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Case report

Persistent buccopharyngeal membrane: Report of a case and review of the literature

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ABSTRACT

A persistent buccopharyngeal membrane is a rare entity with 23 reported cases in the literature. Patients often present at birth in airway distress with diagnosis initially unrecognized. The otolaryngologist is frequently consulted in the delivery room to establish a secure airway after attempts at direct laryngoscopy are unsuccessful. Historically, definitive treatment has proved challenging. This article presents a new case and thoroughly reviews the literature for previous cases. Embryology, presentation, and treatment options are outlined.

1. Case report

A 6.5 pound female was born to a G2 P1 African American female whose pregnancy was complicated by polyhydramnios. The mother was induced for postdates at 40 4/7 weeks and underwent an uncomplicated vaginal delivery. The patient's Apgar score was eight at 1 and 5 min. Soon after birth the patient became cyanotic and bag mask ventilation failed to raise her blood oxygen saturation level. Multiple attempts at trans-oral intubation were unsuccessful, and the otolaryngologist performed a fiberoptic nasotracheal intubation.

Despite a secure airway, the patient's blood oxygen saturation level remained at 50%. An emergent echocardiogram showed transposition of the great arteries, an atrial septal defect, and a patent ductus arteriosus. The patient was then transferred to the Children's Hospital Los Angeles for tertiary care. Upon arrival, a balloon atrial septoplasty followed by an arterial switch was performed, successfully raising her blood oxygen saturation level.

Head and neck examination revealed a blind oral pouch with a membrane connecting the soft palate to the anterior tonsillar pillars and base of tongue (Fig. 1). The uvula was affixed to the anterior surface of the membrane. Attempts at direct laryngoscopy revealed no connection between the oral cavity and pharynx, with the membrane preventing visualization of the nasotracheal tube in the patient's oropharynx. The tongue was otherwise normal and there was no evidence of cleft palate.

Magnetic resonance imaging (MRI) was performed. Sagittal T1-weighted MRI image illustrates this complete membrane (Fig. 2). A CT was performed after contrast was placed into the patient's oral cavity (Fig. 3). Again shown is a blind oral pouch with no connection to the pharynx.

A diagnosis of persistent buccopharyngeal membrane (PBM) was made and a tracheotomy was performed. Transnasal fiberoptic examination of the oropharynx, hypopharynx, and larynx revealed normal anatomy.

The decision was made to defer definitive treatment of the PBM until the patient was 4 months old and had gained weight. A gastrostomy tube was placed and the patient was transferred to a skilled nursing facility. However, the patient became septic after transfer due to a presumed gastric laceration and passed away. An autopsy was not performed.

2. Embryology

In early development the primordial mouth, or stomodeum, is a depression in the surface ectoderm and separated from the primitive pharynx and foregut by the buccopharyngeal membrane [1]. This is a bilaminar membrane with ectoderm on one side and endoderm on the other. As the primitive face grows in three dimensions a stress is placed on this membrane causing it to break down, typically around day 26 [1,2]. The theoretical attachments of the membrane are superiorly to the soft palate, laterally along the anterior tonsillar pillars, and inferiorly at the junction of the anterior two-thirds and posterior one-third of the tongue [3]. Biopsies of the persistent buccopharyngeal membrane have shown

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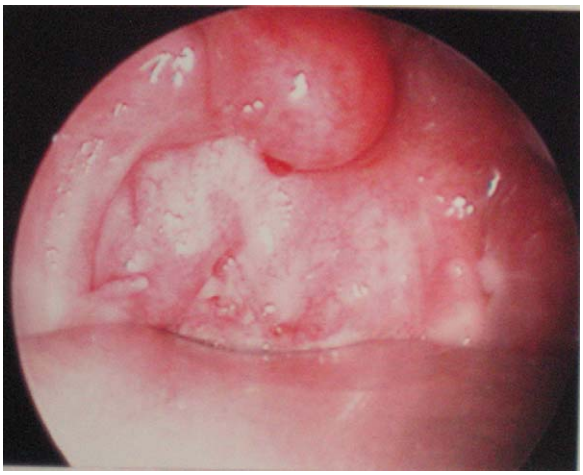

Fig. 1. Persistent buccopharyngeal membrane. Superiorly uvula is affixed to membrane. Right anterior tonsillar pillar is in view. Tongue is retracted inferiorly.

fat and muscle, with the theory that mesodermal penetration in utero of the membrane leads to its persistence [2].

