Alveolar lymphangioma diagnosed in a white Irish neonate: a previously unreported finding

Précis

A case of alveolar lymphangioma in a white Irish neonate is presented. The relevant background, clinical presentation, diagnosis and management are discussed.

Abstract

Historically, alveolar lymphangiomas have been reported exclusively in the oral cavities of black infants. To the author's knowledge this is the first report of alveolar lymphangioma in a white Irish neonate. The paper presents multiple alveolar lymphangiomas found in the oral cavity of a white Irish neonate in a Dublin children's hospital. The child's medical background, differential diagnosis, management options and outcome are discussed.

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Introduction

Alveolar lymphangiomas are rare, benign soft tissue conditions of the oral cavity. First described by Levin in the 1970s, the lesion has exhibited a strong racial predilection, with an incidence of 2.2-4% reported in black neonates and no cases reported in white neonates.[1,2,3] Increasing immigration into Ireland may result in infants with this condition presenting to the general dentist. A case series of this condition in two healthy black neonates was published in this journal in 2009.[4]

Case report

History

A 15-day-old white Irish female neonate was referred from her paediatric neonatology team to the dental department of a tertiary care children's hospital regarding bilateral cystic swellings in the upper and lower jaw. She was born at term 37+4 weeks by spontaneous vaginal delivery. Maternal history was that of a healthy 40-year-old white Irish woman with a medical history of psoriasis and cholecystectomy. Paternal background was that of a white Irish male. The infant was noted to have dysmorphic features at birth, including frontal bossing, depressed nasal bridge and overlapping digits. A cleft of the soft palate was diagnosed by the neonatal team. From a respiratory perspective, the infant was diagnosed with obstructive sleep apnoea and nocturnal continuous positive airway pressure (CPAP) was required. As she was unable to feed orally, a nasogastric tube was inserted. There was a history of tonic clonic seizures at birth, which resolved. There was both central and peripheral hypotonia. Genetic testing detected a chromosomal imbalance including a gain on Xp22.33-Xp22.11 and a 14q deletion.

Clinical presentation

The patient's mother first noticed the oral lesions around the time of birth. No change was observed by her in the size, shape or location from birth to time of assessment at just over two weeks of life. Clinical examination revealed bilateral, soft, fluid-filled swellings at all first primary molar sites along the crest of the maxillary and mandibular ridges. All lesions were dome shaped, with the upper right lesion mobile. Fluid within the lesions had a clear yellow, straw-like colour. A cleft of the soft palate was also noted (**Figure 1**).

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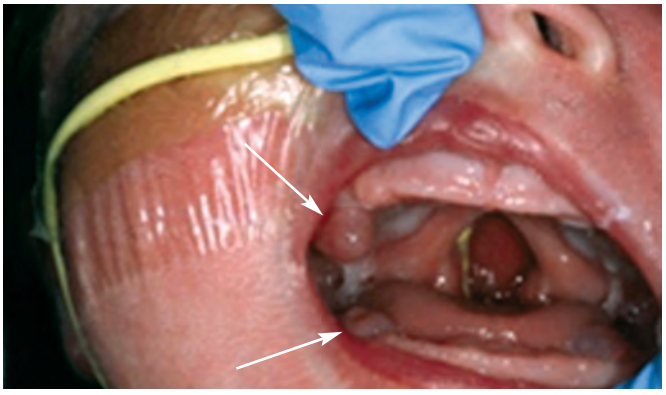

FIGURE 1: Photograph of right maxillary and mandibular ridge at initial presentation (15 days old) showing lesions on the central aspect of the alveolar ridge. A cleft of the soft palate is also apparent.

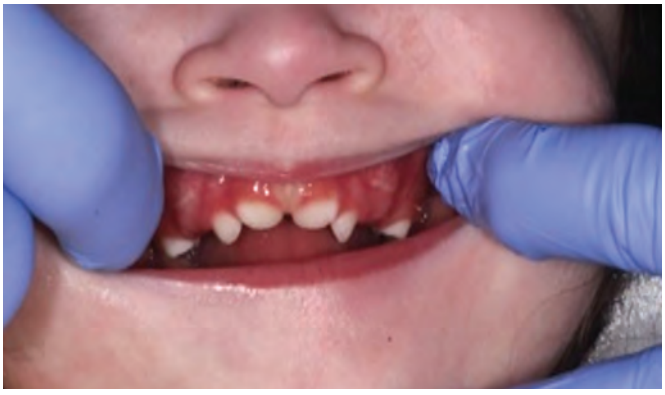

FIGURE 2: Review at 16 months showing resolution of maxillary right and left lesions, and eruption of maxillary first primary molars and incisors.

Table 1: Differential diagnosis considerations for alveolar lymphangioma.

Differential diagnosis	Clinical appearance/ characteristics	Location	Distribution
1. Eruption cyst	Bluish, fluctuant, dome-shaped swelling Crowns of teeth under lesions	Mandibular incisor region but can occur at any site of an erupting tooth	Unilateral, may cross midline
2. Extravasation mucocoele	Bluish, fluctuant, dome-shaped swelling	Lip, floor of mouth	Unilateral
3. Congenital epulis of newborn	Pink, firm, lobulated mass	Anterior maxillary gingiva	Unilateral

Table 2: Clinical criteria for diagnosis of alveolar lymphangioma.

