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Endoscopic and CT-scan evaluation of rhinosinusitis in cystic fibrosis*

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SUMMARY

In order to obtain an accurate evaluation of nasal and paranasal sinus disease in cystic fibrosis patients, 75 patients with a proven cystic fibrosis diagnosis have been investigated using the following standard techniques: questionnaire, ENT examination, endoscopical examination, sinus X-rays, and in 31% of the cases a CT scan. The analysis of results shows that nasal obstruction is the most frequent symptom (32%) and that nasal polyps are present in 43% of the cases. Endoscopic examination seems to be of great interest in giving a more accurate picture of nasal and paranasal sinus disease. Analysis of CT scan images leads the authors to describe a new and specific entity in nasal and paranasal sinus disease in cystic fibrosis patients: the pseudomucocele. In this report, the authors discuss the various aspects of pseudomucocele, attaching particular importance to the CT scan results.

Key words: sinusitis, nasal polyps, cystic fibrosis, endoscopy, CT-scan, pseudomucocele

INTRODUCTION

Rhinosinusitis is a common feature in children and adults with cystic fibrosis (CF). If sinus disease does not play a key role in the life expectancy of these patients, it most certainly increases the morbidity of their illness, with a deterioration of their quality of life, of their general state of health and perhaps with a worsening of their bronchopulmonary disease (Crockett et al., 1987). Although pulmonary and gastrointestinal disease remain the most preoccupying features, in our opinion rhinosinusitis, with its effects on everyday life, should be taken into greater account in CF.

In order to evaluate the impact and extent of rhinosinusitis in CF patients, the nasal and paranasal sinus (NPS) condition of 75 patients with a positive sweat test was studied. These patients, whether an ENT problem was suspected or not, were systematically referred to our ENT out-patient clinic by the different teams taking care of CF patients in the Necker Enfants-Malades Hospital at Paris.

PATIENTS AND METHODS

Seventy-five patients with a proven diagnosis of CF were evaluated over a period dating from November 1990 to October 1992, with clinical, endoscopical and radiological examination of their nose and paranasal sinuses and with, where necessary, a computed tomographic (CT) scan examination.

History and symptoms of NPS disease for all patients were recorded using a standardized questionnaire. Questions were related to functional signs (nasal obstruction, headache, purulent rhinorrhoea and olfactory troubles) and to the subjective relation established by the patient or his parents, between NPS disease exacerbation and fever, headache attacks or bronchopulmonary exacerbation.

A standard ENT examination was made and if possible an endoscopic examination of the nose and meatus was performed.

Endoscopic investigation was conducted under local anaesthesia using a rigid panoramic view endoscope (30°-view angle, 4-mm diameter Storz endoscope) or, in the youngest patients, a flexible 4-mm endoscope (Olympus). For each patient we recorded the presence and localization of the following features: inflammation and/or oedema, nasal polyps and purulent secretions. We also recorded the anatomical extension of NP using the following classification: (1) stage I: presence of one or more small NP in the middle meatus; (2) stage II: presence of

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NP reaching but not protruding beyond the middle turbinate's inferior rim; (3) stage III: NP protruding in the nasal fossa.

Initially the intention was to obtain systematic X-rays (in Waters' and Caldwell's incidence) for each patient but, the discriminative value of these exams being rather poor, this aspect of the study was abandoned (only 40 cases underwent X-rays).

Axial and coronal CT scans were performed, mostly using the limited-slice CT method (Gross et al., 1991), systematically on all patients registered on a pulmonary or heart-pulmonary transplant list, and also on patients whose NPS disease was considered severe enough to raise the question of NPS surgery.

In a number of cases, we took some bacteriological samples in the middle meatus under endoscopic control.

RESULTS

Sex and age

The mean age of our population was 11.9 years, ranging from 13 months to 31 years. The sex ratio was 59% females and 41% males.

Symptoms

Nasal obstruction was said to be present to a certain degree by 61 patients (81.3%) and was considered to be permanent and a real source of discomfort by 24 patients (32%). Permanent purulent rhinorrhoea was present in 15 cases (20%). Olfactory troubles were not easily analyzed in the youngest patients, but only five of all patients (6.6%) reported having completely lost their sense of smell. Headaches (of frontal localisation in almost all cases) were a daily occurrence in 17 cases (22.6%). Visible facial deformation was found in two children (5 and 6 years old), who had a noticeable enlargement of the nasal root and telorbitism.

