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CLINICAL RECORD

Unilateral blindness: a unique complication of choanal atresia surgery

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Abstract

Objective: To report a unique case of unilateral blindness secondary to transnasal endoscopic surgery and stenting for right choanal atresia.

Case report: A 24-year-old man was referred with right eye blindness and acute headache, occurring immediately after transnasal endoscopic surgical repair of unilateral right choanal atresia with placement of an endonasal stent. Maxillo-facial computed tomography with three-dimensional reconstruction showed the endonasal stent entering the right nostril, passing through the lamina papyracea into the orbit and running anterior to the optic foramen towards the superior orbital fissure. Despite stent removal and medical treatment (ceftriaxone and dexamethasone), permanent right eye blindness secondary to an irreversible lesion of the optic nerve was diagnosed. At three-month follow up, an uncommon, complete fibrous obliteration of the right nasal fossa was noticed.

Conclusion: To the best of our knowledge, this is the only published report of unilateral blindness following transnasal endoscopic stenting for right choanal atresia. Causes of this complication, and ways of avoiding it, are discussed.

Key words: Nasal Surgical Procedures; Nasal Cavity; Adult; Choanal Atresia; Complications; Blindness

Introduction

Choanal atresia is a narrowing or obliteration of the posterior nasal aperture, in the latter case leading to failure of the posterior nasal cavity to communicate with the nasopharynx.[1] It is the second most common congenital nasal disorder after dermoids, and may present in complete or incomplete, and bilateral or unilateral, forms. Bilateral choanal atresia is a medical emergency and requires surgical treatment during the first days of life to allow nasal breathing in the infant. In contrast, unilateral choanal atresia may remain undiagnosed until childhood or even adulthood, the symptoms being mainly nasal obstruction, mucoid nasal discharge, anosmia, disturbed sleep and daytime fatigue.[2,3]

Different surgical techniques have been proposed for the repair of choanal atresia, including transnasal, trans-palatal and trans-septal approaches. Technical advances and growing experience with endoscopic nasal surgery have favoured the development of the transnasal approach, with a resultant reduction in the risk of choanal atresia surgery complications.[3,4]

We describe a case of unilateral blindness occurring secondary to transnasal endoscopic stenting of right choanal atresia. To the best of our knowledge, this paper represents the only published report of this complication. The causes of this event, and ways to avoid it, are discussed.

Case report

In April 2011, a 24-year-old man was referred to our department from a peripheral hospital because of right eye blindness and acute headache, both presenting immediately after transnasal endoscopic surgical repair of unilateral right choanal atresia with placement of an endonasal stent. The patient had undergone LeFort I osteotomy for surgical correction of a maxillary deformity, six years earlier, and had also suffered accidental facial trauma two years before that.

On admittance to our centre, the patient complained of a moderate, diffuse headache and was well orientated in space and time. Vital parameters were as follows: heart rate, 100 beats per minute; breathing rate, 17 breaths per minute; and blood pressure, 140/95 mmHg. The patient reported complete blindness in his right eye. Oedema of the right upper and inferior eyelids was evident. The anterior extremity of the endonasal stent, inserted during choanal atresia surgery, could be seen protruding from the right nostril. Ophthalmic examination confirmed complete right eye blindness. No other neurological deficit was noticed.

Maxillo-facial computed tomography (CT) with three-dimensional reconstruction was performed. This showed the endonasal stent entering the right nostril, passing through the lamina papyracea into the orbit, and running anterior to the optic foramen towards the superior orbital fissure (Figure 1). The CT also showed severe narrowing of the right nasal fossa, together with bony right choanal atresia.

The stent was removed. Intravenous antibiotics (ceftriaxone 2 g/day) were administered for 15 days. A tapered steroid regime (dexamethasone) was administered. No cerebrospinal fluid rhinorrhoea or epistaxis were noticed. The

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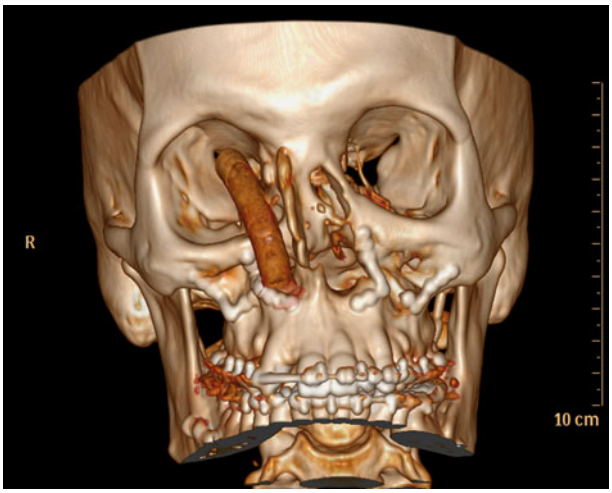

FIG. 1

Maxillo-facial computed tomography scan with three-dimensional reconstruction showing the endonasal stent entering the right nostril, passing through the lamina papyracea into the orbit and running anterior to the optic foramen towards the superior orbital fissure.

patient was neurologically monitored for 20 days after stent removal, and no sign of meningeal irritation was recorded.

