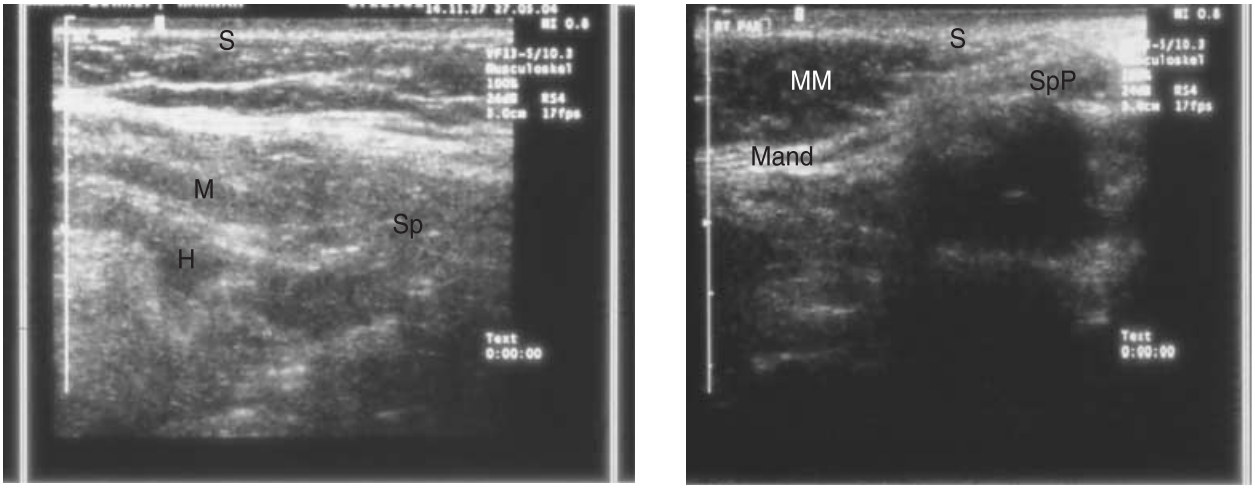
Discussion

Salivary gland agenesis is a rare disorder, which can appear singly or in combination with other genetic disorders. It can exhibit a complex pleotropic-autosomal dominant trait, or as is more likely in this case, as a new dominant anomaly secondary to a spontaneous mutation. The precise gene mutation involved in agenesis has not yet been identified.

Diagnosing salivary gland agenesis may not be straightforward as was demonstrated in this case. Some cases are detected early in childhood, however, later presentations especially in middle age may be confused with Sjögren's syndrome, which is more likely in this setting [5]. The child in this case had never experienced having saliva, and did not consider her situation as abnormal. Further potential for misdiagnosis may occur where a unilateral facial swelling actually represents the normal salivary gland in the absence of the contra-lateral gland. In the case presented, prolonged xerostomia led to a variety of opportunistic oral bacterial and viral infections, but being a rare anomaly, salivary agenesis may not feature on the initial differential diagnosis.

Imaging was fundamental in the detection of this anomaly. Investigations commonly associated with salivary gland disease involve plain radiography, sialography, ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI). The aims of imaging in this young girl had to maximize compliance, involve minimal use of ionizing radiation coupled with a need to delineate morphology and function. To this end, ultrasound and radionuclide imaging provided the optimum modalities.

Scintigraphy involves the injection of a radioactive (gamma emitting) tracer into the patient and



Longitudinal section, Lt submandibular area. Axial section, Rt parotid area.

Fig. 3. Ultrasound scan showing absence of salivary glands. H, hypoglossal muscle; M, mylohyoid muscle; Mand, mandibular ramus; MM, masseter muscle; S, skin surface; Sp, space where submandibular gland should be; SpP, space where parotid should be.

measuring its uptake in a gamma camera. The injected 'tracer' is technetium (Tc 99m) pertechnetate, which binds to the Na-K-Cl membrane transport system of the acinar cells of the salivary gland. The gamma camera measures the amount of radiation output from the tissue that directly correlates to the tissue uptake. Usually, the 'quantity' of emission is expressed as a change in colour, i.e., increasing red that equals a 'hot spot'. In this case, a grey scale is used so that the darker the shade, the more radiation this area is emitting and is incident upon the gamma camera.

With regards to salivary glands, uptake correlates with functional gland tissue and hence no uptake indicates absence of functional salivary tissue. Thyroid gland also takes up the technetium and gives a normal comparison to the salivary glands acinar cell membrane. If these cells are absent (because of gland agenesis) then there is nothing to bind to. Results from the scintigraphy demonstrated a normal uptake pattern in the thyroid gland. No such areas of uptake are seen in the parotid or submandibular gland areas on the scintiscan. This finding was corroborated on the ultrasound scan where no salivary tissue echo can be noted.

The absence of major salivary glands provides the clinician with an intriguing diagnostic challenge and the importance of recognizing this anomaly is obvious. It is within the dental speciality remit to have a working understanding and be aware of the differential diagnosis of dry mouth in the child patient.

The involvement of a multidisciplinary team with early referral to specialist dental services did ensure that a full preventive dental regimen was set in place at age 3 years to minimize the detrimental effects that a lack of saliva has on teeth. It should be noted, however, that glandosane is acidic and should not be considered as a first-line salivary substitute in a dentate patient.

A lesson to be learned from this case must be that of education of all members of the primary care team to the possibility that salivary gland agenesis (partial or total) can occur and should be considered in the differential diagnosis of the 'nondrooling baby'. Lifelong therapy utilizing salivary substitutes, topical fluoride, strict adherence to diet, and regular dental check-ups are essential to prevent the early loss of teeth and potential difficulty in coping with dentures.

What this case adds

- A 3-year-old girl presents with severe xerostomia, oral infections, and caries with an unusual underlying aetiology.

Why is this case report important to paediatric dentists

- Salivary gland agenesis is uncommon.
- Salivary gland agenesis can be imaged with a variety of imaging modalities that minimize radiation exposure to the child.

Acknowledgements

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