Bilateral congenital oral mucous extravasation cysts

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Abstract

This report documents the bilateral presentation of oral mucous extravasation cysts on the left muccobuccal fold and right buccal mucosa of a neonate. The lesions were noted at birth and subsequently enlarged to the point that they interfered with eating. The left lesion ruptured but persisted as an exophytic fibrotic mass. Both lesions were surgically removed at eight months and the diagnosis was confirmed by histopathologic examination. Post-operative follow-up after nine months shows no recurrence. The presentation and diagnostic considerations are discussed. (Pediatr Dent 21:286-289, 1999)

Oral mucous extravasation cysts (MECs) can develop following disruption of minor salivary gland ducts and resultant extravasation of mucous secretions into the contiguous connective tissues. The ectopic mucous pool becomes delineated by a compressed zone of granulation tissue which, clinically, presents as a fluctuant or firm, often bluish, mucosal swelling. If the mucous accumulates immediately in the subepithelial region, it presents as a clear vesicle. Common analogous, but not specific, terms referring to the same pathologic process are mucous retention phenomenon or mucocele. MECs are distinct from the well-delineated, epithelium-lined mucous retention cysts, which occasionally develop from proximal expansion of a blocked duct. On a related note, the term "ranula" is used to describe a diffuse swelling in the floor of the mouth caused by either a MEC or, less commonly, a mucous retention cyst derived from the major sublingual or submandibular salivary glands. [1-4]

MECs can persist for highly variable periods ranging from days to weeks or even months. They are often characterized by sudden deflations as the mucous pool ruptures through the distended mucosa. The swellings recur if the duct connection to the surface is not re-established and the secretions continue to be directed into connective tissues. In an unknown fraction of cases, spontaneous resolution does not occur and surgical management is indicated. Although MECs occur most commonly in children and young adults, on rare occasions, these lesions have been reported in neonates and present an unusual differential diagnosis.[1,2,5,6] The differential diagnosis of congenital MEC primarily involves other exophytic congenital lesions. [7-9] This group includes the gingival cyst of the newborn (dental lamina or alveolar cyst), palatal cyst of the newborn (Epstein's pearls, Bohn's nodules), eruption cysts, congenital epulis, melanotic neuroectodermal tumor of infancy, and vascular hamartomas or neoplasms.

The gingival and palatal cysts of the newborn are common, easily identified, and self-resolving. They present as palatal nodules in 58%-64% of neonates and as gingival nodules in 11%-53% of neonates . Their putative origin is from embryonic epithelial rests, which differ according to site. Gingival cysts (origin: dental lamina) present on the alveolar ridges. Epstein's pearls (origin: epithelial remnants from palatal shelf fusion) present on the posterior mid-palatal raphe. Bohn's nodules (origin: embryonic glandular epithelial remnants) present at the junction of the hard and soft palate.[7-9] Additionally, eruption cysts occur on rare occasions in neonates. Jorgenson et al.[7] found such cysts in about 0.1% of healthy neonates. These lesions present as gingival swellings over erupting teeth caused by accumulation of fluid in the space between the tooth and follicle. The clinical presentation is characteristic, and again, the lesions are usually self-resolving with tooth eruption. The remaining lesions are unusual soft tissue masses, which could represent a greater challenge in diagnosis.

The congenital epulis is a rare smooth-surfaced soft mass, which occurs on the alveolar ridges of neonates. Large granular cells, possibly derived from primitive mesenchymal cells, comprise the lesion. It usually stops enlarging after birth and conservative excision is adequate treatment.[10, 11] A second rare soft tissue mass, which presents congenitally or in the first year, is the melanotic neuroectodermal tumor of infancy. This is a dark, locally destructive, lesion of neural crest origin, which most commonly presents in the anterior maxilla. It is comprised by a biphasic combination of larger melanin producing cells and smaller darkly staining neuroblastic-appearing cells. High urinary levels of vanillymandelic acid, consistent with a tumor of neural crest origin, are often found. Although simple excision is usually adequate, recurrence has been noted in about 15% of cases. Occasionally, there is even metastatic spread sometimes resulting in death.[12, 13]

