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Potential Pitfalls in the Work-up and Diagnosis of Choanal Atresia

Carl M. Black, David Dungan, Evan Fram, C. Roger Bird, Harold L. Rekate, Stephen P. Beals, and John M. Raines

Summary: To increase an awareness of the developmental anatomy of the nasal cavity as it applies to the radiologic work-up of choanal atresia and frontoethmoidal cephaloceles, we report two cases that demonstrate potentially serious imaging pitfalls. Two neonates with nasopharyngeal obstruction were imaged with CT and MR. Both patients had surgically proved bilateral bony choanal atresia. In addition to choanal atresia, CT showed a radiolucent, or nonossified cribriform plate and mucoid secretions within the nasal fossa, adjacent to the cribriform plate, which approximated the attenuation of brain parenchyma. In one of the patients, a preoperative diagnosis of nasopharyngeal encephalocele resulted in surgical exploration. At surgery, however, the cartilaginous cribriform plate was found to be intact.

At birth, the entire midline of the face may be a radiolucent stripe of cartilage situated between paired ossifications in the lateral masses of the ethmoids. On computed tomography (CT), this radiolucent, fibrocartilage midline can simulate a midline defect (1,2). We report two examples of potentially confusing imaging studies in neonates undergoing work-up for a congenital nasopharyngeal obstruction. An awareness of potential imaging pitfalls as well as a knowledge of normal developmental midline facial anatomy can help avoid misdiagnoses or unnecessary surgery.

Case Reports

Two neonates underwent imaging work-up for congenital nasopharyngeal obstruction. Each had a coronal CT study of the nasopharynx as well as magnetic resonance (MR) imaging. One patient, in whom a frontoethmoidal encephalocele was diagnosed before surgery at an outside institution underwent surgical exploration.

Case 1

A preterm boy was born at an estimated gestational age of 35 weeks. At birth he became pale, flaccid, limp, and bradycardic with no respiratory effort and required intubation. He was later extubated but immediately had recurrent airway obstruction that required reintubation. Attempts at passing nasal catheters were unsuccessful, and the infant was examined with CT and MR imaging for nasopharyngeal obstruction.

Axial and coronal 1.5-mm CT scans were obtained through the nasopharynx without contrast enhancement. An apparent midline osseous defect was seen in the cribriform plate, and brain parenchyma appeared to extend into the nasal fossa (Fig 1A). A narrowed posterior nasopharynx suggested bilateral bony choanal atresia. MR studies showed a well-demarcated lesion in the nasal fossa that was hyperintense on T1-weighted images (750/16/1[repetition time/echo time/excitations]) and slightly hypointense on intermediate- and T2-weighted images (2500/30–90/1) (Fig 1B). Coronal T1-weighted images (750/16) clearly showed this "lesion" to be separate from brain parenchyma. The nasal septal cartilage was identified and no obvious defect was seen in the roof of the nasal fossa. Bilaterally, the posterior nasal cavity contained high-signal-intensity material (Fig 1C). After nasal suction, repeat MR imaging showed gas-filled nasal passages with resolution of the high-signal-intensity material in the nasal fossa (Fig 1D). No clear communication was evident between the nasal passages and the oropharynx. This patient subsequently underwent uncomplicated, transnasal surgical repair of bilateral posterior choanal atresia.

Case 2

A full-term boy had respiratory difficulty and blood clots in the nares bilaterally at birth. Attempts to pass a nasal catheter were unsuccessful, and examination of the right nares showed fluid and a possible cystic mass. Imaging work-up and subsequent surgical exploration were performed at an outside institution. T1-weighted MR images (750/16) showed high signal intensity in the posterior nasal cavity (Fig 2A). No communication to the floor of the anterior cranial fossa was seen, and the brain appeared normal. Coronal and axial CT scans showed a nonossified floor of the anterior cranial fossa, which was thought to represent a bony defect (Fig 2B). No definite cerebrospinal fluid leak or communication was seen at cisternography. The combined imaging findings from CT, MR imaging, and cisternography were interpreted preoperatively as most compatible with a nasal cephalocele.

Surgical exploration revealed an intact cribriform plate and floor of the anterior fossa. Nasal exploration showed deformity of the nasal wall without an associated mass lesion. A repeat CT scan showed bilateral narrowing of the posterior nasal cavity, consistent with bilateral bony choanal atresia (Fig 2C). Nasal surgery confirmed the diagnosis of bilateral choanal atresia, which was then corrected surgically.