Clinical subjective correlation

Twenty-six patients (34.6%) found a clear relationship between exacerbation of their NPS disease and their bronchopulmonary surinfections.

Headaches increased dramatically during NPS disease exacerbation in 19 patients (25.3%), and in 11 patients (14.6%) this exacerbation was accompanied by moderate fever.

Endoscopic examination

An endoscopic examination of the nose was carried out in 49 cases (65.3%). In 23 cases (30.6%) nasal endoscopy was not performed for technical reasons and in only three cases (4%) did the patient refuse to undergo the examination. All the endoscopic examinations were performed easily, causing no apparent pain or discomfort for the patient. Of the 49 endoscopic examinations, only nine (18.3%) were found to be normal and 40 (81.7%) abnormal.

Nasal polyps (NP) at clinical and/or endoscopical examination

NP were present in 32 of the 75 cases (43.7%) and were detected in 25 of the 49 patients (51%) who underwent nasal endoscopy. Patients having NP were male in 12 cases (37.5%) and female in 20 cases (62.5%). The mean age of patients with NP was 13.5 years with a minimum of four years and a maximum of 31 years. NP localization and extension are summarized in Table 1.

Table 1. Clinical features of nasal polyps in 32 CF patients in whom polyps were observed among 75 CF patients (43.7%); number (n) and percentage (%) of patients for each localization and stage.

	localization		stage	
	unilateral	bilateral	I-II	III
n	17	15	28	4
%	53.1	46.8	87.5	12.5

Nasal mucosa oedema or inflammation, purulent secretions

Oedema (reddish and bulging aspect) and/or inflammation (red colour and bleeding easily) of the nasal mucosa was found in 27 of the 75 patients (36%). In 88.3% of the cases, these findings were limited to the middle meatus, and in 11.7% of cases they affected the whole nasal fossa. Purulent secretions were found in 36% of the cases and were always detected in the middle meatus, never in the sphenoethmoidal recess. Bacteriological samples were taken in only six cases: one culture was negative, but the five others revealed the presence of *Pseudomonas aeruginosa.*

X-rays

Systematic standard X-rays were obtained during the first year of the study but were later dropped. Whatever the symptoms, CT scan images or surgical findings we always recorded the same images: both the maxillary sinuses were invariably totally opaque, frontal sinuses were absent or poorly developed, and the ethmoidal sinuses were difficult to analyze. These non-instructive results led us to abandon this part of the study and in the future to demand a CT scan if images of the sinuses were really deemed necessary.

CT scan

A CT scan of the nasal fossa and paranasal sinuses was carried out in 23 cases (30.6%) with axial and coronal sections. Images in the nasofrontal duct plane seemed difficult to obtain in these mainly young patients whose excessive bronchopulmonary secretions led to problems in maintaining the position required. All patients presented an opacity of the maxillary sinus (100%), while 78.2% of patients presented an opacity of the anterior ethmoidal sinus, 82.6% an opacity of the posterior ethmoidal sinus and 97.3% an opacity of the sphenoidal sinus (Table 2). In most cases the sinuses were opaque in a bilateral fashion (Table 2).

Table 2. Localization with its unilateral (u) or bilateral (b) feature of sinus opacities observed in 23 CF patients on CT-scan; number (n) and percentage (%) of patients for each localization.

	maxillary sinus	anterior ethmoidal sinus posterior	ethmoidal sinus	sphenoidal sinus
n	23 (u=0; b=23)	18 (u=0; b=18)	19 (u=2; b=17)	22 (u=4; b=18)
%	100	78.2	82.6	97.3

On analysis, we found the usual image of homogenous opacities partially or wholly filling the sinus cavities. These opacities may of course be just as well related to mucosal oedema as to polyps and/or secretions. Another image (Figures 1 and 2), consisting of a two-tone opacity (heterogeneous hyperdense centre with relative hypodense surroundings) with a tendency to expand (causing the pressing back or even osteolysis of the sinus wall), was also found in a large number of patients. This two-tone opacity was found in 86.95% of all patients with a CT scan. When this opacity was detected, it was affecting the maxillary sinus in 100% of cases, the ethmoidal sinus in 55% and the sphenoidal sinus in 60%. In the patients presenting this image who underwent surgical intervention, we found under endoscopic examination a bulging aspect of the nasosinus wall. This aspect had been underestimated during the previous endoscopical examination performed in the out-patient clinic. Because these images broadly reproduce the negative of a typical CT-scan image of mucocele, we named it "pseudomucocele".