The vascular hamartomas or neoplasms represent proliferations of lymphatic or blood vessels. Various subtypes have been defined based on clinical presentation as well as distinctive histologic features. The latter include vessel type and caliber, endothelial prominence, and architectural organization of the vascular components. Congenital oral hemangiomas can present as raised red/blue lesions, which blanch with diascopy. Congenital oral lymphangiomas can present superficially, most commonly on the tongue, as exophytic pebbled clear vesicular lesions whereas deeper lymphangiomas present as poorly defined masses without surface changes.[1,14] Of particular relevance is a form of lymphangioma called the alveolar lymphangioma.[15-17] The alveolar lymphangioma has been documented exclusively in about 4% of African-American neonates and occurs, often in multiple quadrants, on gingiva from the posterior maxillary

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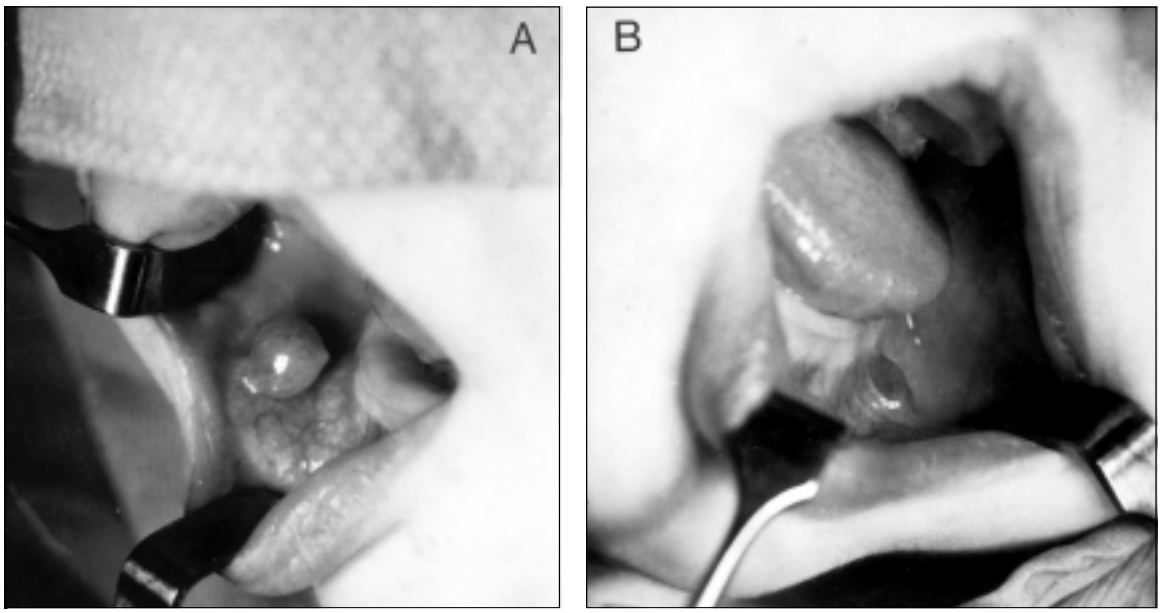

Fig 1A and 1B. Clinical photographs showing the right (A) and left (B) lesions arising respectively from the buccal mucosa and the muccobuccal fold. The left lesion had previously ruptured but has persisted as an elevated mass.

ridge or the mandibular posterior lingual alveolar ridge. It presents as a bluish fluid-filled dome-shaped swelling, which typically collapses on incision. Thus, the clinical presentation is suggestive of MEC and could represent an important differential diagnosis. Since the site of origin is gingiva, which does not contain mucous glands, lesion location is a significant consideration. However, before excluding the possibility of MEC, it would be important to confirm that the lesion has not extended from the contiguous posterior lateral hard palatal mucosa and soft palate or the lingual mandibular alveolar mucosa and retromolar pad area. Both of the latter regions do contain minor mucous glands.

This report documents and discusses a case in which a male neonate presented with bilateral exophytic lesions in the mandibular muccobuccal fold area. After consideration of the other previously noted lesions, these were considered clinically to represent congenital MECs. There was a family history of neurofibromatosis adding further complexity to the clinical interpretation since oral manifestations occur in up to 72%-92% of cases and congenital oral neurofibromas have been documented.[18-20] However, further clinical stigmata of neurofibromatosis were not found and in the absence of other anomalies, this possibility was not considered likely. After excision of the lesions, the clinical diagnosis of MEC was confirmed with histopathologic exam.

