SECOND BRANCHIAL CLEFT ANOMALY PRESENTING AS A RUDIMENTARY PINNA IN THE NASOPHARYNX OF A NEWBORN

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Abstract
We describe the case of a 4-day-old girl who presented with an epiglottic cyst that was later identified as a rudimentary pinna attached to the soft palate.

Introduction
The branchial apparatus develops during the third to seventh weeks of intrauterine life. The ectodermal side of each invagination is known as the branchial cleft. While normal development of the second branchial cleft entails the formation of the skin of the anterior triangle of the neck, deviations from this norm may occur anywhere from the anterior triangle to the tonsillar fossa.

The pinna itself normally develops from six aural hillocks that are derived from both the first and second arches. It was once thought that the pinna receives equal contributions from both arches, but it is now believed that the first arch gives rise to the tragus and that the remainder of the pinna is formed from the second arch. The fact that the muscles attached to the pinna are all innervated by the facial nerve, which originates from the second branchial arch, further attests to the dominant role that this arch plays in the development of the external ear.

In this article, we describe the case of a newborn who presented with an epiglottic cyst that was later identified as a rudimentary pinna attached to the soft palate.

Case report
A 4-day-old girl was referred to Hermann Hospital at the Texas Medical Center in Houston. She had been born at another hospital after an uneventful, full-term pregnancy. At 20 hours of life, she became tachypneic and began to exhibit signs of airway obstruction. The infant’s father noticed what he described as a mass whenever the girl opened her mouth to cry. The infant was nasotracheally intubated, and examination revealed that an epiglottic cyst was present (figure 1).

The patient was taken to the operating room, where the mass was found to be attached to the right posterosuperior surface of the soft palate. The lesion was removed, and on gross examination, it resembled a rudimentary pinna with fully intact cartilage and overlying skin (figure 2). A provisional diagnosis of nasopharyngeal hamartoma was made. The specimen was submitted for pathologic study and was diagnosed as a second branchial cleft remnant. There was no evidence of a cyst, sinus, or fistulous tract in the patient’s neck.

Discussion
Almost one of every five cervical masses in the pediatric population is of branchial arch origin. The second branchial arch accounts for as many as 95% of these cases. However, the vast majority of these masses arise in the form of a branchial sinus, fistula, or cyst; presentations such as the one that occurred in our patient are exceedingly rare.

Second branchial cleft anomalies were reported to occur as pharyngeal masses by Thaler et al, who described 2 patients with a cystic mass in the tonsil. The explanation they put forward was that the persistence of the branchial plate without mesodermic arch interposition allowed for ectodermic-endodermic apposition without a fistula into the oropharynx.

Before we attempt to explain the embryologic anomaly in this particular patient, it is important to note that this infant had two normal, fully formed ears that had undergone intrauterine maturation without any apparent aberration. It could be postulated that the hillocks on the right side divided at an early stage to form two pinnae and that one of these underwent altered development, resulting in its appearance at an ectopic site. The location of this accessory pinna might be explained by the mechanism previously discussed by Thaler et al—that is, small areas of ectodermic-endodermic apposition pulled the pinna into the pharynx. However, the presence of cartilage in...
the rudimentary pinna of our patient precluded any such appositions from existing to a significant degree, which renders this explanation somewhat unlikely.3

The developing branchial apparatus spans the distance from the lumen of the foregut to the skin. Therefore, we must understand that a small alteration in its development could lead to an unusual presentation. As we understand more about this development, we hope to be in a better position to explain such anomalies.

References