CASE REPORT

Paediatric nasal polyps in cystic fibrosis

Mohd Afiq Mohd Slim, David Dick, Keith Trimble, Gary McKee

Department of Ear, Nose and Throat, Royal Victoria Hospital, Belfast, UK

Correspondence to
Mohd Afiq Mohd Slim,
chain1993@yahoo.com

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SUMMARY

Patients with cystic fibrosis (CF) are at increased risk of nasal polyps. We present the case of a 17-month-old Caucasian patient with CF who presented with hypertelorism causing cycloplegic astigmatism, right-sided mucoid discharge, snoring and noisy breathing. Imaging suggested bilateral mucocoeles in the ethmoid sinuses. Intraoperatively, bilateral soft tissue masses were noted, and both posterior choanae were patent. Polypectomy and bilateral mega-antrostomies were performed. Histological examination revealed inflammatory nasal polyposis typical of CF. The role of early functional endoscopic sinus surgery (FESS) in children with CF nasal polyposis remains questionable as the recurrence rate is higher, and no improvement in pulmonary function has been shown. Our case, however, clearly demonstrates the beneficial upper airway symptom relief and normalisation of facial appearance following FESS in a child with this condition.

BACKGROUND

Cystic fibrosis (CF) is the most common autosomal recessive disease in the Caucasian population occurring in about 1:2400 newly born in the UK.¹ It affects organs with an epithelial lining, most notably the lungs and the pancreas.² A mutation that encodes the CF transmembrane conductance regulator (CFTR) protein produces an abnormally viscous secretion that subsequently causes obstruction, tissue inflammation and destruction.²

Owing to the pathophysiology of CF, the upper airway suffers the same fate as the lower airway as these are lined by the same epithelial cells.³ This manifests in the upper airway as nasal polyps and chronic rhinosinusitis.^{4–5} Nasal polyps can be seen in up to 86% of children with CF, but only in 6% of those under 6 years of age.^{6–11} Those with the $\Delta F508$ mutation are also more prone to have polypoidal manifestation.^{12–13} Recognising this association is important (figure 1) as the symptoms of chronic rhinosinusitis which are secondary to nasal polyps tend to go unrecognised, necessitating a high index of clinical suspicion.^{5–9–14} The presence of nasal polyps has no impact on pulmonary function, overall outcome and nutritional status.⁹ Unfortunately, polyps in patients with CF are associated with early *Pseudomonas aeruginosa* colonisation, more acute pulmonary infections, and hospitalisation.^{9–12} To the best of our knowledge, this is the seventh report addressing the presence of bilateral soft tissue masses occupying the nasal cavity and sinuses in a child with CF.

CASE PRESENTATION

A 17-month-old Caucasian boy with a background history of CF ($\Delta F508/N1303K$), diagnosed from

the neonatal screening programme, was referred to the otorhinolaryngology (ORL) service for a 1-year history of persistent right-sided mucoid nasal discharge associated with snoring, noisy breathing, nasal speech, and an 11-week history of frequent dry cough and sneezing. His mother also had concerns about his facial appearance.

He is a second child with one elder sister without CF. He had an uncomplicated vaginal delivery with up-to-date vaccinations and was achieving all of his cognitive developmental milestones. Prior to our review, he had frequent bouts of upper respiratory tract infection (URTI), two of which required hospitalisation despite already being on regular oral prophylactic antibiotics (flu-cloxacillin), nebulised isotonic saline and Dornase α . He was also being managed for failure to thrive that was secondary to pancreatic insufficiency.

On examination, there was marked hypertelorism with proptosis of the right eye. This was associated with significant astigmatism (8 Dioptre) and reduction in visual acuity following a formal assessment (6/24 with forced preferential looking) by an ophthalmologist. Anterior rhinoscopy revealed bilateral mucoid nasal secretion with difficulty in passing of a suction catheter and a nasogastric tube through the right nostril. The remaining ORL examinations were unremarkable. Awake nasal endoscopy was not performed here as plan imaging was indicated; operative intervention was deemed necessary afterwards. So, a CT scan of the paranasal sinuses was requested.

