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Case Report

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Multiple hamartomas of the oral cavity with bifid tongue and cleft palate: A unique observation

Moaied A. Hassan

Department of Pediatric Surgery, Basrah Children's Specialty Hospital, Basrah, Iraq

Correspondence*: Moaied A. Hassan, Department of Pediatric Surgery, Basrah Children's Specialty Hospital, Basrah, Iraq. **E-mail**: moaied70@yahoo.com

KEYWORDS

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ABSTRACT

Background: Hamartomas are developmental tumor-like malformations, comprising a mixture of cytologically mature tissues that are indigenous to their anatomical location. Within the oral cavity, these lesions are usually multifocal, relatively small, and asymptomatic, and have been frequently linked to present as a manifestation of the oro-facial-digital-syndrome.

Case Presentation: A 2-day-old female newborn was referred with two masses protruding from the oral cavity, feeding difficulties, and dribbling of milk during feeding. The patient had no dysmorphic features and no obvious associated anomalies. On careful examination, there was one mass protruding from the undersurface of a bifid tongue with another larger mass that seemed to originate from the hard palate. Complete surgical excision of both masses and repair of the bifid tongue were done to relieve the baby's feeding difficulties. Histopathological examination of both specimens confirmed the diagnosis of multiple hamartomas.

Conclusion: The occurrence of multiple and large oral hamartomas in newborns might result in feeding difficulties that necessitate urgent surgical intervention. The association of lingual and palatal hamartomas, with bifid tongue and cleft palate, is a unique observation that is not reported in the English literature.

INTRODUCTION

Hamartomas are rare malformations that may develop anywhere in the body and consist of various tissue elements that are histologically consistent with those normally found in the anatomical location they develop in.[1] They develop most frequently in the lung, pancreas, spleen, liver, and kidney, with only a rare occurrence in the oral cavity.[1,2] Oral hamartomas can develop as isolated lesions, or in association with other systemic syndromes. [2,3] Multiple hamartomas that develop in the same patient are usually referred to as hamartomatosis or pleiotropic hamartomas. [1]

Herein, we present a unique association of bifid tongue, and cleft palate with lingual and palatal hamartomas in a newborn, describing its clinical presentation and surgical management.

CASE REPORT

A 2-day-old female newborn was referred to our institution with two masses protruding from the oral cavity, feeding difficulties, and dribbling of milk while feeding. The mother had an uneventful antenatal period, and there was no family history of previous birth defects.

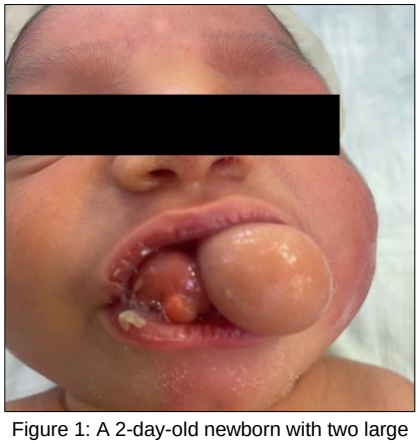

Figure 1: A 2-day-old newborn with two large masses protruding from the oral cavity.

On examination, there were no signs of respiratory distress. The patient had no dysmorphic features and

no obvious associated anomalies. There was one mass protruding from the undersurface of a bifid tongue with another larger mass that seemed to originate from the hard palate (Fig.1). The baby was admitted to the neonatal intensive care unit and stabilized. Echocardiography and abdominal ultrasonography excluded associated cardiac and renal anomalies. Complete surgical excision of both masses under general anesthesia along with repair of the bifid tongue was done to relieve the baby's feeding difficulties, and to avoid any possible intervention with respiration (Fig.2 & Fig.3).

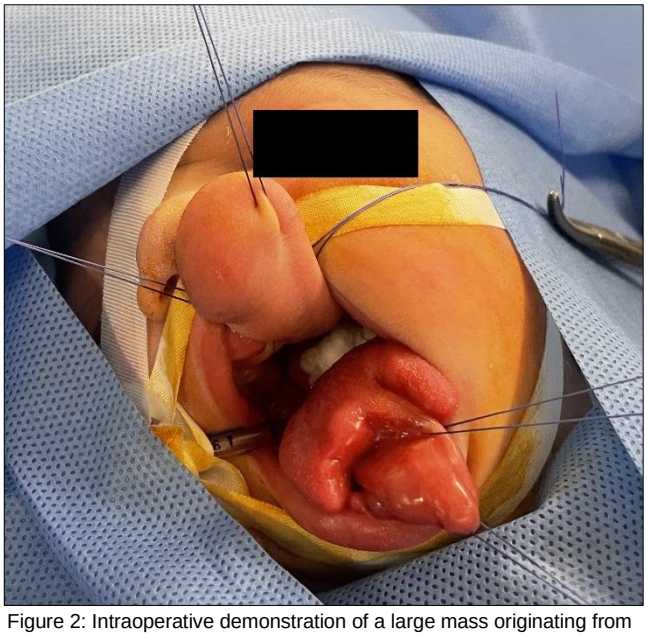

Figure 2: Intraoperative demonstration of a large mass originating from the palate, bifid tongue, and sublingual mass

