Case Report

Congenital Bilateral Ectopic Parotid Glands: Case Report with Literature Review

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Abstract

Congenital ectopic bilateral parotid glands are extremely rare, to date only two cases have been reported in the literature. Our patient, a 5-day-old male, presented with bilateral palpable cheek swelling. On imaging, the absence of bilateral parotid glands in parotid space and their ectopic location, anterior to the masseter muscle, was seen. Our case emphasizes ectopic parotids as an important differential among conditions presenting with bilateral cheek swelling in children. We have also compared the findings of previously described cases and their management with our case.

Keywords: Bilateral ectopic parotid glands, computed tomography, congenital, ectopic parotid, parotid aplasia, ultrasound

Introduction

Parotid glands are the largest salivary gland and are located behind the angle of the mandible and below the external auditory canal. They lie posterior to the masseter muscle and anterior to the mastoid and the sternocleidomastoid muscle. The parapharyngeal and carotid spaces are located medial to it.[1] Parotid gland is divided for surgical purposes into superficial and deep lobe based on the anatomical location of the retromandibular vein and facial nerve, which travel through the parotid parenchyma. Part of the gland located lateral to them is the superficial lobe and medial to them is the deep lobe. Anatomical variations of parotid glands are rare. Usual variants include accessory parotid tissue with or without aplasia of the main gland. Aplasia can be partial or total, unilateral or bilateral, and associated with several developmental syndromes. Other variants include the ectopic location of parotid which may be unilateral or bilateral.[2-4] Among the various variants, the ectopic location of parotid is important to diagnose as pure aplasia can result in failure of proper salivation leading to symptoms such as xerostomia, oral candidiasis, and dysphagia. Ectopic bilateral parotid glands are a very rare cause of bilateral cheek swelling and are usually incidentally detected. Other causes of bilateral palpable cheek swellings in a neonate include hypertrophy of the masseter muscles, diffuse inflammatory pathologies, infections like parotitis, and rarely neoplastic causes.[1] We present a case of an asymptomatic 5-day-old neonate with congenitally ectopic location of bilateral parotid glands with a review of the previously reported similar cases.

Case Report

A 5-day-old, term, a male baby was born by normal vaginal delivery and was clinically asymptomatic. The child received vaccinations at birth as per schedule. The patient was brought to the hospital by his parents when they noticed the presence of bilateral palpable cheek masses just posterior to the angle of the mouth. On examination, the swellings were firm on palpation and were not mobile in any direction. The baby did not cry on touching the swelling and was not irritable. The blood investigations done for the patient, hemogram, C-reactive protein, and erythrocyte sedimentation rate were all within the normal range. Bilateral cheek region ultrasound was requested by the clinician. Ultrasound [Figure 1] showed the absence of parotid glands at their eutopic location within

Received: 21-08-2022 **Revised:** 27-09-2022 **Accepted:** 17-11-2022 **Available Online:** 19-01-2023

Access this article online

Quick Response Code:

Website: https://journals.lww.com/jmut

DOI: 10.4103/jmu.jmu_85_22

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How to cite this article: Chawla S, Bothra M. Congenital bilateral ectopic parotid glands: Case report with literature review. J Med Ultrasound 2024;32:167-9.

the parotid fossa [Figure 1a and b] and bilateral symmetrically placed hyperechoic masses anterior and superficial to the masseter muscle [Figure 1c and d]. The child had a normal opening of the parotid ducts at the level of the upper second molar on oral examination with normal salivation. A diagnosis of bilateral ectopic parotid gland was suspected on USG. To confirm the findings, the patient was taken for computed tomography (CT) [Figure 2] of the head and neck which confirmed the findings of ultrasound by showing hypodense masses at a similar location and the absence of normal parotid glands. There were no associated bony facial anomalies. Since the child was asymptomatic, a decision to conservatively manage the patient was taken by the clinician. Parents were informed regarding this anatomical variation and benign nature of this condition. They were also counseled to notice the probable clinical symptoms in the child and the need for regular follow-up.

