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Case Report

Asymptomatic unilateral aplasia of the left parotid gland: an unusual entity and case report ☆

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ABSTRACT

Congenital agenesis of major salivary glands is considered a very infrequent condition and typically appears to be a coincidental finding. It can be present as sporadic case or may be combined with aplasia or hypoplasia of other salivary glands or the lacrimal glands, or as a part of syndromes. Only 23 cases documented in the literature to date, while the youngest patient was 50 days old. Plenty of radiographic useful techniques and treatment is closely related to the clinical manifestations; therefore. We present a case of 52 year old female who was referred to our radiology department for recurrent numbness of the left upper limb, experienced over the previous three months. Complete absence of the left parotid gland was incidentally demonstrated at the brain MRI scan. Based on the patient's past medical history, physical examination and demonstrated radiographic techniques it was an asymptomatic, no-syndromic and no-familious unilateral aplasia of the parotid gland. In the present report, we aimed to underline that this rare condition may be asymptomatic and co-exist with other medical conditions and syndromes.

Introduction

Agenesis of the major salivary glands is a rare and misdiagnosed entity, generally combined with other congenital abnormalities of the first and second branchial arch, as well as craniofacial deformities [1,2]. This entity can be total or partial, unilateral or bilateral, symptomatic or asymptomatic, and with or without the presence of papilla of Stensen's duct [1]. Furthermore, agenesis of major salivary glands can be part of other syndromes [3]. To the best of our knowledge only 23 cases of unilateral parotid aplasia, have been documented in the literature [1,4]. Clinical presentation of unilateral parotid aplasia may include obvious signs of xerostomia and an increased rate of oropharyngeal infections [5]. The first imaging modality for evaluating the parotid region is a bilateral ultrasound; however, Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are more sensitive and equally as effective in demonstrating the pathology of

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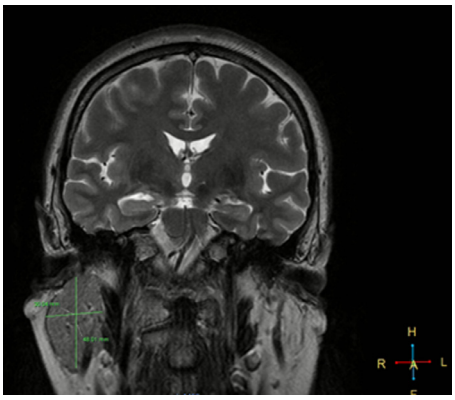

Fig .1 – Coronal T2 MRI revealed absence of left parotid gland and presence of right parotid gland, measuring 4.8 x 3cm, within the normal range.

major salivary glands [6]. Treatment of parotid gland aplasia is closely related to its clinical manifestations [5].

Herein, we present a rare case of a 52-years-old female patient with asymptomatic unilateral agenesis of the left parotid gland with bilateral presence of the Stensen's duct.

Case

52-year-old female was referred to our radiology department for recurrent numbness of the left upper limb, experienced over the previous 3 months. A brain and cervical MRI scan were performed, with no suspicious findings that could correlate with patients symptoms. However, complete absence of the left parotid gland was incidentally demonstrated at the brain MRI scan. The finding was confirmed by neck US examination. The rest of the salivary glands were depicted as normal and no other pathological findings were found according to the MRI (Fig. 1) and ultrasound findings (Fig. 2). Based on the patient's past medical history, it was an asymptomatic, nosyndromic and no-familial unilateral aplasia of the left parotid gland. Following a physical examination by the Otorhinolaryngology department (Fig. 3A), the bilateral hemifacial contour

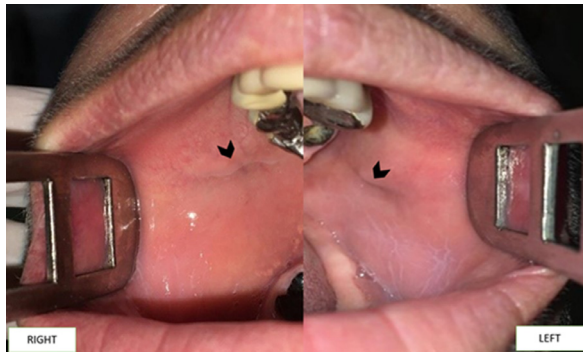

Fig . 3 – (A) The bilateral hemifacial contour was symmetric. (B) Bilateral presence of Stensen's duct (arrowheads) was noticed.