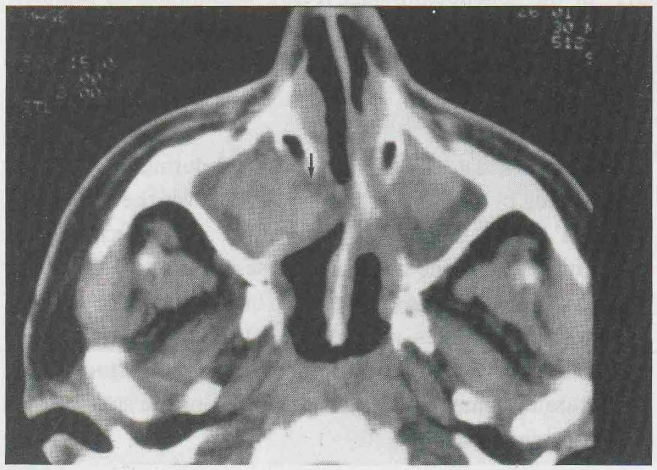

Figure 1. Pseudomucocele aspect of both maxillary sinuses on an axial CT-scan of a 10-year-old girl. Note the erosion of the nasosinus wall (arrow).

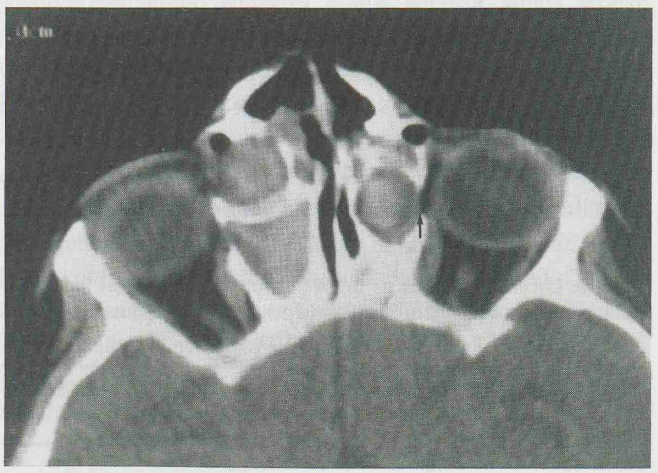

Figure 2. Pseudomucocele aspect of the anterior ethmoid cells on an axial CT-scan of a 5-year-old boy. Note the erosion of the ethmoid-orbital wall (arrow) and the enlargement of the nasal root.

DISCUSSION

Chronic rhinosinusitis is a common feature in CF patients, and well documented in medical literature (Shwachman et al., 1962, 1977; Drake-Lee and Morgann, 1989; Denoyelle et al., 1990; Kerrebijn et al., 1992). Generally, NPS disease exists either as chronic purulent sinusitis or as nasal polyposis, or both. If the symptoms of NPS disease are well documented, little information is available concerning their prevalence in CF patients or their effects on the daily life of these patients. In this study, 75 patients suffering from CF whatever their age, degree of illness and nasal complaint, were investigated systematically in order to clarify NPS disease symptoms and lesions in CF. Nasal obstruction was the major symptom seriously affecting patients in one third of all cases. Purulent nasal discharge and headaches were frequently reported, but were a real source of permanent discomfort in only 20% of patients. The proportion of patients having real trouble with their nasal symptoms is higher than that previously noted in other studies (Schwachman et al., 1977; Denoyelle et al., 1990) and close to that recently reported by Cuyler (1992) and Brihaye et al. (1994). Nearly 35% of the patients established a link between NPS disease and pulmonary disease exacerbations. The way this figure was obtained is too subjective to establish a cause-effect relationship between NPS disease and pulmonary disease exacerbations, a relationship, moreover, that remains a subject of controversy (Drake-Lee and Morgan, 1989; Denoyelle et al., 1990; Umetsu et al., 1990).