Discussion

Embryologically, parotid glands are the first salivary glands to develop between 4 and 8 weeks of gestation from epithelial bud of the floor of the primitive mouth which invaginates into the ectomesenchyme, proliferates, and forms the secretory parenchyma.[5] Any disruption in this development process can result in variant anatomy and morphology of the parotid glands, most of them being unilateral.[2,6] Most of the previously reported anatomical variations are described as the aberrant or heterotopic location of the parotid gland. Approximately 20% of the normal population have accessory parotid gland which is a nodule of normal glandular tissue, <3 cm and separate from the main gland. It is located superficial to the masseter muscle and is connected to Stensen's duct at that level.[1] Apart from this, heterotopic locations are reported in the literature at numerous locations such as the mandible, masseter muscle, hypophysis, lymph nodes, external and middle ear, thyroid gland, parathyroid gland, thyroglossal duct, lower neck, supraclavicular location, and rarely distant from the head and neck region.[2,7]

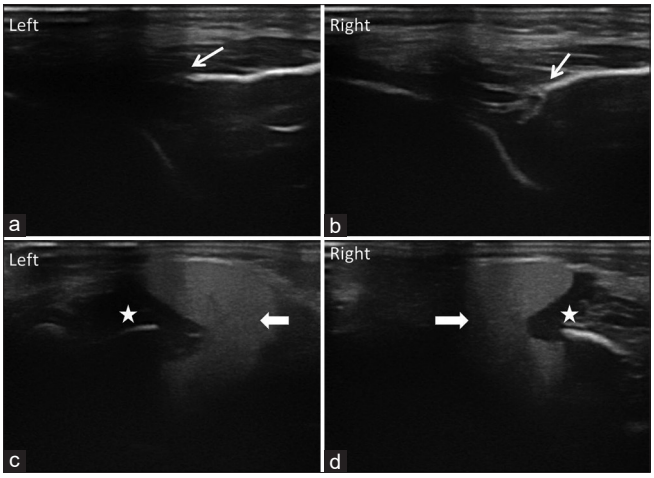

Figure 1: Coronal images anterior to the tragus shows the absence of bilateral parotid glands in bilateral parotid fossa posterior to the ramus of mandible (thin white arrows in figures a and b). Axial images done at the site of swelling shows hyperechoic ectopic parotid glands (thick arrow in figures c and d) anterior and superficial to the masseter muscle (white stars in figures c and d)

The complete absence of bilateral parotid glands at the eutopic location and the presence at ectopic location bilaterally is however very rare. Only two similar previous cases have been reported. The latest case was published in 2021 by Xie *et al*.[2] They performed a systematic review and found only one similar case in the literature which was published by Sun *et al*.[7] as a composite of case series with syndromic associations. Our case is the third in the series and findings of our case in comparison to previously reported cases are highlighted in Table 1.

Parotid gland aplasia refers to defective development or congenital absence of main parotid gland tissue. Few of these cases show hypertrophy of the accessory parotid tissue associated with aplasia of the main parotid glands, however, it's pure ectopia or a hyperplastic response of accessory parotid glands, is still not clear.[2,6,8] These cases can be differentiated from our case as accessory parotid glands are located lateral and superficial to the masseter muscle unlike the anterior location in relation to masseter muscle in our case. Deficient saliva production in patients with bilateral aplasia predisposes them to various oral sequelae such as dental caries, candidiasis, periodontal diseases, profound xerostomia, ascending sialadenitis, and halitosis. There is also an association with various developmental diseases such as first and second branchial clefts, mandibulofacial dysostosis, Klinefelter syndrome, hemifacial microsomia, and Down's syndrome.[2-4] Our case did not have any associated syndromic association.