was symmetric. On palpation, the right parotid gland and the rest of the salivary glands were normal. Intraoral examination revealed a sufficient amount of saliva in the oral cavity, with bilateral presence of Stensen's duct (Fig. 3B). After stimulation, normal salivary flow through the right Stensen's duct was observed, while this could not be seen on the left side. We didn't proceed to another radiographic technique for ethical reasons, due to lack of symptoms in our case.

Discussion

Congenital agenesis of major salivary glands was first described by Gruber in 1885. The true incidence of cases involving one of the 2 parotid glands can be determined by taking into consideration not only symptomatic cases, but also asymptomatic ones [1,2]. Unilateral agenesis of the parotid gland typically appears to be a coincidental finding [1]. As far as the parotid gland is concerned, unilateral congenital absence of the parotid gland is considered a very infrequent condition, yet its actual incidence is difficult to ascertain since it is generally asymptomatic and may go unnoticed [5]. Parotid glands have an ectodermal origin; therefore, a disruption

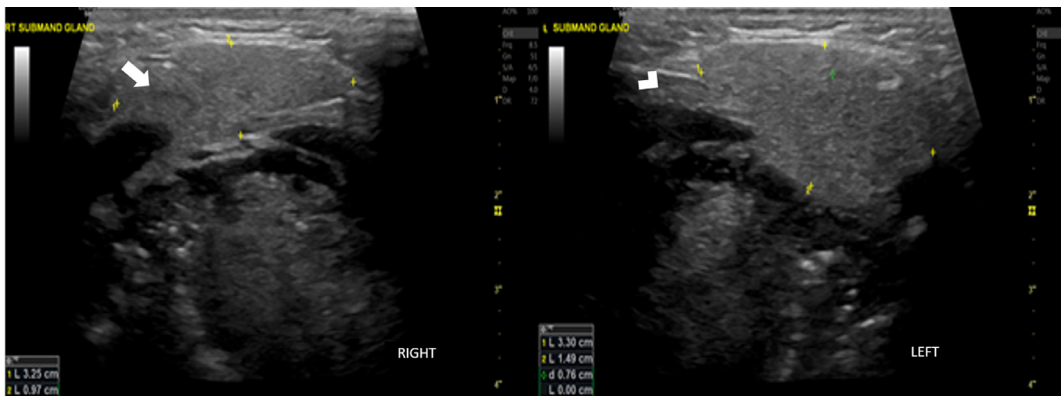

Fig . 2 – Neck ultrasound revealed normal appearance and size of right (thick white arrow) and left (thick white arrowhead) submandibular gland. (Color version of figure is available online)

during the 4th to 8th week of gestation may lead to parotid gland aplasia. This can be bilateral or unilateral, partial or total, and with or without the presence of an accessory gland [2]. Parotid gland aplasia may be a sporadic case or conversely, may be combined with aplasia or hypoplasia of other salivary glands or the lacrimal glands, as a part of an autosomal dominant disorder [2,3]. Moreover, parotid gland aplasia can be part of syndromes such as Down, Klinefelter and lacrimoauriculodentodigital (LADD) syndrome 1, or as a familial case [7,6,8]. This aplasia of the parotid gland is also combined with other congenital abnormalities of the first and second branchial arch, as well as craniofacial deformities [5,3,9,10].

A review of the literature conducted in 2016 identified a total of 22 cases of symptomatic and asymptomatic congenital unilateral aplasia of the parotid gland [1]. To the best of our knowledge, one additional case has been reported in the literature up to mid-2020. This case was a 4-year-old female with complete absence of the right parotid gland, hypoplasia of the parotid gland on the opposite side, and hypertrophy of the sublingual gland based on MRI findings [4].