Figures for nasal polyposis in CF patients vary widely from one study to another from 8–44% (Shwachmann et al., 1962, 1977; Denoyelle et al., 1990; Kerrebijn et al., 1992; Brihaye et al., 1994). There are two factors which probably explain these variations: firstly, some of these studies have been reported 20 or 30 years ago when life expectancy for CF patients and means of investigation were very different, and secondly there are differences in patient age from one study to another. In the present study, NP were present in 43.7% of patients, a figure which is close to those published in recent studies (Kerrebijn et al., 1992; Brihaye et al., 1994). The mean age of patients with NP (13.5 years) was significally different ($p < 0.05$, unpaired t-test) from the mean age of patients whithout NP (10.8 years), suggesting that NP occur more frequently in older CF patients (Shwachmann et al., 1977; Drake-Lee and Morgan, 1989; Brihaye et al., 1994). However, we often observed NP in young CF children. NP were not more frequent in male patients than in female patients as reported by others (Drake-Lee and Morgan, 1989; Brihaye et al., 1994). Surprisingly, NP were detected unilaterally in 53% of the cases, in accordance with another study (Denoyelle et al., 1990) but in contrast with others (Reily et al., 1985; Brihaye et al., 1994). Perhaps a more accurate examination with middle turbinate luxation would reduce this figure. We found a large invasion of nasal fossa by NP in only 9% of cases, most cases being stage I or especially stage II polyposis. We were expecting a higher score of stage III polyposis, as reported by others (Brihaye et al., 1994), which would have been more consistent with the usual notion of NP aggressiveness in CF.

The second aim of our study was to evaluate whether modern tools of NPS investigation (CT scan and endoscopy) could contribute to a more accurate evaluation of NPS disease in CF. Endoscopic examination presented us with no problems, irrespective of patient age, and only three children refused it. Endoscopic examination allows a better view of the whole nasal fossa and meatus (Brihaye et al., 1994; Mackay and Djazaeri, 1994) and has shown a higher frequency of NP than classical anterior rhinoscopy. Endoscopic signs of pseudomucocele (middle projection of the nasomaxillary wall) were initially underestimated as, before recording CT scans results, they were not actively researched: we only noticed an occasional narrowing of the nasal fossa. The middle projection of the nasomaxillary wall in CF patients occurred in 12% of the cases in a recent study (Brihaye et al., 1994). It is now clear that the aspect of the nasomaxillary wall must be evaluated systematically, as proposed by others (Brihaye et al., 1994). Not surprisingly, CT scan was of great interest in appreciating NPS invasion by NP, oedema or purulent secretions. Standard X-rays were of little relevance, as already suggested by many authors (Crockett et al., 1987; Drake-Lee and Morgan, 1989; Denoyelle et al., 1990; Umetsu et al., 1990; Kerrebijn et al., 1992; Mackay and Djazaeri, 1994). We found high rates of sinus opacities on CT scans, as recently reported by Brihaye et al. (1994), but most of these exams were performed on patients who had NPS disease severe enough to discuss a surgical treatment. The most interesting fact is that CT-scan analysis led us to describe this new pathological sinus entity which we have named pseudomucocele. Recently, some authors (Brihaye et al., 1994; Mackay and Djazaeri, 1994) reported similar pathological CT-scan images in CF patients, images for which the most likely diagnosis was a mucopyosinusitis (Brihaye et al., 1994). The results of the present study do agree with these findings, but we propose to distinguish these particular pathological changes by the term pseudomucocele. Neither in medical literature nor in our own experience have we noted such images in non-CF NPS disease. We think this strongly suggests that pseudomucocele is a specific entity of NPS disease in CF. In most cases, when the maxillary sinus was filled with a pseudomueocele, a rather extensive bone destruction of the nasomaxillary wall was observed. This was not the case in other sinus cavities, except in one case where the orbital wall was erroded (pseudomucocele of an anterior ethmoidal cell; Figure 2). The destruction of the bony walls in chronic sinusitis in CF patients could be the result of osteitis (Mackay and Djazaeri, 1994) or of pressure necrosis (Brihaye et al., 1994). In the patients that were surgically managed, exposure of the sinus cavities affected by a pseudomucocele showed a thick inflammatory and resistant shell following the contours of the sinus walls (corresponding to the hypodense surround seen on CT scan). The opening of this shell allowed difficult suction of its contents, consisting of thick and chewing-gum-like mucopus (corresponding to the heterogeneous hyperdense centre seen on CT scan). When referring back to symptoms initially reported by patients with pseudomucocele, we found the usual complaints including nasal obstruction, purulent rhinorrhoea and headaches. In one case, a 5-year-old boy, who had numerous and extremely expanded pseudomucoceles (Figure 2), there was a considerable enlargement of the nasal root and telorbitism. Histopathological examination of the pseudomucocele showed only intense inflammatory reaction with glandular hyperplasia. The physiopathological mechanisms of pseudomucocele still remain unclear and for a better understanding require further investigation. Sinus ventilation and drainage troubles, in relation with mucus viscosity impairment and in association with chronic local infection, are certainly involved. Concerning treatment, only a few patients (n=7) with pseudomucocele underwent functional endoscopic sinus surgery. Surgery was decided when the severity and frequency of symptoms (nasal obstruction, purulent rhinorrhoea and headache) were not sufficiently improved by medical therapy (local and general antibiotics, nasal lavage with saline serum, and sometimes local and general corticosteroids) and seemed to be strong enough to affect the quality of life (sleep, work, and eating), the general state of health (weight stagnation) and/or the frequency of bronchopulmonary exacerbations of the disease. In terms of symptoms, endoscopic findings and CT-scan findings, improvement was noted for all of them, but the follow-up period remains too short (<2 years) to draw any conclusion.

