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REVIEW

Nasal and sinus disease in cystic fibrosis

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Summary Paranasal and sinus disease is present in almost every patient with cystic fibrosis. However, symptoms are rarely reported. Some aspects including polyposis and microbial flora change with patient age. Endoscopy and computerised tomography have broadened our understanding of how this disease affects the sinuses, including an increased recognition of polyposis than previously thought and identification of several disease specific entities such as abnormalities of the lateral nasal wall and uncinate process. Few randomised, controlled trials evaluating medical or surgical treatments of CF sinus disease exist. Sinus surgery may provide some benefit, though there are no established selection criteria for appropriate candidates. A link between sinus disease and lower respiratory tract function may contribute to general health and survival following lung transplantation. Complications of sinonasal disease in CF are rare and include mucoceles and periorbital abscesses.

Although nasal and sinus disease is almost universal in cystic fibrosis (CF), somewhat surprisingly the prevalence of otitis media appears to be no greater than in an age-matched general population.[1] Because nasal and sinus disease usually coexist, we will refer to this as 'sinonasal disease' throughout this manuscript. Sinonasal disease, including polyposis, may be the presenting symptom of CF. Mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene in CF carriers (parents of patients with CF) appear to be independently associated with a higher prevalence of sinonasal disease; a total of 36% of carriers reported chronic rhinosinusitis compared with 13–14% in the general population.[2]

Since the mucosa of the upper respiratory tract, including the sinuses, and the mucosa of the lower respiratory tract are similar, disease may be similar in both locations or sinonasal disease could influence the severity of pulmonary disease. This view of the 'universal airway' has been demonstrated in patients with several pulmonary conditions, such as asthma and chronic obstructive pulmonary disease. In these diseases, an improvement in sinus health is reflected by an improvement in the lower airway disease.[3,4] This has not been well studied in CF but the implications of this relationship combined with increasing lifespan make an understanding of sinonasal disease important to the care of these patients.

CLINICAL PRESENTATION OF SINONASAL DISEASE IN CF

Despite significant disease, less than 10% of CF patients report sinonasal symptoms.[5] In a survey of consecutive paediatric patients at a CF centre, the average subject answered 'no problem' to 'mild to slight problem' for most

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sinonasal symptoms.[6] Even when they did complain, the complaint did not correlate with disease severity seen on endoscopic examination or on computed tomography (CT) .[5] The lack of complaints may be secondary to the chronicity of the symptoms or the overshadowing of these problems by more troublesome symptoms in the lung or gastrointestinal tract. The paucity of complaints has made it difficult to assess the outcome of sinus treatments, medical or surgical, as therapeutic outcome assessments usually rely on symptom scores.

One of the most common complaints is unilateral or bilateral nasal obstruction, which is present in up to 81.3% of patients.[7] Rhinorrhoea is reported to be present in over half of CF subjects.[8] Daily headache and anosmia are a problem for, at most, 51% and 27% of subjects, respectively.[8] Other symptoms include morning cough, halitosis and frequent throat clearing. Symptoms for those with polyps are the same as for those without, although the incidence of hyposmia may be higher in the group with polyps.[9] Approximately one-third of older persons with CF have broadening of the nasal bridge. Although most CF patients had normal nasal mucosa, mucopurulent material seemed to be present in the anterior nasal cavity of one-third of patients.[10] Cobblestoning of the posterior pharynx and adenoid hypertrophy could also be seen.

Endoscopic sinus exams on CF patients are almost always abnormal.[7,8] Among 248 CF patients referred to an otolaryngology clinic, congestion of the turbinates was noted in 88%, and most had abnormalities of the lateral nasal wall. One-third of patients with a mean age of 3 years had nasal polyposis and 15% had bulging of the lateral nasal wall. A total of 25% of older patients (mean age of 17 years) had an isolated prominent uncinate process.

NASAL POLYPOSIS IN CF

The reported prevalence of nasal polyposis in CF patients varies from 7% to 48%.[11] This variation may be accounted for by differences in subject populations or diagnostic methods. Initial reports of polyp prevalence in CF ranged from 6.7% to 26.7%, however diagnosis was made only by physical exam of the external nares.[1,10,12] These early studies were performed primarily in a younger population. Most polyps in CF are small[13,14] and over 60% are not visible outside the middle meatus.[13,15] Endoscopy significantly improves detection of nasal polyps to 33–56.5% (Fig. 1).[7–9,15–17] In 1995, it was reported that 51% of children at a CF clinic (mean age 11.9 years) had nasal polyps on endoscopy and that the mean age of those with polyps was significantly older than those without polyps.[7] For CF patients, the risk of developing polyps may increase with age.

Children with CF who have visible nasal polyps or recurrent polyposis have less severe lung disease (as measured by pulmonary function) than age-matched peers.[18] We speculate that polyps represent a proliferative airway repair mechanism and thus the absence of nasal polyps in patients with more severe disease may represent replacement of normal tissue with scarring or 'remodelling' with associated loss of normal repair capacity (Fig. 2).

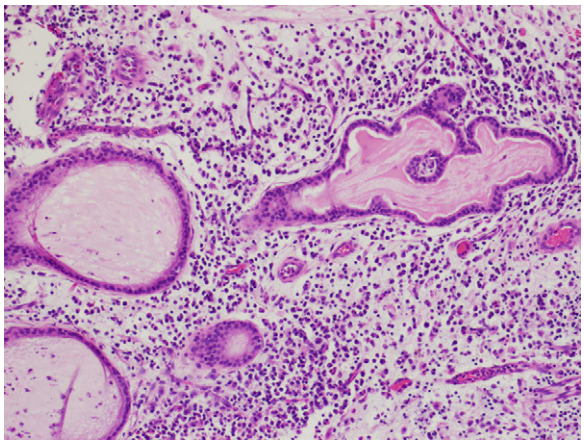

Figure 1 Sinus tissue from a cystic fibrosis patient with markedly dilated glands filled with eosinophilic mucus and submucosa with predominantly chronic inflammatory cells (plasma cells and lymphocytes) with and less frequent neutrophils. (H&E, original magnification 100×).

COMPUTED TOMOGRAPHY OF CF SINONASAL DISEASE

Sinus computed tomography (CT) provides great detail of bony and soft tissue anatomy and has replaced planar radiographs for the diagnosis of sinus disease. Several CT sinonasal anomalies are characteristic in the CF population, such as bulging or displacement of the lateral nasal wall and demineralisation of the uncinate process.[7,8,19,20]

Hypoplasia or aplasia of the paranasal sinuses has been uniformly reported in patients with CF.[20] Minimal development of frontal and sphenoid sinuses has been reported

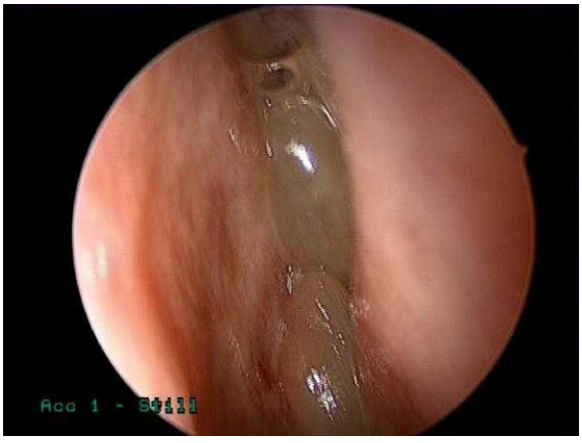

Figure 2 Typical intranasal examination of a cystic fibrosis patient with nasal polyps.