Congenital Lesions of Epithelial Origin

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KEYWORDS

• Midline cervical cleft • Dermoid cyst • Pilomatrixoma • Foregut duplication
• Preauricular pit dermoid

KEY POINTS

- Congenital lesions of the epithelium have varied presentations, including clefts, cysts, sinuses and pits, and masses.
- Management ranges from simple observation for asymptomatic lesions to surgical excision with complex repair.
- Nasal dermoids may take a multidisciplinary approach with neurosurgical involvement for intracranial extension.
- Syndromes associated with these lesions must also be considered in the evaluation of the pediatric patient with these defects.

INTRODUCTION

Most masses of the head and neck in children are benign in nature. Most commonly, these lesions are congenital, inflammatory, or infectious in cause. Defects of epithelial origin are the result of failure of fusion during embryologic development, failure of involution of primordial remnants during embryologic development, and defects of ectodermal development during childhood.

CONGENITAL MIDLINE CERVICAL CLEFT

Epidemiology

Congenital midline cervical cleft (CMCC) is a rare developmental defect of the anterior neck with fewer than 100 cases reported in the literature.[1] These defects account for less than 2% of congenital cervical malformations.[2] There is some predilection for caucasian female children, and it can occur as a concomitant lesion with thyroglossal duct cysts and branchial cleft anomalies with an incidence of 1.7% in this population.[3]

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No familial inheritance pattern has been identified, and they are thought to be a sporadic defect.[2]

Cause/Embryology

Although the exact cause of CMCC is unknown, it is generally thought to be the result of failure of midline fusion of the branchial arches.[4] In normal embryologic development, during the third and fourth week of gestation, mesenchymal tissue migrates between the branchial arches, pushing the ectoderm outward and resulting in fusion of the arches. A disruption in this process results in a midline defect. Failure of fusion of the second (hyoid) arches results in CMCC, whereas defects of the first (mandibular) arches result in clefts of the lower lip, tongue, and mandible, as well as CMCC.

Presentation

CMCCs are present at birth, although they become more apparent over time. They may be overlooked or misdiagnosed early on.[5] These lesions may present at any point in the anterior midline neck from the mandible to the sternum.[1] There are 3 primary anatomic components in these lesions: a superior skin tag, a midline cleft of moist atrophic epidermis lacking adnexal structures, and a caudal sinus or duct at the inferior aspect of the cleft, which may drain mucous.[1] The senior author has operated on many of these lesions, and in his experience, the skin tag associated with this lesion may present at the superior or inferior aspect of the defect. It is also noted that CMCCs are associated with a subcutaneous amorphous fibrous cord that traverses the sternum to the midline of the mandible. This cord as well as a paucity of dermis contributes to a contracture that results in a wry neck. In the senior author's experience, there is nearly always a subcutaneous cord along the tract of the cleft, which may extend as high as the mandible and as low as the sternum. This cord is the result of maldevelopment of the median raphe of the strap muscles.[5] CMCC may be a solitary deformity or may be accompanied by other lesions, such as thyroglossal duct cyst, branchial cleft cyst, ectopic bronchogenic cyst, cleft lip, mandible, or tongue, absence of hyoid bone or thyroid cartilage, cleft sternum, or congenital heart disease.[1]

With time, the cleft heals with a scar, resulting in a web.[1] The web may ultimately result in neck contracture, limited mobility, and functional impairment, or torticollis. In more severe cases, this webbing can result in micrognathia, or a bony spur of the mandible or sternum (**Fig. 1**).

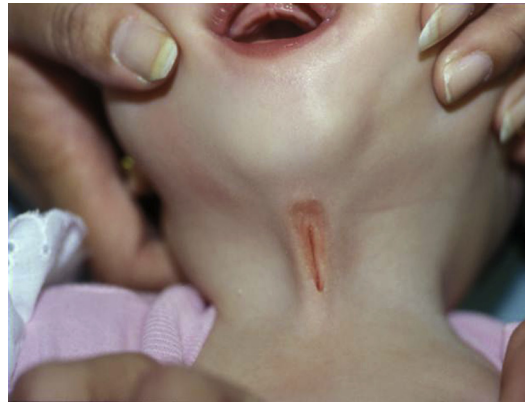

Fig. 1. Example of CMCC without an associated skin tag. The subcutaneous cord and associated contracture are evident.