Pathologies affecting the accessory parotid glands are an important differential in patients presenting with bilateral

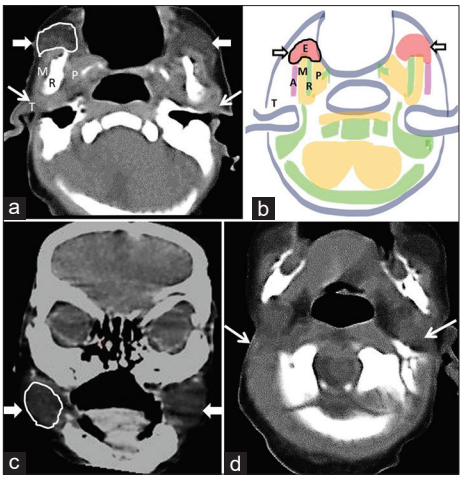

Figure 2: Axial and coronal sections of noncontrast CT scan show bilateral hypodense ectopic parotid glands (thick arrows and white line encased structures in figures a and c) anterior to the bilateral ramus of mandible, superficial to masseter muscle (M). Thin arrows in figures a and d show the absence of normal parotid gland in the parotid fossa bilaterally. Diagrammatic illustration of the axial noncontrast CT in figure b showing the anatomical landmarks and classical location of ectopic parotic glands (E) and accessory parotid glands (A). R: ramus of mandible; T: Tragus of the ear; P: pterygoid muscles. CT: Computed tomography

Table 1: Comparison of previously reported cases of bilateral ectopic parotid glands

Author and year	Age and gender	Clinical presentation	Imaging modalities	Ectopic location of parotid	Associated deformities	Management
Sun *et al*., 2013[7]	5 month old Male	Lateral facial cleft on the right side; underwent cheiloplasty at 5 months age. Recalled for examination at age of 2 years	CT-Bilateral absent parotid gland in parotid fossa with ectopic location of bilateral parotid glands	Below bilateral zygoma	Bilateral zygomatic arch slightly enlarged	No further treatment
Xie *et al*., 2021[2]	12 year old Female	Bilateral spontaneously developed buccal swelling in the upper masseteric regions for 8 years	CT-Left parotid gland was absent, right parotid gland was dysplastic with ectopic location of bilateral parotid glands	Superficial to ipsilateral masseter muscle	Asymmetry between bilateral angle of the mandible, ramus, and condylar process with the left side of the face being poorly developed	No further treatment. Regular follow-up mandated
2022 (index case)	5-day-old male	Bilateral cheek palpable masses	Ultrasound, CT-Bilateral absent parotid gland in parotid fossa with ectopic location of bilateral parotid glands	Anterior and superficial to masseter muscle	None	No further treatment. Regular follow-up mandated

CT: Computed tomography

cheek swelling with infections being the most common etiology.[2] Other lesions in the similar location which might be considered possible differentials in a neonate include congenital lesions such as venous or lymphatic malformations, masseteric muscle hypertrophy, diffuse inflammatory/infective diseases, and few other neoplastic lesions.

Since the physical examination is often inconclusive, imaging evaluation is done to confirm the diagnosis. Ultrasound is usually the first investigation performed followed by cross-sectional imaging with modalities such as CT and magnetic resonance imaging (MRI).[2,8] In our case, ultrasound showed the absence of parotid glands in bilateral parotid fossa with bilaterally symmetrical hyperechoic lesions anterior and superficial to the masseter muscle. It has the advantage of being radiation safe which is important in our case as the patient is neonate and is clinically asymptomatic. CT and MRI are reserved for complicated cases and in symptomatic or asymptomatic patients to look for associated craniofacial anomalies and syndromic associations.

Management is based on the severity of symptoms. If the patient has symptoms such as pain, fistula formation secondary to infection or cosmetic concerns, and surgical removal of the ectopic gland are done. In a patient with bilateral ectopic parotid glands, bilateral removal is not done for the risk of decreased salivary secretion. On the other hand, if the patient is asymptomatic like in our case, no further treatment is required. Regular follow-up with ultrasound imaging is usually performed to look for any subsequent pathology which might develop and its treatment.

Conclusion

We report an extremely rare case of congenital bilateral ectopic parotid glands in a newborn presenting as painless bilateral masses in cheeks without any associated bony deformities which have never been reported in previous cases. Our case highlights the importance of ultrasound as the first investigation to be performed in such cases and the role of cross-sectional imaging in addition to confirming the diagnosis is limited to the evaluation of complicated cases, ruling out other pathologies, and looking for associated syndromic associations. In asymptomatic patients, no treatment is required; however, frequent follow-up is mandated.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient's parents have given their consent for the images and other clinical information to be reported in the journal. The parents understand that the name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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