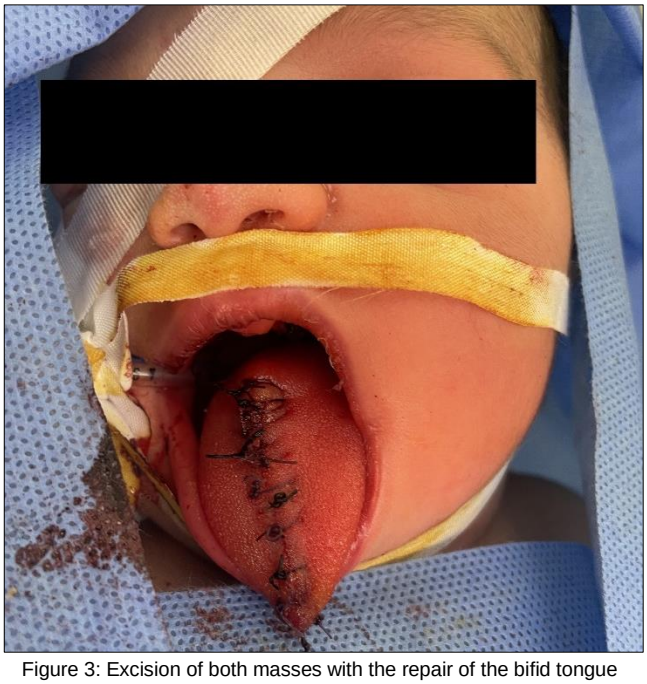

Figure 3: Excision of both masses with the repair of the bifid tongue

The palatine mass (4×3×2.5 cm) was pedunculated and its excision was straightforward. The sublingual mass (1.5×1.5×1 cm) was a bit more sessile; however, its excision was not difficult. After excision, it became obvious that the baby had a cleft palate. The patient had experienced uneventful postoperative recovery, tolerated feeding well on the first postoperative day, and was discharged on the third postoperative day. Histopathological examination of both specimens confirmed the diagnosis of multiple hamartomas. At 6 months of follow-up, the baby continues to have a normal feeding pattern and fair lingual appearance with no sign of recurrence.

DISCUSSION

The term hamartoma, which is derived from the Greek word "hamartia" was first coined by Albrecht in 1904 to describe developmental tumor-like malformations comprising a mixture of cytologically mature tissues that are indigenous to the anatomical location they are found in.[1,4]

Stamm and Tauber were the first to describe lingual hamartoma in 1945.[3] Hamartomas of the oral cavity are unusual and may show a wide spectrum of clinical presentations as well as histological characteristics and growth patterns.[5] According to the published literature, most hamartomas of the oral cavity are multifocal, relatively small, and asymptomatic, and have been frequently linked to present as a manifestation of the oro-facial-digital-syndrome.[2] Our patient presented with feeding difficulties at birth resulting from the multifocal, relatively large lesions that obstructed the oral cavity and thus necessitated urgent surgical intervention to obviate any possible airway problems. As a thorough evaluation did not reveal any associated defects, we consider the index patient to have developed the malformation sporadically. In their review, Kreiger et al. reported that the dorsal anterior tongue was the most common site affected [6,7], whereas Liu et al. reported that lingual hamartomas develop most frequently at the base of the tongue.[6,8]

Lingual hamartomas that are located in a posterior position, adjacent to the foramen cecum, in particular, should be evaluated thoroughly to exclude the possibility of being lingual thyroids, as excising them will render the patients hypothyroid.[8] The anterior position of the lingual hamartoma in the index case has obviated the need for further imaging or thyroid scans in the preoperative setting.

Takagi et al. reported that lingual hamartoma is the most common congenital tumor associated with the bifid tongue.[9] They proposed that a non-syndromic bifid tongue is secondary to the mechanical interference from the congenital tumor during the embryologic development of the tongue which will prevent its eventual fusion. Furthermore, failure of the embryologic precursors of the tongue to fuse will prevent proper morphogenesis of the palate resulting

in the formation of a cleft palate. This is attributed to the mechanical relationship between the two during embryologic development.[2,9] With regard to the patient presented in this study, we support the hypothesis of mechanical interference as the most likely etiology. The concomitant development of palatal hamartoma in our patient further supports the proposed hypothesis.

The patient was managed by surgical excision of both sublingual and palatal masses, along with repair of the bifid tongue, deferring repair of the cleft palate to another date. Histopathological examination of both specimens confirmed the diagnosis of hamartomas.

This is the only association of lingual and palatal hamartomas, bifid tongue, and cleft palate in the English literature.

To conclude, non-syndromic hamartomas of the oral cavity in newborns are extremely rare. The occurrence of multiple and large lesions might result in feeding difficulties that necessitate urgent surgical intervention.

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