Chronic sinusitis in cystic fibrosis

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Introduction

Rhinosinusitis occurs commonly in children with cystic fibrosis as a result of the altered viscosity of secretions. It presents with nasal congestion, 'post nasal drip' and may be associated with facial pain, headache and hyposmia. Chronic infection results in swelling and oedema of the mucous membranes lining the nose and paranasal sinuses and the lining of the ethmoidal sinuses may prolapse down as polyps further obstructing the middle meatus and leading to a vicious cycle.

Differential diagnosis

Another rare cause of chronic rhinosinusitis in children is primary ciliary dyskinesia (PCD), a syndrome characterized by bronchiectasis, sinusitis and situs inversus which was first described by Kartagener in 1933 though it was not until 1976 that Afzelius[1] and Pedersen and Mygind[2] revealed that this was associated with an abnormality of ciliary motility. Poor or absent ciliary motility leads to sinusitis, bronchiectasis and dextrocardia (in 50% of the patients) due to random rotation of the archenteron during embryonic development. In adults bronchiectasis, sinusitis and reduced fertility may also be seen in patients with normal ciliary activity but abnormally viscid mucus - Young's syndrome[3]. Examination of the semen reveals azoospermia due to lack of sperm transport down the genital tract at the level of the caput epididymis where the sperm may be found in viscous fluid.

Choanal atresia, if bilateral, will result in symptoms at birth, unilateral atresia however can be surprisingly difficult to diagnose. It should always be considered in cases of unilateral nasal obstruction. Other mechanical factors which need to be excluded are adenoidal hypertrophy and foreign bodies.

Any child presenting with nasal polyps should raise the suspicion of cystic fibrosis until proved otherwise. Even after this has been excluded the possibility of a meningocoele, encelaphalocoele, dermoid cyst, glioma or foreign body should be considered before proceeding to removal of 'simple polyps' as unlike adults these are very rare in children.

Other conditions which may give rise to chronic infection of the sinuses would include rare infection by specific organisms such as tuberculosis, rhinoscleroma (*Klebsiella rhinoscleromatis*) leprosy, yaws (*Treponema pertenue*) or chronic glanders (*Loefflerella mallei*). The nose and sinuses may also be chronically infected by fungi and yeasts particularly in immunocompromised patients.

Examination

Physicians who are not used to examining the nose with a headlight and mirror may find it easier to use an auroscope with large speculum. Even when examining the nose with a headlight and nasal speculum, it is difficult to examine the middle meatus and posterior half of the nose. A better view can be obtained with an endoscope which, even with small children, is usually tolerated well. Endoscopy can be performed with a small flexible nasendoscope or a 4 mm or 2.7 mm, 30° angled rigid Hopkins Rod.

Polyps may be attached by a narrow pedicle or appear as sessile 'cobblestoned' mucosal lining. The anterior end of the middle turbinate may swell to present a polypoidal appearance, but the inferior turbinate should not be confused with polyps as the latter are mobile and insensitive to gentle probing. Young children, and sometimes adults who have been left untreated for some time may develop broadening of the bridge of the nose and lateral displacement of the eyes (frog face).

Imaging

Plain radiography of the paranasal sinuses in patients with CF will usually reveal small or undeveloped frontal sinuses and hazy maxillary sinuses with a 'ground-glass' appearance.

The frontal sinus does not develop in most cases until the age of six years or older. It is very variable in size and is absent in 1% of the normal population. For the purposes of this paper we reviewed the plain radiographs of 47 patients with CF and compared these with 47 'non-cystic' patients matched for age. This revealed no discernable frontal sinus in 36% of cases with CF. The size of the frontal sinus was measured on the plain AP view using cell slide summagraphic analysis. Figure 1 shows the overall difference in the size of the frontal sinus in the two groups for each age range. From this it can be seen that the frontal sinus, even when present is usually considerably less well developed than normal. All 47 patients with CF were 'positive' in that they revealed