NPS disease is a common feature in CF patients, whose frequency is presently on the increase, possibly because the life-expectancy of these patients is improving and also because the ENT surgeon is more involved in CF patient care. Endoscopic examination and CT-scan seem of great interest for an accurate evaluation of NPS disease in CF patients. Nasal polyposis, which seems especially frequent (44%), and chronic purulent sinusitis are the two clinico-pathological entities usually described in CF. CT scans of sinus cavities led us to describe a new entity: the pseudomucocele. The pseudomucocele would seem to be frequent in and specific to CF patients. Further investigations are necessary for a better characterization of the clinical, developmental and physiopathological profile of such lesions.

REFERENCES

1. Brihaye P, Clement PAR, Dab I, Desprechin B (1994) Pathological changes of the lateral nasal wall in patients with cystic fibrosis (mucoviscodosis). Int J Pediatr Otolaryngol 28: 141–147.
2. Crockett DM, McGill TJ, Healy GB, Friedman EM, Salkeld LJ (1987) Nasal and paranasal sinus surgery in patients with cystic fibrosis. Ann Otol Rhinol Laryngol 96: 367–372.
3. Cuyler JP (1992) Follow-up of endoscopic sinus surgery on children with cystic fibrosis. Arch Otolaryngol Head Neck Surg 118: 505–506.
4. Denoyelle F, Garabedian EN, Sardet A, Tournier G, Bellity A (1990) Atteinte de la sphere O.R.L. dans la mucoviscidose. Ann Oto-Laryng (Paris) 107: 200–204.
5. Drake-Lee AB, Morgan DW (1989) Nasal polyps and sinusitis in children with cystic fibrosis. J Laryngol Otol 103: 753–755.
6. Gross GW, McGeady SJ, Kerut T, Ehrlich SM (1991) Limited-slice CT in the evaluation of paranasal sinus disease in children. AJR 156: 367–369.
7. Kerrebijn JDF, Poublon RML, Overbeek SE (1992) Nasal and paranasal disease in adult cystic fibrosis patients. Eur Respir J 5: 1239–1242.
8. Mackay IS, Djazaeri B (1994) Chronic sinusitis in cystic fibrosis. J R Soc Med 87 (Suppl 21): 17–19.

9. Reily JS, Kenna MA, Stool SE, Bluestone CD (1985) Nasal surgery in children with cystic fibrosis: Complications and risk management. Laryngoscope 95: 1461–1463.
10. Shwachman H, Kukychi I, Mueller H, Flake C (1962) Nasal polyps in patients with cystic fibrosis. Pediatrics 30: 389–401.
11. Shwachman H, Kowalski M, Khaw K-T (1977) Cystic fibrosis: A new outlook. Medicine 56: 129–149.
12. Umetsu DT, Moss RB, King VV, Lewiston NJ (1990) Sinus disease in patients with severe cystic fibrosis: Relation to pulmonary exacerbation. Lancet 335: 1077–1078.

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