3. Review of literature

A review of the literature revealed 23 reported cases, including ours (Table 1). Twelve of the patients were females and 11 males. All but four of the patients presented within the first month of life. Fourteen patients had a total membrane and nine presented with perforations in the membrane ranging from slit-like to 2 cm in size.

Eight patients, including ours, were born to mothers whose pregnancy was complicated by polyhydramnios [4,9,11,13–16]. Persistence of the buccopharyngeal membrane inhibits the fetus from swallowing amniotic fluid as in esophageal and duodenal atresia, all of which result in polyhydramnios. Seven patients, including ours, presented with cardiac anomalies which potentially complicate oxygenation issues until addressed [4,6,8,11,14,16]. A list of congenital anomalies associated with each patient is presented in Table 2.

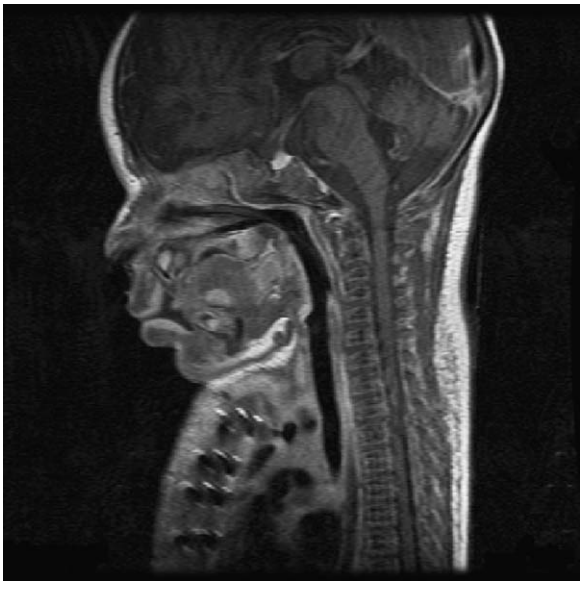

Fig. 2. Sagittal T1 weighted MRI demonstrates a complete buccopharyngeal membrane.

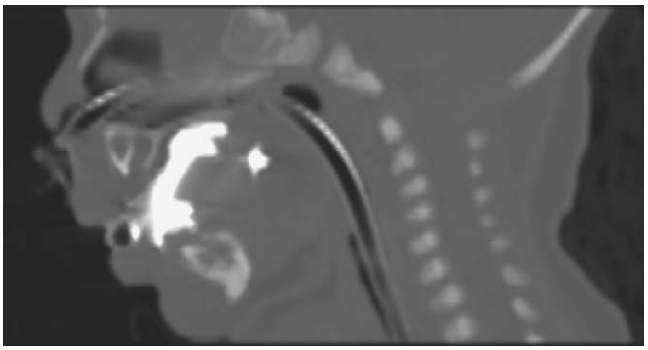

Fig. 3. Sagittal CT of the neck with oral contrast demonstrates no communication between oral cavity and oral pharynx. Patient is nasotracheally intubated.

The four patients reported with the condition hypomandibular faciocranial dysostosis had a PBM as a constellation of their symptoms [9,12,14,16]. This is thought to be an autosomal recessive condition, as two of the affected are siblings, and another patient was born to consanguineous parents [9,14]. Only one patient of four with this disorder has survived and is tracheotomy and gastrostomy tube dependent. In addition to the PBM the affected patients have anomalies including mandibular hypoplasia, hypoglossia, and a deficient midface.