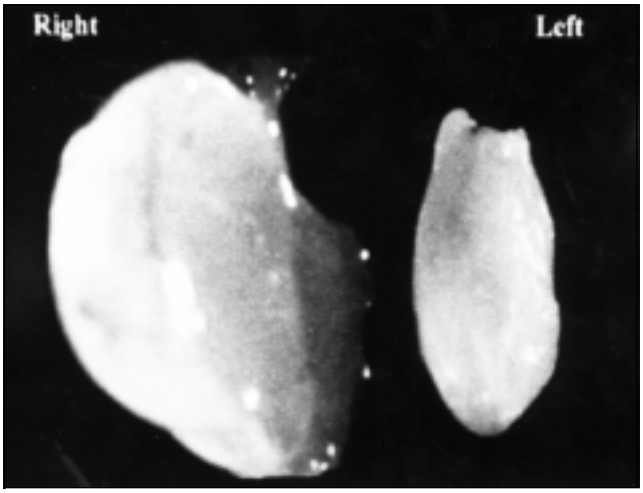

Fig 2. Surgical specimens. There were two soft smooth-surfaced masses measuring 9x8x6 mm (right) and 6x3x2 mm (left). The right lesion has been previously bisected and copious amounts of adherent mucous are evident.

Case report

Bilateral exophytic lesions in the mandibular muccobuccal fold/buccal mucosa region were noted at birth of an otherwise healthy caucasian male infant. The delivery was uneventful with no history of unusual manipulations in the mouth area. The lesions were compressible suggesting the possibility of MEC. Since there was a family history of neurofibromatosis and to investigate possible inheritance of the disease, the patient underwent two procedures involving general anesthesia at four and six months. These procedures were to permit magnetic resonance imaging and hearing testing. These investigations did not indicate any anomalies. However, at about eight months, the mucosal lesions had enlarged to the point that they interfered with eating. The left lesion had ruptured but persisted as a diminished exophytic mass. To manage this problem and to confirm the clinical diagnosis of MECs, the decision was made to excise both lesions. Excisional biopsies of both lesions (Fig 1) were performed at nine months under general anesthesia and the surgical specimens were submitted for histopathologic exam.

The surgical specimens are shown in Figure 2. Bisection of the right specimen showed it was filled with mucous. The left specimen was more fibrotic, but also showed a small cystic mucous filled center. Histopathologic exam (Fig 3) confirmed the clinical diagnosis. The thicker, more fibrotic wall and smaller lumen of the left specimen were interpreted to represent a reparative tissue response to the earlier rupture of this MEC.