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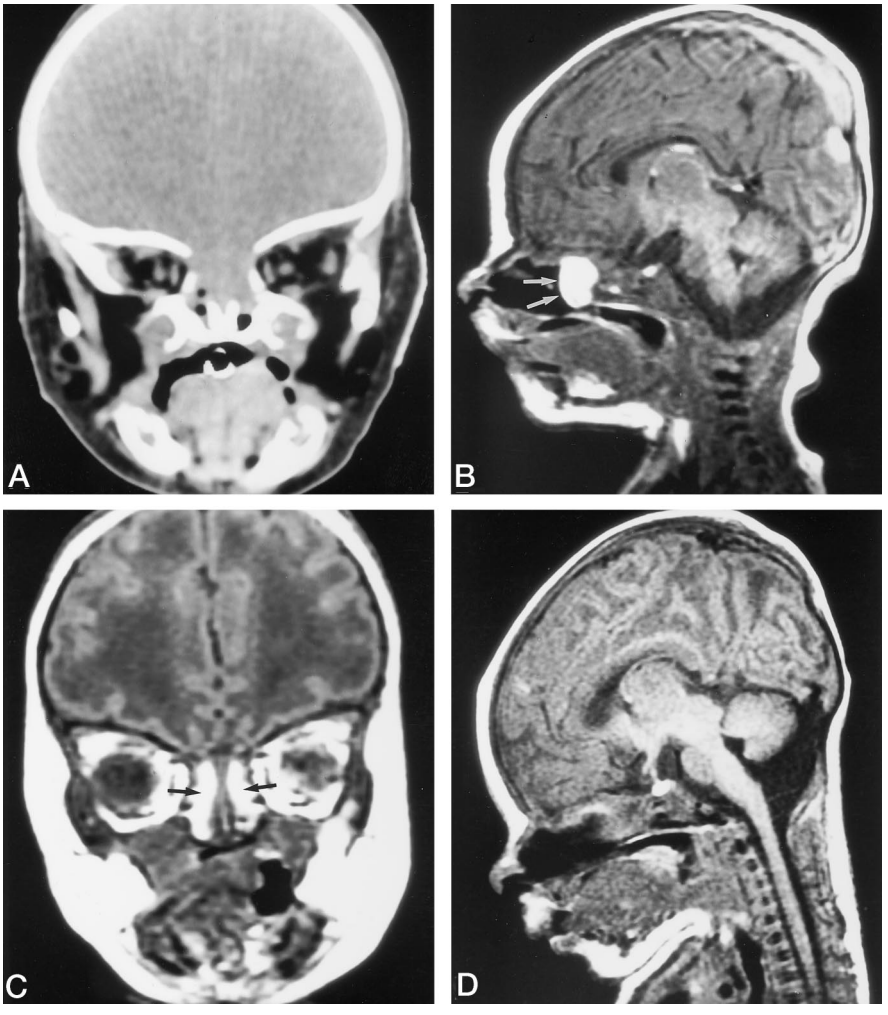

FIG 1. Preterm infant, gestational age 35 weeks.

A, On this coronal CT scan, the nonossified floor of the anterior fossa was mistaken for a midline bony defect.

B, T1-weighted sagittal MR image (750/16/1) obtained before the nasopharynx was suctioned shows hyperintense tissue (*arrows*) in the region of the posterior nasal cavity, suggesting an intranasal mass related to an encephalocele.

C, On a T1-weighted coronal MR image (750/16/1), bilateral high-signal-intensity material in the posterior nasal cavity (*arrows*) lies lateral to the septum.

D, T1-weighted MR image (700/14/1) obtained after suction of nasal secretions shows that the hyperintense material has disappeared, and there is no evidence of an encephalocele or other midline mass.

Discussion

The findings on CT scans in the work-up of these two neonates most likely reflect the normal sequential development of osseous midline facial structures (3, 4). Midline facial development is closely related to development of the cartilaginous nasal capsule. The nasal capsule provides the framework for development of the upper lateral nasal cartilage and the ethmoid bone. Portions of the maxillary bones, premaxillary bone, nasal bones, lacrimal bones, and palatine bone also form in close relationship with the roof and lateral walls of the nasal capsule. Most components of the nasal capsule ossify or atrophy with age. Only the alar cartilages, which surround the nostrils, and the anterior aspect of the nasal septum remain cartilaginous into adulthood.