A PBM presents in one of three scenarios. When a complete membrane, not allowing any communication between the mouth and the pharynx, the patient may suffer from early respiratory compromise. Attempts at direct laryngoscopy are unsuccessful with no normal anatomy visualized. Eleven of the 23 patients, or 48%, required establishment of a secure airway on the first day of life. Eight patients required an emergent tracheotomy, often in the delivery room. Two patients, including ours, were successfully intubated nasally over a fiberoptic bronchoscope and subsequently received a tracheotomy.

Eight patients were able to breathe easily and presented between 1 day and 3 months of age with dysphagia [2,8,18–21]. Physical examination of these patients demonstrated either a partial or total BPM. Five of these patients eventually required a tracheotomy while surgery was performed on the BPM.

Four patients presented with a buccopharyngeal membrane as an incidental finding [3,7,17,22]. These are the four oldest patients to present with this abnormality, aged 3 years to 27 years. All had a partial buccopharyngeal membrane, leaving a connection between the oral cavity and pharynx, permitting normal respiratory and swallowing function [3,7,22].

Definitive treatment of a complete PBM is challenging. Bent and co-authors recommend establishing an airway and feeding access as early as possible. They further suggest repair of the membrane be delayed until the patient is between 2 and 4 months of age. This allows the patient to mature but addresses the membrane early enough to prevent problems with swallowing and speech [11]. Experience has shown that a simple circumferential excision of this membrane is not adequate, almost always leading to stenosis and requiring further surgeries. Attempts at dilating these stenotic openings are equally as unsuccessful.

Longacre, in 1951, reported the successful repair of a persistent buccopharyngeal membrane in a patient who also had a cleft palate. Incisions were made inferiorly along the palatoglossus muscles to the level of the ariepiglottic fold. The palatoglossus muscles were then covered by rotated buccal mucosal flaps. This patient was subsequently able to tolerate oral intake and the tracheotomy tube was successfully removed [21].

After multiple attempts at a resection of a buccopharyngeal membrane with a combination of laser resections and pharyngeal

Table 1
Reported cases of a persistent buccopharyngeal membrane.

Author	Reference number	Year	Gender	Age at presentation	Membrane description	Establishment of airway at birth
Verma		2009	F	0	Total membrane	Nasotracheal intubation
Lee	[4]	2007	F	0	Slit perforation	Oral intubation
Kara	[3]	2007	F	18 years	2 cm central perf	None needed
Takahashi	[5]	2007	M	0	Total membrane	Tracheotomy
Tan	[6]	2006	M	0	Total membrane	Intubation
Ooi	[7]	2005	M	27 years	2 cm central perf	None needed
Lim	[8]	2005	M	1 day	Total membrane	None needed
Thauvin	[9]	2002	F	0	Total membrane	None needed
Kantaputra	[10]	2003	F	0	Total membrane	Tracheotomy
Bent	[11]	1997	F	0	5 mm perforation	Tracheotomy
Agarwal	[2]	1996	M	8 days	Total membrane	None needed
Agarwal	[2]	1996	M	13 days	Total membrane	None needed
Ludman	[12]	1993	F	0	Total membrane	Tracheotomy
Gartlan	[13]	1992	M	0	Pinpoint hole	Tracheotomy
Schimke	[14]	1991	F	0	Total membrane	Tracheotomy
Flannery	[15]	1989	F	0	Total membrane	Tracheotomy
Neidich	[16]	1988	F	0	Total membrane	Tracheotomy
Arcand	[17]	1988	M	3 years	Partial membrane	None needed
Hoffman	[18]	1979	M	1 month	Partial membrane	None needed
Chandra	[19]	1974	F	28 days	Total membrane	None needed
Seghers	[20]	1966	M	1 month	Total membrane	None needed
Longacre	[21]	1951	F	3 months	Partial membrane	None needed
Fridenberg	[22]	1908	M	21 years	Partial membrane	None needed