INVESTIGATIONS

CT of paranasal sinuses demonstrated soft tissue masses from the ethmoid sinuses obstructing the nasal cavity (figure 2). The masses completely blocked both osteomeatal complexes (figure 3) with associated opacification of maxillary sinuses. The masses also caused bilateral lateral displacement of the medial orbital wall, more prominent on the right side, resulting in lateral shift of the right eye (figure 4). A possible bony outline defect on both medial orbital walls was noted, but could not be confirmed due to the nature of the low resolution CT utilised here in view of his age. There was also some soft tissue opacification of the sphenoid sinus.

DIFFERENTIAL DIAGNOSIS

The most common cause of unilateral mucoid nasal discharge in this age group is nasal foreign body. Given his unilateral mucoid nasal discharge with restricted nasogastric tube insertion, a unilateral posterior choanal atresia had to be excluded. Massive antrochoanal polyps or mucocoeles were also considered here because of his CF and

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Figure 1 Pathophysiology of the upper airway in cystic fibrosis.^{5 9}

recurrent URTI background. Meanwhile, changes in his facial morphology also raised the concern of an invasive process such as nasal tumour.

TREATMENT

In view of the deteriorating right-sided vision and proptosis, functional endoscopic sinus surgery (FESS) was carried out with the aim to remove the masses and decompress the sinuses. Intraoperatively, large soft tissue masses obstructing both nasal cavities were noted ([figure 5](#)). Decompression of the mass revealed a thick yellow-green toothpaste like discharge ([figure 6](#)) suggesting mucocele. Further examination also revealed a

Figure 2 CT—Axial view of the paranasal sinuses showing a mass arising from the ethmoid sinuses filling the nasal cavity.

Figure 3 CT—Coronal view of the paranasal sinuses showing the blockage of the osteomeatal complexes (Green Circle).

Figure 4 CT—Coronal view of the paranasal sinuses showing displacement of the medial orbital wall with lateral shift of the right eye (Green star=Approximate central alignment of the eye).

Figure 5 Pale-colour soft tissue mass in the right nasal cavity.

positive Stankiewicz sign bilaterally, with absence of lamina papyracea ([video 1](#)) subsequently confirming the CT findings. Both choanae were patent. Unhealthy mucosa was stripped, and polypectomies were performed. Mega-antrastomies were

Figure 6 Yellow-green toothpaste like discharge from the ethmoid sinus.

Video 1 Stankiewicz sign from the right side.

fashioned at the maxillary ostia. The anterior ethmoidal arteries and turbinates were preserved. Samples from mass and the discharge were sent for microbiology and histological assessments.

OUTCOME AND FOLLOW-UP

The cultures grew coliform bacteria. Histological examination of the soft tissue mass showed respiratory mucosa with intense mixed inflammatory cell infiltration (plasma cells, lymphocytes, neutrophils) and some entrapped glandular elements of retained secretions, typical of inflammatory nasal polyposis in CF (figures 7 and 8). No granulomatous inflammation or any characteristics suggesting a malignant lesion were noted.

The child had an uncomplicated postsurgical recovery. He was discharged with isotonic saline nasal rinses and betamethasone 0.1% nose drops (2 drops, three times a day) for 5 weeks. At his first 4-week follow-up appointment, the unilateral mucoid discharge had settled and there was also a marked improvement in his facial appearance. His parent also noticed improvement in his speech pronunciation and a complete resolution of his snoring.

Further follow-up at 8 weeks with his ophthalmologist also revealed substantial improvement in his right eye astigmatism (3.5 Dioptre from 8 Dioptre), which confirmed the cycloplegic effect the mass had on visual acuity. He does, however, still require glasses to correct the residual astigmatism. Otherwise,

Figure 7 Retained mucin within the polyp.