The skull develops from three major ossification centers: the basioccipital, the basisphenoid, and the presphenoid. The midline septal cartilage is continuous with the cartilaginous skull base and is nonossified at birth. The septal cartilage borders on the nares anteriorly, the presphenoid ossification center and cranial base posteriorly, the cartilaginous anterior cranial fossa superiorly, and the vomer inferiorly. At birth, the lateral ethmoids, maxilla, hard palate, and vomer are ossified. The cribriform plate begins to ossify between 2 and 8 months. During the first year of life, a fourth ossification center, termed the mesoethmoid, develops in the septal cartilage anterior to the cranial base. The mesoethmoid center forms the perpendicular plate of the ethmoid and begins to ossify between 4 and 11 months. By the sixth year of life, the lateral masses and the perpendicular plate of the ethmoid unite across the roof of the nasal cavity through ossification of the cribriform plate. Prior to complete ossification, the perpendicular plate of the ethmoid and the crista galli intersect, forming a "cristal" cross (1, 4).

In a review of the CT appearance of the midline anterior cranial fossa and nasal septum in 100 children ages 2 days to 18 years, Naidich et al (1) observed that no midline ossifications of the anterior fossa or septum were present in 14% of patients less than 1 year of age. The entire midline face may be a radiolucent stripe of cartilage situated between the paired ossifications in the lateral masses of the ethmoids. This radiolucent line can simulate a midline cleft on imaging studies (1, 4). The ossification pattern of the nasal capsule and surrounding structures is diagrammed in Figure 3.

Usually, the lack of midline facial ossification is not problematic in diagnostic imaging because air abuts the cribriform plate. However, as in the two patients described here, the presence of choanal atresia may lead to accumulation of complex fluid in the nasal cavities. If the fluid attenuation approximates that of adjacent brain parenchyma, a midline encephalocele may be impossible to exclude on the basis of CT examination alone. MR imaging can play a pivotal role in such cases.

A recent case report described findings similar to those reported here (5). A full-term boy underwent CT of the nasal cavities because of respiratory distress and unsuccessful attempts to pass a nasogastric tube. Axial CT scans showed bilateral posterior choanal atresia and a questionable soft-tissue density in the nasopharynx that was subsequently imaged with coronal CT cisternography. No CSF communication was demonstrated. Three-dimensional CT reconstruction of the

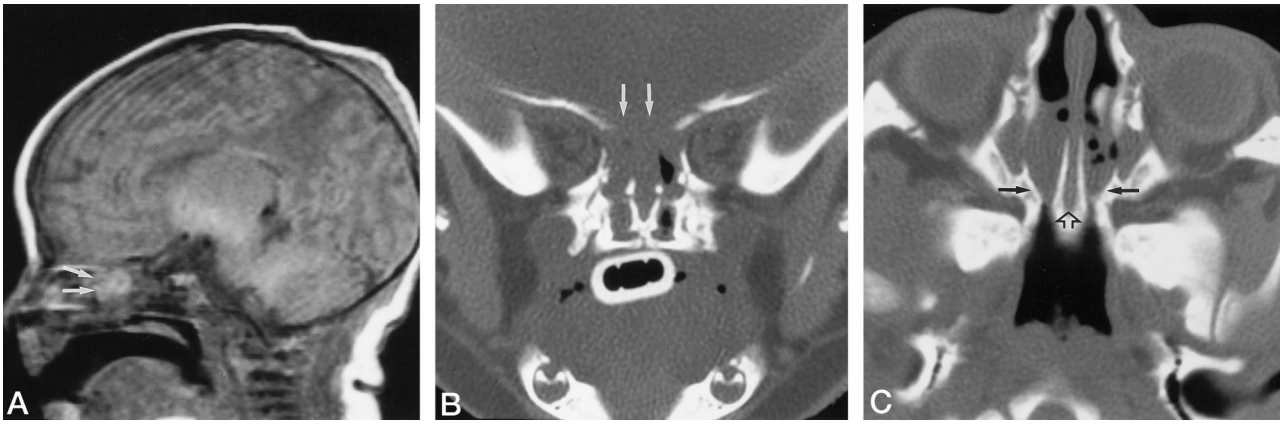

FIG 2. Full-term infant.
A, On a T1-weighted sagittal MR image (750/16/1), nasopharyngeal secretions (*arrows*) are hyperintense, simulating a mass.
B, On a coronal CT scan, the lack of ossification along the floor of the anterior fossa (*arrows*) was mistaken for a bony defect.
C, An axial CT scan shows bilateral choanal atresia, with narrowing of the posterior nasal cavity (*solid arrows*) and a widened posterior septum (*open arrow*).