mucosal flaps, Bent and colleagues used an oropharyngeal stent to prevent restenosis of the buccopharyngeal membrane [11]. One of these two patients was able to have her tracheotomy tube removed and the other remained tracheotomy dependent secondary to micrognathia. Tan and co-authors created a stent initially fashioned from an endotracheal tube and later made of polyurethane [6]. After membrane excision, the stent was placed into the oropharynx and sutured to the cheek. The patient at last report was two and a half years old and able to babble while being tracheotomy and gastrostomy tube dependent.

Takahashi and co-authors described a PBM consisting of an anterior and posterior mucosal flap with an intervening muscle layer [5]. They made incisions similar to the spokes of a wheel to create six mucosal flaps in both the anterior and posterior mucosal layers which were alternatively folded and sutured in a zig-zag pattern. Post-operatively their patient had a patent connection between the oral cavity and oropharynx, was able to orally ingest, breathe, and vocalize, but was still tracheotomy tube dependent. Patients with a PBM have also been reported to have chronic eustachian tube dysfunction possibly requiring tympansotomy tubes [3,11].

Table 2
Congenital anomalies associated with a persistent buccopharyngeal membrane.

Author	Reference number	Associated congenital anomalies
Verma		Transposition of great arteries, patent ductus arteriosus, atrial septal defect
Lee	[4]	Ventricular septal defect, patent ductus arteriosus, vertebral anomalies
Kara	[3]	None
Takahashi	[5]	None
Tan	[6]	Atrial septal defect
Ooi	[7]	None
Lim	[8]	Atrial septal defect, unilateral choanal atresia
Thauvin	[9]	Hypomandibular faciocranial dysostosis
Kantaputra	[10]	Aglossia, micrognathia, microcephaly, absence of mandibular teeth
Bent	[11]	Transposition of great arteries
Agarwal	[2]	None
Agarwal	[2]	None
Ludman	[12]	Hypomandibular faciocranial dysostosis, bicornuate uterus
Gartlan	[13]	Micrognathia, microglossia
Schimke	[14]	Hypomandibular faciocranial dysostosis, patent ductus arteriosus
Flannery	[15]	Mandibulofacial dysostosis, downslanting palpebral fissures, epicanthal folds, broad nasal bridge, costovertebral anomalies
Neidich	[16]	Severe first branchial arch anomalies, atrial septal defect
Arcand	[17]	None
Hoffman	[18]	None
Chandra	[19]	None
Seghers	[20]	Auricular and vertebral anomalies
Longacre	[21]	None
Fridenberg	[22]	None

4. Conclusion

Persistent buccopharyngeal membrane is a rare entity with 23 reported cases. The otolaryngologist is often involved early with an emergent consult to the delivery room for a newborn with respiratory compromise and a difficult airway. Recognition of this lesion is useful as it allows for quick intervention through the multiple means of establishing an airway. Definitive treatment of the lesion is difficult as most patients are tracheotomy and gastrostomy tube dependent.

References

[1] K.L. Moore, T.V.N. Persuad, The Developing Human: Clinically Oriented Embryology, Saunders, 2003.
[2] R. Agarwal, P. Kumar, G.S. Kalra, S. Bhushan, R. Chandra, Persistent buccopharyngeal membrane: a report of two cases, Plast. Reconstr. Surg. 98 (October (5)) (1996) 866–868.
[3] C.O. Kara, I.G. Kara, Persistent buccopharyngeal membrane, Otolaryngol. Head Neck Surg. 136 (June (6)) (2007) 1021–1022.
[4] H.C. Lee, T.J. O-Lee, A. Madan, P. Koltai, Oropharyngeal atresia in a preterm infant: a case report and review of the literature, Int. J. Pediatr. Otorhinolaryngol. 71 (September (9)) (2007) 1485–1489.
[5] N. Takahashi, K. Kiyokawa, H. Rikimaru, K. Watanabe, Y. Inoue, Surgical treatment of pharyngeal atresia, J. Craniofac. Surg. 18 (January (1)) (2007) 208–211.
[6] S.S. Tan, H.K. Tan, C.S. Lim, W.M. Chiang, A novel stent for the treatment of persistent buccopharyngeal membrane, Int. J. Pediatr. Otorhinolaryngol. 70 (September (9)) (2006) 1645–1649.
[7] E.H. Ooi, Z. Khouri, M. Hilton, Persistent buccopharyngeal membrane in an adult, Int. J. Oral. Maxillofac. Surg. 34 (June (4)) (2005) 446–448.

