CASE REPORT

Conservative management of bilateral choanal atresia? Bilateral choanal atresia diagnosed in a 5-year-old girl

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SUMMARY

Choanal atresia (CA) is an obliteration or blockage of the posterior nasal aperture, limiting or obstructing the nasal airway to the nasopharynx. The presentation of unilateral CA commonly occurs at a later age than those children affected by bilateral CA (BCA) and can often persist undiagnosed into adulthood. BCA, on the other hand, typically presents with respiratory obstruction within the first few days of life, or desaturations with or without cyanosis when feeding or during exertion and traditional teaching is that this is a life-threatening emergency. We present the case of a 5-year-old girl referred to our department with nasal obstruction, snoring and some mild rhinorrhoea. After investigation she was found to have a mixed bony and membranous BCA and no other craniofacial abnormalities. The BCA was subsequently repaired using urethral dilators and a drill and the child is currently asymptomatic of any restenosis. This case prompts discussion of the various presentations and options in management of CA as well as allowing us an opportunity to discuss the literature on the subject.

BACKGROUND

Choanal atresia (CA) is an obliteration or blockage of the posterior nasal aperture, limiting or obstructing the nasal airway to the nasopharynx. The presentation of unilateral CA commonly occurs at a later age than those children affected by bilateral CA (BCA) and can often persist undiagnosed into adulthood.¹ BCA, on the other hand, typically presents with respiratory obstruction within the first few days of life, or desaturations with or without cyanosis when feeding or during exertion.² This is due to the obligate nasal airway of the neonate and as such the deformity is almost uniformly diagnosed within the first few days to weeks of life.² CA may be suspected in the clinical setting with an inability to pass nasogastric tubing and is almost invariably confirmed on CT imaging.^{3–5} Traditional teaching suggests that BCA is an inherent neonatal emergency and this case prompts discussion of the various presentations of CA as well as allowing us an opportunity to examine the literature on the participant. This case is an excellent example of the need to consider rarer alternative diagnoses even in the most straight-forward appearing presentations.

CASE PRESENTATION

We present the case of a 5-year-old girl who was initially referred to our services by her local general

practitioner (GP) with symptoms since birth of nasal obstruction, mouth breathing, snoring, poor sleep and tiredness on waking. A trial of topical steroids had no effect on the patient's symptoms and she was referred to our services to consider adenoidectomy.

The patient was born by spontaneous vaginal delivery in Bangladesh and the GP reported no further significant medical history in the initial referral. The family on further history reported that the patient had been suffering since birth with obstructed nasal breathing and mild rhinorrhoea. The family denied any congenital abnormalities found at the time of birth. On initial examination our patient was found to have normal ears bilaterally, no obvious craniofacial abnormalities, an absence of nasal airflow bilaterally and grade 2 tonsils. No other significant findings were apparent on initial examination and the patient was listed for adenotonsillectomy with a presumed diagnosis of obstructive sleep apnoea resulting from adenotonsillar hypertrophy.

At the time of the operation tonsillectomy proceeded without incident but a nasal catheter was unable to be passed through either nares in order to visualise the nasopharynx for adenoidectomy. Further CT scan was requested in order to investigate the apparent BCA. On postoperative follow-up her parents reported a mild improvement of her symptoms but still felt that the patient was continuing to mouth breathe. The examination on this occasion revealed significant nasal discharge as well as diminished nasal airflow on mirror/cold metal testing and the patient was referred to the senior author. During further history with a Bengali patient advocate it emerged that the patient had a very stormy infancy with the mother reporting a need for almost constant feeding owing to the fact that when the patient breast-fed for longer than 2 min consecutively she 'became blue'. It also became apparent that she was also investigated in Bangladesh with CT imaging and that the family understood from the treating team in Bangladesh that she was too small to have any corrective procedure and it was not an urgent matter for the family to correct. As such the patient and her family immigrated to the UK at the age of 11 months and initially pursued the matter no further until consulting the referring GP for nasal obstruction. The original imaging was unfortunately unavailable.

The CT scan of her sinuses revealed a bilateral bony and membranous CA (figures 1 and 2).

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Hence day-case endoscopic BCA repair was planned for early March 2013. Cophenylcaine spray was used topically as nasal preparation. BCA was confirmed using a 120° telescope (figure 3). The CA was mixed membranous and bony on examination. Bilateral choanal membranes were perforated using urethral dilators (figure 4) and dilated up to size 14/16 gauge (figure 5). Diamond burr of 3 mm and 4.2 mm cutting burr were used to widen the bony component of the atresia. No stents were inserted at the time and examination under anaesthesia with or without dilation was planned 4 weeks later. The patient was sent home with ephedrine nasal drops, β -methasone nasal drops, simple analgesia and a short course of prophylactic coamoxiclav.

Three subsequent examinations under anaesthetic have revealed patent airways bilaterally. On all occasions, minimal soft tissue obstruction on the right has required trimming only. The most recent examination revealed a good surgical result bilaterally and showed no signs of stenosis of the airway. On follow-ups there have been no symptoms of nasal obstruction and the patient continues to be asymptomatic.

INVESTIGATIONS

CT scan: bilateral mixed bony and membranous CA.

DIFFERENTIAL DIAGNOSIS

- ▶ Adenoid hypertrophy
- ▶ Rhinitis

OUTCOME AND FOLLOW-UP

Our patient is currently asymptomatic and is awaiting one further examination under anaesthetic to confirm absence of restenosis.