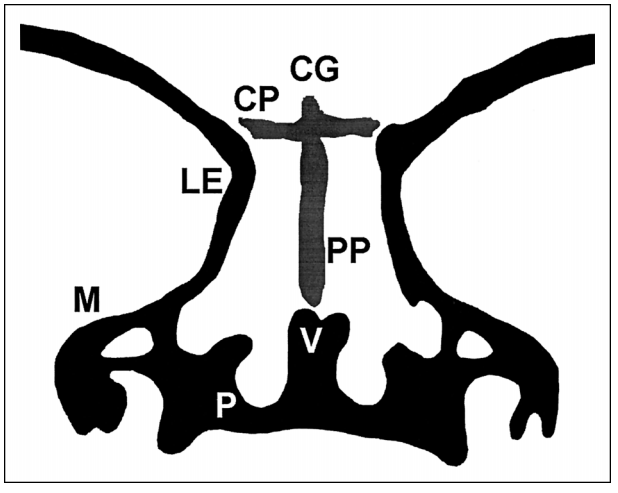

FIG 3. Diagram of a coronal CT scan in a neonate. At birth, the lateral ethmoids (*LE*), maxilla (*M*), hard palate (*P*), and inferior vomer (*V*) are ossified. The cribriform plate (*CP*) begins to ossify between 2 and 8 months. The perpendicular plate of the ethmoid (*PP*) begins to ossify between 4 and 11 months, at which time the ossifications and the crista galli (*CG*) form a "cristal cross" (1).

skull base revealed an "absence of the crista galli, cribriform plate and perpendicular plate of the ethmoid." At age 6 months, the infant underwent repair of the choanal atresia with a transpalatal approach. Telescopic endoscopy revealed no nasopharyngeal mass or encephalocele. The CT findings were attributed to a "malformation" of the anterior skull base associated with choanal atresia.

No consistent association between frontoethmoidal cephaloceles and choanal atresia has been demonstrated (6). However, several abnormalities associated with choanal atresia have been reported. In a study of 130 cases of choanal malformation, 57 had other anomalies. Most of these were from the CHARGE association (C, coloboma; H, heart defects; A, atresia choanae; R, retarded growth; G, genitourinary defects; E, ear defects). Three cases were associated with unspecified types of encephalocele (7).

CT, particularly in the coronal plane, has been advocated as a reliable method of studying nasal encephaloceles. Lundorf et al (8) reported two patients, ages 16 and 28 years, in whom coronal CT scans clearly showed bony midline basal defects. They suggested coronal CT, ideally with intrathecal contrast cisternography, as the examination of choice. However, in these adolescent and adult patients, anterior skull base ossification is complete, so any nonossified component represents a true bony defect.

Bannister et al (9) described a premature infant born at 25 weeks' gestation in whom intranasal sphenoethmoidal encephalocele was shown preoperatively by coronal CT. Difficult passage of an endotracheal tube led to an imaging work-up consisting of transaxial and coronal CT studies. They suggested that only coronal CT was necessary to detect the skull defect, because they "had all the information needed from the CT." Although this case was documented surgically, coronal CT cannot reliably distinguish a true bony defect from a nonossified anterior fossa in a neonate. Three-dimensional CT is also subject to the same limitations as direct coronal or axial CT. The three-dimensional reconstruction is keyed to the density of bone and therefore does not detect the cartilaginous floor of the anterior cranial fossa. In these patients, MR imaging plays a pivotal role in characterizing the nature of "intranasal soft tissue masses" and should be used to confirm findings demonstrated by CT.

Conclusion

An awareness of the normal ossification sequence of midline facial structures may help the radiologist and surgeon avoid the misdiagnosis of a frontoethmoidal defect, thereby also avoiding unnecessary surgery. Within the first year of life, the nonossified floor of the midline anterior cranial fossa has a CT appearance that simulates a bony defect. Usually, this lack of midline facial ossification is not problematic in diagnostic imaging because air abuts the cribriform plate. The presence of choanal atresia, however, may lead to an accumulation of complex fluid in the nasal cavities. If the fluid attenuation approximates that of adjacent brain parenchyma, a midline encephalocele may be difficult to exclude on the basis of CT examination alone. MR imaging can play a pivotal role in such cases. The combination of CT and MR imaging is useful to evaluate the ossified and cartilaginous components of the pediatric nasal-ethmoidal complex as well as suspected intranasal lesions.

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