Inflammatory cell infiltrate just below surface of polyp – lymphocytes and plasma cells mostly

Figure 8 Inflammatory cells within the polyp.

he is due for his first 3-month follow-up with planned imaging or examination under anaesthesia depending on his symptoms.

DISCUSSION

Literature search using the following search strategy (table 1) was conducted using PubMed. Six case publications were identified through the search strategy. Unfortunately, the other three case publications cannot be verified by the authors due to access restriction.^{15–17} Based on the other two previous case reports

Table 1 Search strategy utilised for this case report

Search terms	Items identified
1 cystic fibrosis	43 532
2 (mucocoeles OR nasal polyps OR polyps OR polyp OR polyposis) AND bilateral	701
3 1 AND 2	19
4 child OR children OR paediatric OR infant	2 596 839
5 3 AND 4	13

Table 2 Summary of cases that presented with bilateral soft tissue mass in the nasal cavity for patients with cystic fibrosis

Number	Authors	Year	Sex	Age, months	Point mutation	Presenting symptoms	Soft tissue diagnosis
1	Thomé <i>et al</i> ¹⁸	2000	Male	10	Unknown	Rhinorrhoea, nasal obstruction	Mucocele or pseudomucocele
2	Di Cicco <i>et al</i> ¹⁰	2005	Male	6	ΔF508/G542X	Nasal obstruction, chronic rhinitis	Mucocele
			Male	52	ΔF508/ΔF508	Snoring and apnoea	
			Female	50	ΔF508/ΔF508	Epiphora	
			Female	26	ΔF508/Unknown	Epiphora, nasal obstruction	
			Female	31	ΔF508/ΔF508	Unilateral nasal obstruction	
			Female	53	ΔF508/ΔF508	Nasal obstruction, snoring	
3	Portes <i>et al</i> ¹⁹	2007	Male	24	Unknown	Rhinorrhoea, nasal obstruction, noisy breathing, snoring	Pseudomucocele
4	Current	2015	Male	17	ΔF508/N1303K	Unilateral rhinorrhea, snoring, noisy breathing, cough, sneeze, unable to pass catheter into nostril	Inflammatory nasal polyp

and one case series (table 2), the average age of symptomatic presentation for patients with bilateral soft tissue mass in the nasal cavity for CF is at 29.89 months of age. Further analyses also show the ΔF508 mutation and male (M:F=5:4) as being the predominant factor for nasal polyposis presentation in the CF population.^{7 12 18}

The pathophysiology (figure 1) for nasal polyp formation in CF is different from the non-CF population. Understanding the effect that the viscous secretions have is important as the conventional symptomatic criteria for diagnosing chronic rhinosinusitis in CF might not be a sensitive tool, as only 10–15% of patients with CF will complain about upper airway symptoms despite fulfilling 80–85% of the European position paper criteria on rhinosinusitis.^{4 20} Histologically, nasal polyps in CF consist of dilated mucous glandular structures, lack of eosinophils and a higher volume of neutrophils, mast cells and plasma cells (figure 8).^{5 11} Microscopically, persistent bacteria colonisation is the reason for a prolonged inflammatory response in the sinuses of patients with CF rather than the common viral vector.⁴ *P. aeruginosa* is the commonest occupant, with a 95% prevalence rate.⁴ This highlights the theory of ‘united airway’ where the sinuses act as a reservoir allowing chronic colonisation and subsequently, a starting point for recurrent pulmonary

complications.^{4 14} Nevertheless, the presence of nasal polyps do not seem to predict CF disease severity.¹²