Further ophthalmic assessment was conducted together with cranial magnetic resonance imaging and visual evoked potential testing, confirming the patient's right eye blindness secondary to an irreversible lesion of the optic nerve. The patient was discharged.

He returned for follow up three months later. At this visit, complete fibrous obliteration of the right nasal fossa was noted (Figure 2). The patient was offered neuronavigation-assisted surgical correction of his right nasal fossa atresia; however, he refused all surgical treatment.

Discussion

First described by Johann Roederer in 1755,[1] choanal atresia is a rare disorder with a prevalence of 1 in 5000 to 8000 live births. The female:male ratio is 2:1. The condition is frequently associated (in 20–50 per cent of cases) with other congenital abnormalities, such as the syndrome which includes coloboma of the eye, heart defects, choanal atresia, growth and/or developmental retardation, genital and/or urinary tract abnormalities, and ear abnormalities and deafness (also termed 'CHARGE' syndrome).[2,5] It had previously been thought that 90 per cent of choanal atresia cases are bony and 10 per cent are membranous; however, recent CT and histopathological research has found a higher incidence of mixed bony and membranous anomalies (70 per cent) and pure bony atresia (30 per cent), with no cases of pure membranous anomaly.[6]

Many surgical techniques have been used for the repair of choanal atresia, including transnasal (puncture, endoscopic and microscopic), trans-palatal and trans-septal approaches. A survey on the topic involving American Society of Pediatric Otolaryngology members revealed that endoscopic approaches were favoured because of their high success rate, excellent visualisation, short operative time, and low rates of bleeding and morbidity, compared with other techniques (i.e. trans-palatal).[3,4,7,8] In order to reduce the risk of re-stenosis (reported to occur in 0 to 85 per cent of cases), several

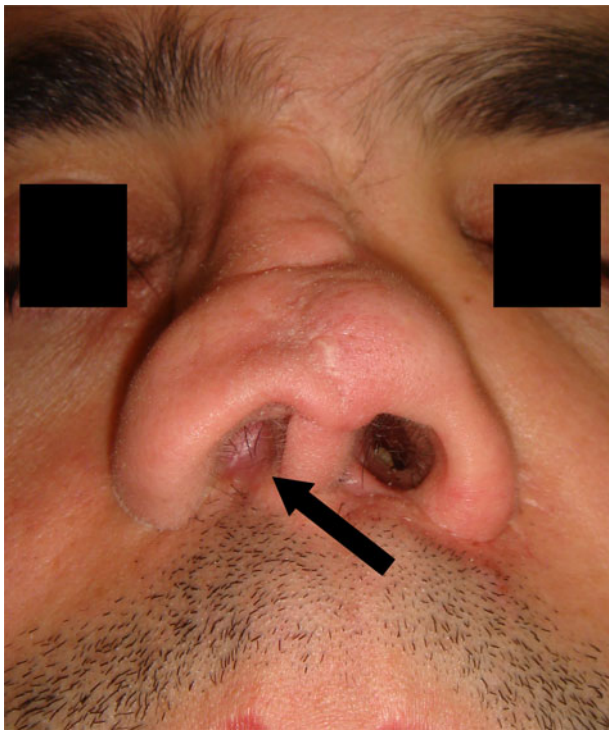

FIG. 2

Clinical photograph showing complete fibrous obliteration (arrow) of the right nasal fossa, three months after stent removal. The scarring and deformity caused by the previous facial trauma can be seen.

authors have described the use of post-operative temporary stenting, although the efficacy of this intervention is controversial.[2,9]

The severe complication we report here was probably due to the surgeon mistaking the lamina papyracea for the bony atresia plate, during endoscopic correction of the unilateral choanal atresia, with consequent drilling of the lamina papyracea and stent insertion into the posterior orbit, irreversibly damaging the optic nerve. The risk of this error may have been increased by the anatomical changes produced by the patient's previous maxillo-facial surgery and facial trauma.

- **The transnasal endoscopic approach is the treatment of choice for choanal atresia repair, due to its safety and efficacy**
- **The reported case suffered unilateral blindness as a complication of endoscopic choanal atresia surgery and stenting**
- **This surgery requires appropriate anatomical knowledge and experience, pre-operative imaging, and, in selected cases, neuronavigation assistance**

This case emphasises the importance of accurate pre-operative planning prior to choanal atresia surgery. In particular, high-resolution CT scanning should be carried out in all cases to establish the mean vomer width, nasal fossa diameter and choanal air space, and to detect other anomalies of the nasal cavity and nasopharynx (which are extremely common in patients with multiple congenital abnormalities or previous surgery).[3,9] In addition to having accurate knowledge of nasal anatomy and wide experience in endoscopic

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nasal surgery, we suggest that surgeons should use neuronavigation assistance during choanal atresia surgery, when operating upon patients whose CT scan indicates anatomical complexity, in order to reduce the risk of complications. Finally, if nasal stenting is undertaken, great attention should be paid during stent positioning, especially in cases with concurrent anatomical malformations.

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Dr L D'Ascanio takes responsibility for the integrity of the content of the paper
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