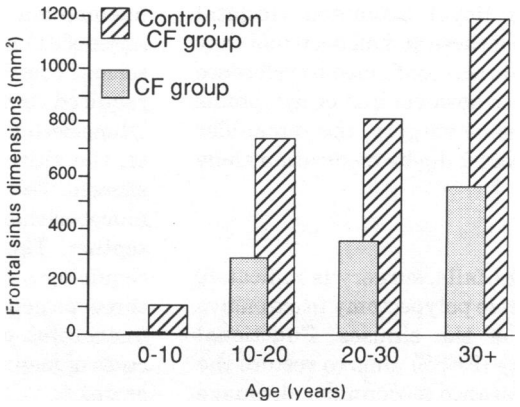

Figure 1. Comparison of frontal sinus dimensions: cystic fibrosis vs control group

increased opacity of the maxillary sinuses usually reported as 'mucosal thickening of the maxillary antra'. Apart from confirming the 'normal appearance for CF patients', little else is gained from plain films and certainly repeated plain films are of no value.

Plain radiographs of the sinuses even in non-cystic patients, can be misleading, particularly in infants and children[4] and in both adults and children it is seldom possible to identify the middle meatus and maxillary ostium (ostio-meatal complex), anatomical variants or accurately judge important surgical features. Zinreich[5] has popularized a technique for computer assisted tomography (CT) of the sinuses in the coronal plane using 'wide windows'. This technique clearly reveals the pathological changes, which in cystic fibrosis is characterized by uniform thickening of the lining of all the sinuses, increased bone density probably associated with an underlying chronic osteitis and broadening of the ostio-meatal area. It is also extremely important to undertake scanning in this manner prior to undertaking endoscopic sinus surgery to identify the anatomical landmarks of each individual patient.

Gross polyposis will result in a 'white-out' of the scan with no anatomical landmarks visible, thinning of the roof of the ethmoid and lamina papyracea and is associated with increased risk for complications and a poor prognosis.

Medical treatment

Treatment of both rhinosinusitis and the resulting nasal polyposis should in the first instance be medical, with broad spectrum antibiotics and topical anti-inflammatory drugs. Nasal polyps respond to systemic and topical corticosteroids. Betamethasone drops in the head down and forwards position (allowing the drops to run via the middle meatus into the ethmoids) have been shown to be effective[6], though in children these should not be used as long-term treatment, as they are absorbed and do have a systemic effect. Maintenance treatment with topical corticosteroid sprays (eg beclomethasone, budesonide or fluticasone) may be useful in preventing recurrence after medical or surgical treatment.

The organisms most commonly cultured from the sinuses in our group of CF patients have been *Pseudomonas aeruginosa* and *Staphylococcus aureus*. Long term treatment with systemic antibiotics for both upper and lower respiratory tract infection frequently results in antibiotic resistance and the choice of antibiotic for each patient will be dependent on culture and sensitivities.

A recent questionnaire of 200 patients (or their parents) attending the Royal Brompton Hospital resulted in 118 replies. Of these 46 had been told they had nasal polyps (and this was confirmed by reference to their notes), 16 of these however had no symptoms and only 12 had undergone surgery, the remainder having been treated successfully by medication alone (topical steroids).

Surgical treatment

When medical treatment fails, surgery is indicated. This may vary from simple polypectomy to extensive surgical exenteration of the sinuses. Functional endoscopic sinus surgery (FESS) aims to restore the natural mucociliary clearance mechanism, drainage and aeration of the sinuses, by a minimally invasive technique maintaining as much of the normal anatomy as possible. The surgical technique commences anteriorly concentrating particularly on the ostio-meatal complex, the anterior ethmoid, its infundibulum and the middle ethmoids and may progress posteriorly, superiorly and laterally, but only as far as necessary.

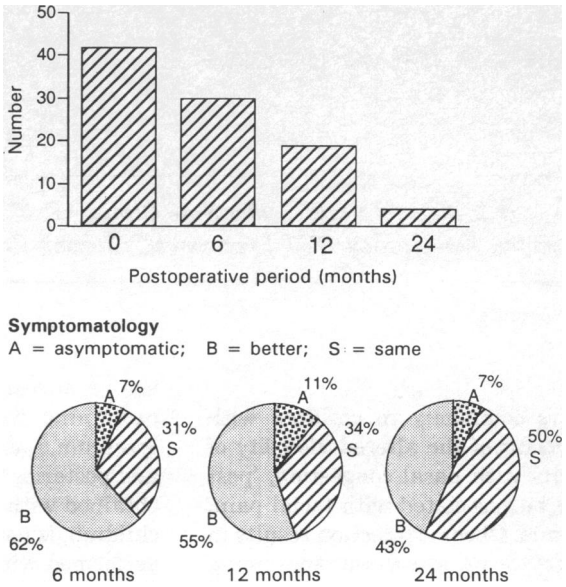

Figure 2. Postoperative follow-up of patients with cystic fibrosis following functional endoscopic sinus surgery