Clinical diagnostic criteria	Description
1. Colour/shape/surface	Blue/dome-shaped/fluctuant
2. Site and number	Alveolar ridge, multiple
3. Distribution	Bilateral pattern
4. Size	Average 3-4mm diameter
5. Content	Fluid (clear/yellow)

Diagnosis

The clinical appearance excluded lesions of an inflammatory or infective origin, and the swellings were deemed to be developmental in nature (**Table 1**). A clinical diagnosis of alveolar lymphangioma was made (**Table 2**) and agreed by two consultant paediatric dental surgeons.

Management

A conservative approach was adopted, with clinical observation and photographic recording to allow for comparison at each review. Consideration was given to other treatment modalities including aspiration/drainage and excision, but these were deemed unsuitable at such an early stage on a background of a complex neonate. The patient was reviewed periodically. At one-week review there was a significant reduction in the size of the lesions at all sites but especially in the upper arch. The following review two weeks later showed further reduction in lesion size, with an epithelialisation of the outer layer apparent. The review period was extended and one month later there was almost complete resolution, with the lower left lesion remaining most prominent. The patient continued to be reviewed regularly and at six months there was a complete resolution of all lesions. Follow-up continued to monitor dental development, and at the recall at 16 months, a normally developing oral cavity was noted (**Figures 2** and **3**).

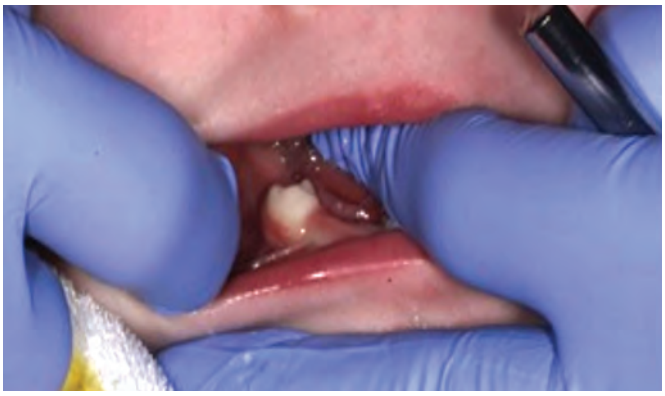

FIGURE 3: Review at 16 months showing mandibular first primary molar erupted and full resolution of lesion.

Discussion

Although some authors recommend excisional biopsy, these lesions are reported to spontaneously involute with time, and therefore no further investigations were undertaken.[2,3] Alveolar lymphangiomas have not been reported in the white population. In the black population there is a 2:1 male-to-female distribution.[2] Clinical presentation is similar to a mucocoele or an eruption cyst characterised by a bluish colour and domed, fluid-filled

appearance. They are characteristically located at the first primary molar site of the alveolar ridge and favour a bilateral distribution.[3] Those affected typically have multiple lesions, with the mandibular alveolar ridge a favoured site.[3] Aetiology is unknown; however, the strong racial predilection and site specificity are suggestive of a developmental, possibly genetic derivation.[3] Surgical management is generally unnecessary, but has been suggested where there are feeding difficulties.[5] Reported biopsies of such lesions have resulted in their collapse and release of a clear fluid.[2] Microscopic examination shows lymphocytic structures arranged in an alveolar pattern, supported by connective tissue.[6] Reassurance to parents is important, with monitoring necessary over time as regression may take several months.[2,3]

This case is an example of a documented lesion in an unreported population. Genetics may play a role, with the 14q deletion associated with distinctive facial features including wide-set eyes and microretrognathia.[7] Reported 14q oral features include a narrow arch and a high palate.[8] Genetic testing from this case reported that this oral finding had yet to be linked with the identified chromosomal deletion. The patient's medical background was complex with acute respiratory difficulties at time of initial referral, and therefore a conservative, non-surgical method was deemed most appropriate. A multidisciplinary approach with prioritisation of care was adopted. Involved teams included respiratory, ophthalmology, plastics, dental and dietetics. As with any patient with complex medical challenges, acute issues were first addressed, with stabilisation of her respiratory condition a priority. Close liaison with the medical team was vital to confirm the ongoing appropriateness of the conservative approach. Communication and reassurance to parents regarding the rationale for conservative oral management was explained. All lesions spontaneously regressed with no intervention needed.

Conclusion

This case represents a unique finding of multiple alveolar lymphangiomas in a white neonate. As with other case reports, the lesions' clinical features were sufficiently characteristic to permit an accurate clinical diagnosis without having to resort to invasive investigations. Although rare, such a lesion may present to general dentists in Ireland, necessitating a clear understanding of the condition's presentation and management.

References

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CPD

1. What is the reported prevalence of alveolar lymphangioma?

- ○ A: 2-4%
- ○ B: 10%
- ○ C: 20%

2. What strong ethnic predilection does alveolar lymphangioma exhibit?

- ○ A: Caucasian
- ○ B: African
- ○ C: Asian

3. How does this condition typically behave?

- ○ A: Spontaneous involution and resolution with time
- ○ B: Increase in size with local invasion
- ○ C: Recurrence following removal