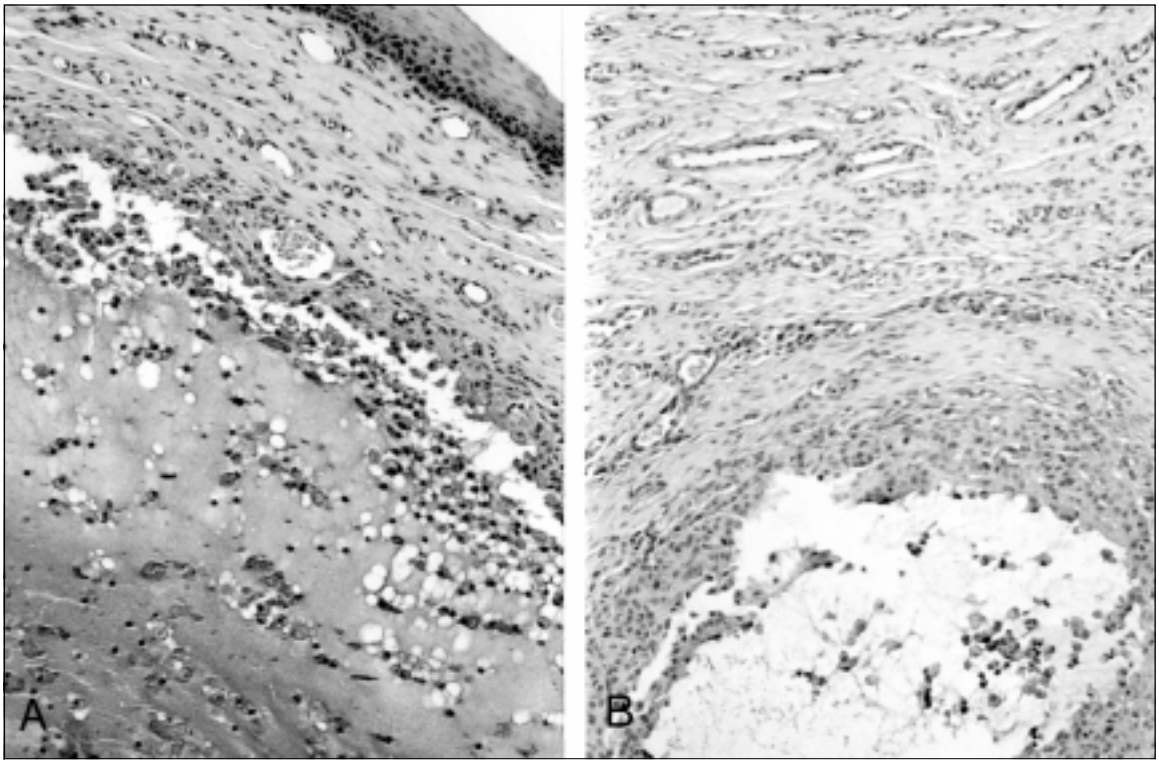

Fig 3A and 3B. Light photomicrographs of 5 um sections from the surgical specimens shown in Figure 2. Both specimens showed similar histologic features although the left lesion was more fibrotic. Figure 3A (right) shows a cyst formed by walls of granulation and chronically inflamed fibrous tissue. The lumen contains mucous which exhibits a mild infiltrate of acute and chronic inflammatory cells. The left specimen (Fig 3B) has a much smaller lumen with thicker fibrous walls. (Original magnification 150X; Southgate's Mucicarmine stain.)

Post-operative evaluation showed normal healing and follow-up after nine months shows no signs of recurrence.

Discussion

MECs occur most commonly on the lower lips and to a lesser extent the buccal mucosa, anterior ventral tongue, floor of mouth, retromolar regions, and posterior palatal mucosa. The age predilection to children and young adults is thought to be related to the greater propensity for facial injury and concomitant minor gland ductal injury in these age groups.[1,2,21] The basis for congenital MEC presentation is not clear but this pathogenic mechanism would suggest in utero oral mucosal injury. We speculate that it is possible that in utero thumb sucking might represent the precipitating factor in congenital cases.

Congenital MEC presentation appears unusual. In particular, we were unable to find other reports of bilateral congenital presentation. Finkelstein et al.[6] indicated in a case report and literature review of 1460 oral MEC cases that only 18 had been documented in newborns and infants up to one year of age. Only two of those cases, including their own, were clearly documented at birth. Subsequently, there has been at least one further report of a congenital MEC involving the inferior surface of tongue.[22] However, the incidence of congenital MECs is unknown since most MECs are relatively asymptomatic and an unknown fraction spontaneously resolve; these would not be readily noted. Thus, it is possible that this is a more common phenomenon than the literature indicates. Of possible relevance, Jorgenson et al.[7] in a study reporting oral lesions from 2164 healthy neonates, noted 15 with raised fluid-filled lesions of unknown diagnosis as well as a congenital ranula. The descriptions are clinically suggestive of MEC and might represent a 0.74% incidence.

Conclusion

The clinical diagnosis of bilateral congenital MECs in our case was suggested after considering multiple other congenital lesions with an exophytic component. After excisional biopsy, this was confirmed by histologic exam. The excised MECs did not contain gland tissue, a feature which sometimes is associated with a recurrence risk.[1,2] However, after nine months of follow-up, the involved areas continue to remain healthy and there is no evidence of further anomalies.

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Abstract of the Scientific Literature

Conversational Skills of Children with Cleft Lip and Palate

This study determined if there is a difference in measures of conversational participation between preschool and school age children with and without cleft lip and palate. Twenty children (ten preschoolers and ten school age) with a unilateral cleft lip and palate were compared to a matched group with no clefts. The children were evaluated for articulation, receptive and expressive language, and pragmatic skills. In addition, the examiner engaged the subjects in spontaneous conversation play. Results showed "no significant differences between the preschool and school age children with cleft lip and palate and their non cleft peers in level of conversational participation. However, individual child comparisons revealed less assertive profiles of conversation participation for 50% of the preschool and 20% of the school age children with cleft lip and palate."

Comments: Craniofacial teams may be able to improve the quality of socialization for the child with a cleft lip and palate who has difficulty with conversational skills. JEP

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