Among the 22 cases documented in the review, 11 (50%) patients were male and 12 (50%) were female. At time of diagnosis, the youngest patient was 50 days old [8] and the oldest was 75 years of age [9], with an average age of 34.7 years [1]. The unilateral absence of the right parotid gland was almost twice as frequent as the left side (14/8 cases) [1,4]. The papilla of Stensen's duct was present in only 1 case from the cases that noticed it [1]. In the other cases, the parotid papilla was absent ($n = 12$), or the presence of the parotid papilla was not documented ($n = 9$) [1]. Hypertrophy of the contralateral parotid gland was found in approximately 6 cases (27.2%) and sialosis of the contralateral parotid gland was found in only 1 case (4.54%, 1). An accessory parotid gland was found in 4 cases (18.8%, 1). 2 cases with abnormalities of the mandible were reported (9.09%, 1), and 1case was found with simultaneous hypoplasia of the parotid gland on the opposite side, hypertrophy of the sublingual gland, presence of bifid tongue, supernumerary teeth on the alveolar border of the maxilla, a solitary mandibular incisor and a high-arched palate [4]. The remaining cases were asymptomatic or had findings irrelevant to parotid aplasia, such as metastatic neuroblastoma [1]. Association with other pathologies of the head and neck area, as well as the presence of an accessory parotid gland could not be identified in our case, while papilla of Stensen's duct was present.

Unilateral agenesis of the parotid gland may be clinically silent. Oftentimes, it may manifest as an absence of the papilla of Stensen's duct or as facial asymmetry, due to sialosis or hypertrophy of the contralateral parotid gland [10]. Nonetheless, depending on the presence or absence of adequate salivation from the residual glands, or in cases of bilateral absence of the parotid glands, the clinical presentation may include obvious signs of xerostomia, such as dental or periodontal diseases, dry and peeling lips, erythematous mucosa of the oral cavity and increased rate of oropharyngeal infections. Furthermore, in extreme circumstances, patients may be confronted with adversities in swallowing, chewing or even speaking, due to decreased salivary flow [5].

If the unilateral agenesis of the parotid gland is symptomatic, clinical examination could be the first step for diagnosis, particularly in the absence of the papilla of Stensen's duct. It should be noted that the Stensen's duct was present in only 1 case documented in literature, plus our case [1].

In asymptomatic cases the absence of parotid gland is discovered incidentally with ultrasound, CT or MRI and there is no need for further evaluation unless there are other suspicious findings.

In symptomatic cases there are plenty of radiographic useful techniques for parotid gland imaging. Plain Radiography is useful only to detect ductal calculi, calcifications, and adjacent osseous lesions [6]. A bilateral ultrasound is the first imaging of choice for evaluating the parotid region [6]. However, CT and MRI are more sensitive and equally effective for demonstrating the pathology of major salivary glands [6]. In these cases, it is important to rule out other associated conditions and syndromes for the most suitable management of patient [1]. In cases of sialadenitis unrelated to sialolithiasis sialography (conventional/fluoroscopy, CT or MRI) may be used [6]. Rarely used technique is Radionuclide imaging using Sodium pertechnetate (Tc[99]) and Positron emission tomography (PET) imaging using 2-deoxy-2-[18F] fluoro-d -glucose (FDG), based on actively concentrated and secreted by salivary gland cells, methods that can show also the presence or absence of the parotid tissue [1,6].

Treatment of the parotid gland aplasia is closely related to the clinical manifestations; therefore, asymptomatic unilateral gland agenesis requires no specific treatment [10]. However, therapy modalities of severe xerostomia and its sequelae include discipline in oral health maintenance and strict adherence to dietary rules. Additionally, the lifelong use of fluoride mouthwashes, saliva substitutes and sufficient liquid consumption is critically important to achieve constant hydration of the oral parotid cavity [5,10].

Conclusions

In conclusion, unilateral aplasia of the parotid gland may be symptomatic or asymptomatic, the only finding or co-exist with other congenital abnormalities.

Patient Consent Statement

Formal consents are not required for the use of entirely anonymised images from which the individual cannot be identified- for example, xrays, ultrasound images, pathology slides or laparoscopic images, provided that these do not contain any identifying marks and are not accompanied by text that might identify the individual concerned.

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