In view of the high prevalence of bacterial colonisation in the sinuses of patients with CF and the possible complications that can result, early treatment focusing on the decolonisation of the sinuses has been recommended.^{4 21} Parenteral medication limits treatment efficacy as the sinuses have poor blood perfusion.⁴ Instead, direct topical delivery of medications into the sinuses allows a higher concentration of drug to be delivered to the target site. This prolonged contact of the medication with the sinus mucosa makes it more efficient and effective while minimising systemic exposure.²² Based on a mathematical model, the conventional method of nasal lavage will only be effective in CF sinuses following FESS, as only then will the narrowed osteomeatal complexes be made large enough to clear the viscous secretion that drives the chronic inflammation and provides a medium for colonisation.²² Meanwhile, nasal sprays deliver large size droplets without any aerosol effect to the sinuses that tend to deposit on the anterior portion of the nose.²² Thus, these factors (figure 9) need to be addressed to allow effective drug delivery into the sinuses.^{21 23 24}

In children, findings such as demineralised bilateral uncinate processes and medial displacement of the lateral nasal wall in the

Figure 9 Factors effecting effective drug delivery into the sinuses.^{21 23 24}

CT of paranasal sinuses should raise the suspicion of CF.⁶ The CT scan of the paranasal sinuses in patients with CF, however, invariably shows anatomical alteration when compared with the non-CF population.^{14 25} Fortunately, there is no proven correlation between image findings with the chronic rhinosinusitis disease severity.^{6 9 14} Interestingly, one study has shown an inverse correlation between the endoscopic and the CT findings of the sinuses with the CF disease severity score, the Shwachman and Kulczycki scoring system.²⁵ This is, however, not an indication to perform a routine CT scan of the paranasal sinuses in all patients with CF if FESS is not to be performed. Pre-FESS CT scan of the paranasal sinuses allows preoperative planning and anatomical evaluation to be performed, as well as determining the extent of the surgery.^{9 14} MRI has a very limited pre-FESS role due to its inability to evaluate the bony structures despite allowing better soft tissue differentiation and no exposure to radiation.^{4 9 14}

Currently, the evidence behind the management of upper airway in CF is not well established.⁵ This includes the first line management plan and the timing for FESS. Conventional management of chronic rhinosinusitis calls for medical management as the first line management, and FESS only for intractable disease.^{6 9 20} Nasal saline lavage has been shown to improve symptoms control in non-CF chronic rhinosinusitis.^{5 22} Nebulised Dornase- α assists in decreasing the mucous secretion viscosity by DNA fragmentation, thus improving nasal symptoms and endoscopic findings.^{5 26} Topical steroid application is able to decrease the polyp size, but has limited effect on nasal symptoms due to the neutrophilic drive in CF nasal polyps.^{5 26} One might argue that due to the pathophysiology involved in CF, anatomical alteration of the osteomeatal complexes via FESS should be performed first to assist in the drug delivery into the sinuses. It seems performing FESS early in a child does not have any detrimental effect on the craniofacial development as first thought.²⁷ Reviews have also shown a good safety profile for FESS in patients with CF.^{8 28 29} FESS relieves nasal symptoms, decreases hospitalisation and assists with *P. aeruginosa* eradication. This subsequently reduces pulmonary complications and reduces the number of parenteral antibiotics needed.^{4-6 8 9 29 30} However, no effect on the pulmonary function was noted.^{4-6 8} Owing to the persistent underlying pathophysiology in CF, extensive nasal polyps will have high recurrence rate that potentially require further revision FESS in the future.^{5 29} This, however, could be minimised by performing serial antibiotic lavage.⁸

Learning points

- Have a low threshold for suspecting cystic fibrosis in a child presenting with nasal polyps.
- Management of sinonasal disease in cystic fibrosis could have a beneficial effect on the patient's overall morbidity.
- Be aware of the new aerosolised delivering system that can also influence the health of the upper airway in cystic fibrosis.
- Recognise the benefits and limitations of functional endoscopic sinus surgery in the management of sinonasal disease in cystic fibrosis.
- Be alert to parental concern about change in facial appearance, which should prompt urgent evaluation by paediatric ophthalmology or otorhinolaryngology.

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