DISCUSSION

CA can be unilateral or BCA and BCA has been documented in numerous mammalian species including humans, horses and on one occasion, a llama.⁶ The comprising tissue may be bony or membranous,²⁻³ with a preponderance for mixed composition,³ however, it has also been suggested that with improving imaging technology it is becoming apparent that the vast majority, if not all, cases have some form of bony occlusion.⁵ The condition is commonly associated with congenital syndromes such as CHARGE (30%), which is characterised by at least four of

Coloboma, heart abnormalities, CA, retarded growth, genital and ear abnormalities.⁷⁻⁸ While these are the classically reported findings in CHARGE, there have been additional clinical findings added to the spectrum including dysmorphic features, rhombencephalic dysfunction and arhinencephaly.⁸⁻⁹ CA and BCA can appear in isolation in an otherwise healthy child. The incidence of CA in the UK is between 1:5000 and 1:8000 births with females more common than males.⁹

The vast majority of patients with BCA are diagnosed in the neonatal period. While there are cases in the literature of patients diagnosed with BCA in adulthood,¹⁻² these have predominantly been in developing nations where access to health-care is less available and lower priority symptoms such as nasal discharge and mouth breathing may be less likely to receive the same extent of investigation as those in developed nations. It is extremely unusual for a patient to present in a developed nation with a BCA at the age of 5. As the neonate is an obligate nasal breather,¹⁰ it is not surprising that the family report a requirement for near-constant breast-feeding, as feeding for over 2–3 min was found to provoke cyanotic episodes. More surprising than this is the apparent decision by the treating team to treat this case expectantly and to await weight gain prior to performing the BCA repair. This runs contrary to traditional teaching and suggests that in some cases the condition may not be inherently life-threatening. Given the difficulties encountered with feeding and caring for our patient in her first year of life, however, it is clear that the family would have been saved significant unnecessary distress with earlier resolution of her BCA. On reflection of her management in the UK this case is an excellent reminder of the need to consider rarer diagnoses for nasal obstruction, including children where the most common diagnoses are adenoidal hypertrophy and rhinitic disease.¹¹ In this instance the complete absence of nasal airflow should prompt the consideration of an anatomical abnormality.

The patient underwent CA repair bilaterally as part of a single procedure. Initial opening of the atretic membrane was performed with urethral dilators and subsequently further opened with a 3 mm diamond burr drill. There were no stents used in the procedure nor was there any use of mitomycin C (MMC), a compound which has been suggested to be useful in the past in minimising restenosis of CA repair.¹² Traditional methods and teaching in CA repair suggest that postoperative measures should include placements of stenting devices in order to

Figure 1 Axial CT illustrating bilateral choanal atresia with bony as well as membranous composition.

Figure 2 Sagittal plane CT showing complete obstruction of the choanal opening on the left.

Figure 3 Intraoperative photograph: view of the choanae and nasopharynx from the oropharynx with a 120° Hopkins rod.

Figure 5 Completed repair of bilateral choanal atresia.

decrease the chances of restenosis, however, in their recent review of CA repair, Cedin *et al*¹² found no significant differences between repair methods nor use of MMC with regard to rates of restenosis nor with regard to total number of procedures.¹² In the review it was noted that between four and six total procedures per patient was typically required in order to achieve complete repair with no restenosis.¹² Our case, using a combined oronasal endoscopic approach with no stenting or adjuvant medications has to date undergone a total of four procedures including the initial repair, in keeping with the expected numbers of required procedures as reported in previous literature.

Perhaps equally striking in this case is the importance of patient advocacy and the irreplaceable value of a thorough history. As the parents of our patient spoke passable English they were not offered the option of a Bengali advocate on many occasions and felt that they were able to 'struggle through.' As a result of this, many salient points were missed in the history, which would have hastened diagnosis as well as saving the patient unnecessary investigations and operations. Our patient had classic symptoms suggesting BCA, however, these were difficult to elucidate due to the difficulties in communication and the family's possible expectations of a paternalistic approach to the healthcare of their daughter. It is difficult to elucidate the exact events in diagnosis and treatments offered to our patient in Bangladesh, however, it is reasonable to expect that the family's low levels of education may have contributed to confusion around the treating team's management plan. In any case,

there was insufficient explanation to communicate to the family the need for intervention and the implications of non-treatment. As such it is a salient reminder of the vulnerabilities of the paediatric patient, the patient from non-English speaking background and the importance of effective communication of the treating team's diagnosis and management plans.

This case allows us the opportunity to explore a few key points in BCA and also gives an opportunity to examine the current literature with regard to CA repair. It also suggests that on very rare occasions the condition may be managed in a non-emergent fashion. Despite this it is the author's opinion that the condition should in the vast majority of cases be managed as a neonatal emergency requiring early intervention in order to allow improved feeding and respiratory function. There is no established gold-standard technique for opening of CA at this point in time in the literature and in the authors' experience we have found the use of urethral dilators and drill to be a satisfactory and safe method of CA repair with similar rates of reoperation to other techniques.

Learning points

- ▶ Choanal atresia has an incidence of between 1 in 5000 and 1 in 8000 live-births and the most common association is CHARGE with a 30% correlation.
- ▶ Bilateral choanal atresia is normally diagnosed in the first days of life with respiratory obstruction or episodic desaturations when feeding which is improved with crying and is traditionally considered a neonatal emergency.
- ▶ The diagnosis of choanal atresia is made by CT imaging.
- ▶ There is no current evidence to suggest that any of the known methods of choanal atresia repair have better outcomes than others.

Figure 4 Initial dilation with urethral dilators.

Contributors All the authors were instrumental to the authorship and review of the submitted case report.

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