An audit of 611 patients undergoing endoscopic sinus surgery under the care of the senior author has been undertaken between January 1990 and September 1993. Of these, 42 patients had CF, 15 of whom had previously undergone heart-lung transplantation. Figure 2 shows the results for the patients symptom scores based on their subjective assessment and was scored as 'A' for asymptomatic, 'B' for better, 'S' for same or 'W' for worse. The overall results (for cystics and non-cystics) reveal that 86.59% considered themselves asymptomatic or improved. The CF patients particularly the more severe group who had required heart-lung transplantation responded least favourably, though 69% regarded themselves as better (or 'cured') at 6 months, 66% at 12 months and 50% at 2 years.

Complications

In a series of over 4000 cases, Stammberger[7] reported only two cases of cerebrospinal fluid (CSF) rhinorrhoea, no intracranial complications and no ophthalmic complications. Wigand[8] reporting on 220 patients undergoing complete ethmoidectomy reported a CSF leak in two patients (0.9%) and one case of orbital haematoma (0.5%). In the authors' consecutive series of 611 patients undergoing FESS there were three serious complications. An orbital haematoma which required immediate decompression via an external ethmoidectomy. One case of CSF rhinorrhoea occurring on the third postoperative day, when the patient sneezed. This was managed endoscopically with a free mucoperichondrial graft from the opposite side of the septum. The third complication was a dural tear requiring a septal flap via an external incision. All three patients made a satisfactory and otherwise uneventful postoperative recovery. There were no cases of meningitis, brain abscess, blindness, diplopia or death.

Nasal packing is avoided and usually all that is required is a small (Merocel) sponge soaked in 1 : 1000

adrenaline placed high up in the ethmoids. This is usually removed in recovery and the patient returned to the ward with no packing. Postoperative bleeding is seldom a problem; of the 611 patients in this series, three required a postoperative transfusion. It is relevant however, that all three had cystic fibrosis. Another complication specific to the CF group was one case of intestinal obstruction following the use of an analgesic containing codeine. Postoperative pain is very variable following this surgery, many patients experience no pain at all, occasionally however, the pain can be intense and in a previous audit 9 out of 411 patients had experienced more than the discomfort they expected.

In a survey of British otolaryngologists undertaken in January 1993 a questionnaire was sent to 653 members of the British Association of Otolaryngologists, 375 replies were received (57.4%) of which 138 surgeons were routinely undertaking FESS. These surgeons estimated that between them they had carried out endoscopic sinus surgery on 15 000 patients with the following serious complications: CSF leak (25), orbital haematoma requiring decompression (4), arterial ligation (2), temporary blindness (2), pneumatocoele (1), permanent blindness (1) and one case of transient diplopia. This gives an incidence of serious complications of 0.24%.

Conclusion

In answer to the question how big a problem is rhinosinusitis in patients with CF, the answer radiologically would be 100%, as all CF patients will reveal changes both on plain radiographs and on CT. The former are of little value in the assessment of sinus disease.

Control of upper respiratory tract infection may be an important factor in preventing reinfection of the lower respiratory tract and this is probably of particular significance in those patients undergoing heart-lung transplantation. Endoscopic sinus surgery provides a minimally-invasive surgical approach for the management of nasal polyposis and chronic sinus disease and although the results in CF patients are disappointing when compared with non-cystic patients, they are at least comparable to more aggressive forms of surgery.

Postoperative bleeding is more likely in CF patients and requires pre-operative assessment. Postoperative pain relief should be considered carefully to prevent intestinal obstruction.

The mainstay of treatment for this group remains medical with antibiotic and anti-inflammatory drugs, while minimally invasive surgery plays a useful adjunctive role. Many patients who have undergone 'conventional' surgery in the past will find the lack of nasal packing and no external scars combined with a short hospital stay a less daunting proposition.

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