Late presentation of choanal atresia

Introduction

Choanal atresia is a rare congenital anomaly in which the normal communication between the posterior nasal cavity and the nasopharynx fails to develop. In a third of cases, it occurs bilaterally and presents as an emergency at birth because newborn infants are obligate nasal breathers. The remaining cases usually present in infancy or childhood as chronic unilateral nasal discharge and obstruction. This report highlights a late presentation in which the diagnosis was delayed despite several consultations.

Discussion

Choanal atresia (derived from Greek meaning imperforated or occluded funnel) occurs in 1 per 6000 to 8000 births, with a 2:1 female predominance and the right side more often affected than the left. The atresia plate is bony or mixed in 90% of the cases and membranous in 10%. While the condition is usually sporadic, choanal atresia affecting siblings, identical twins and successive generations has been reported (Chia et al, 2002).

In 20–50% of cases, there may be other anomalies such as cleft palate, tracheoesophageal fistulas or Treacher–Collins syndrome. The most frequently recognized group of anomalies is the CHARGE association: Coloboma, Heart anomaly, Atresia choana, Retarded growth and development, Genital hypoplasia, Ear anomalies or deafness (Rao et al, 1986; Chia et al, 2002).

Four theories have been suggested to explain the embryological basis of its aetiology:

1. A persistence of the buccopharyngeal membrane from the foregut
2. A persistence of the nasobuccal membrane of Hochstetter
3. An abnormal persistence or location of mesoderm forming adhesions in the nasal choanal region
4. A misdirection of mesodermal flow secondary to local factors (Hengerer and Strome, 1982).

In this case, the choanal atresia caused a chronic right-sided rhinitis and nasopharyngitis, which resulted in an ipsilateral reactive lymphadenopathy. As this case demonstrates, the findings on clinical examination are subtle because it is often difficult to visualize the atresia. Other unusual presentations include unilateral maxillary sinusitis which does not respond to antibiotics (Nemechek and Amedee, 1994; Yanagisawa and Citardi, 1994).

The gold standard for evaluating choanal atresia is computed tomography (CT) scanning with axial images providing the best diagnostic and prognostic information. Pooled nasal secretions may hamper evaluation of the membranous component of the obstruction and can be confused with a soft tissue mass. Ideally, one should topically decongest the nose and suction out pooled secretions before the CT scan (Booth and Drake-Lee, 1991).

Bilateral atresia is treated immediately after birth while unilateral atresia can be managed electively after diagnosis. Four approaches to surgical repair have been described: transnasal, transpalatal, transseptal and transantral (Chia et al, 2002). A stent is usually left in place for 6–8 weeks following surgery. Endoscopic transnasal repair provides excellent visualization and enables accurate surgery to be performed on patients of all ages (Liktor et al, 2001). This has the advantage of shorter operative time, decreased blood loss and preservation of the growing plate. The transpalatal approach provides easier access to the posterior septum but requires a longer operation, potentially increased blood loss and a longer convalescent period for the patient.

Conclusions

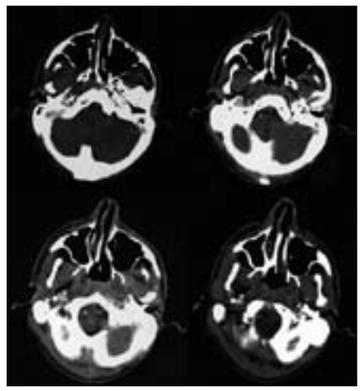
This case illustrates an unusually late presentation of choanal atresia. The non-specific symptoms meant that the diagnosis was missed on several occasions despite nasal endoscopy. This is an important lesson for trainee otolaryngologists, demonstrating that examination of the nose is not complete until the postnasal space has been inspected. For GPs and paediatricians, this shows that choanal atresia should be part of the differential diagnosis of any child or young adult presenting with long-term unilateral nasal symptoms. BJHM

Case Report

A 21-year-old man was referred to the ear, nose and throat outpatients with a 2-month history of sore throat and a right-sided neck lump, which had persisted despite antibiotics. On direct questioning, he was found to have a long history of right-sided nasal congestion and rhinorrhoea. He had previously had a nasal polyp removed from the right side 10 years before, but with no resolution in his nasal blockage. On examination there was a 1.5 cm soft, mobile, non-tender mass in the right anterior triangle. Direct inspection of the oral cavity and oropharynx was normal, while anterior rhinoscopy revealed a deviated nasal septum to the right. Blood tests were normal and an infectious mononucleosis screen was negative.

Following a further course of antibiotics, he was reviewed in clinic showing some improvement in his throat symptoms while the neck mass remained unchanged. Flexible nasendoscopy was normal on the left side, but the scope could not be passed on the right. He was offered surgery for his nasal obstruction and under general anaesthesia, a rigid scope was introduced into the right nasal cavity and the diagnosis was made. A subsequent computed tomography (CT) scan confirmed right-sided choanal atresia (*Figure 1*).

Figure 1. Axial computed tomography scans showing right choanal atresia.



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