[8] E.H. Lim, S. Tan, S.L. Lim, Anesthesia for a neonate with persistent buccopharyngeal membrane and unilateral choanal atresia, Paediatr. Anaesth. 15 (June (6)) (2005) 509–511.
[9] P. Kantaputra, P. Tanpaiboon, Thyroid dysfunction in a patient with aglossia, Am. J. Med. Genet. A 122 (October (3)) (2003) 274–277.
[10] C. Thauvin-Robinet, T. Rousseau, N. Laurent, C. Durand, C. Maingueneau, V. Cormier-Daire, et al., Hypomandibular faciocranial dysostosis in consanguineous parents revealed by ultrasound prenatal diagnosis, Prenat. Diagn. 22 (August (8)) (2002) 710–714.
[11] J.P. Bent III, F.N. Klippert, R.J. Smith, Management of congenital buccopharyngeal membrane, Cleft Palate Craniofac. J. 34 (November (6)) (1997) 538–541.
[12] M.D. Ludman, M.J. Vincer, C. Cron, M. Aguiar, M.M. Cohen Jr., Hypomandibular faciocranial dysostosis: another case and review, Am. J. Med. Genet. 47 (September (3)) (1993) 352–356.
[13] M.G. Gartlan, J. Davies, R.J. Smith, Congenital oral synechiae, Ann. Otol. Rhinol. Laryngol. 102 (March (3 Pt 1)) (1993) 186–197.
[14] R.N. Schimke, K.S. Claflin, J.H. Seguin, T.L. Bennett, B.E. Finley, L.M. Levitch, et al., Hypomandibular faciocranial dysostosis: report of an affected sib and follow-up, Am. J. Med. Genet. 41 (October (1)) (1991) 102–104.
[15] D.B. Flannery, Syndrome of imperforate oropharynx with costovertebral and auricular anomalies, Am. J. Med. Genet. 32 (February (2)) (1989) 189–191.
[16] J.A. Neidich, L.A. Whitaker, M. Natowicz, D.M. McDonald, R. Schnur, E.H. Zackai, Aglossia with congenital absence of the mandibular rami and other craniofacial abnormalities, Am. J. Med. Genet. Suppl. 4 (1988) 161–166.
[17] P. Arcand, J. Haikal, Persistent buccopharyngeal membrane, J. Otolaryngol. 17 (April (2)) (1988) 125–127.
[18] R.A. Hoffman, Persistent buccopharyngeal membrane, Arch. Otolaryngol. 105 (May) (1979) 286–287.
[19] R. Chandra, V.N. Yadava, R.N. Sharma, Persistent buccopharyngeal membrane. Case report, Plast. Reconstr. Surg. 54 (December (6)) (1974) 678–679.
[20] M.J. Seghers, Une malformation rare: l'imperforation oropharyngienne, Acta. Paediatr. Belg. 20 (2) (1966) 130–137.
[21] J.J. Longacre, Congenital atresia of the oropharynx, Plast. Reconstr. Surg. (1946) 8 (November (5)) (1951) 341–348.
[22] P. Fridenberg, Congential detachment of the faucial pillars and isolation of palatoglossus muscle, Laryngoscope 18 (7) (1908) 567–571.