

## Nasosinusal Polyposis Revealing Primary Ciliary Dyskinesia: A Case Report

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### Abstract

The histological and functional compositions of lung and nasal tissue contain similarities as well as distinct differences. Certain nasosinusal and bronchial involvement are associated with many diseases, among them primary ciliary dyskinesia where bronchial involvement in the form of bronchiectasis and nasosinusal involvement in the form of chronic rhinosinusitis are both often present. We report the case of a 35-year-old patient with nasosinusal polyposis revealing primary ciliary dyskinesia.

**Keywords:** Nasosinusal polyposis, Rhinitis, Sinusitis, Bronchiectasis

### Introduction

While the upper and lower respiratory tracts are anatomically distinct, it appears that in many pathological situations their involvement is concomitant, as is the case with rhinitis and rhinosinusitis associated with asthma. But that of nasosinusal polyposis and dilation of the bronchi remains rare. The concept of rhinobronchitis introduced the idea that the upper and lower airways are in fact a single airway and that diseases affect the entire respiratory system [1]. Numerous epidemiological studies have examined and confirmed this association, giving rise to the concept of "one airway, one disease" The clearest example of this association is rhinitis and asthma [3]; most asthmatics have both conditions, and treatment for rhinitis may be beneficial for asthma [3,4].

Primary ciliary dyskinesia (PCD) is a disease that combines pulmonary and otolaryngologic symptoms. Early diagnosis helps prevent bronchiectasis. We report the case of a patient hospitalized for treatment of nasosinusal polyposis subsequently revealing primary ciliary dyskinesia [5].

### Case report:

This is a 35-year-old patient, from a consanguineous marriage (first cousin first degree), asthmatic not followed, the beginning of the symptomatology goes back to his first years of life by the installation of bilateral nasal tilting obstruction associated with clear anterior rhinorrhea and sometimes purulent, without a notion of epistaxis, as well as an almost constant productive cough, aggravated in fits during episodes of superinfection. The course is marked by worsening nasal obstruction and symptoms, which remain resistant to treatment, with the onset of permanent anosmia, and closed rhinolalia. The examination finds an enlargement of the nasal pyramid, nasal flow absents on both sides with a negative Cottle's sign. On nasal endoscopic examination, we note the presence of translucent polyps reaching to the lower edge of the inferior turbinate on both sides (stage III).

A nasosinusal and thoracic tomodensitometry, with an injection of the contrast product for the complement of the lesion assessment, was requested and objectified at the facial level, and opacity of tissue density filling the anteroposterior sinus complexes on both sides, with thinning of the frame. the bone of the sinus walls (Figure 1). Otherwise, at the thoracic level, an aspect of dilation of the lower polar bronchi more important on the right

was noted (Figure 2).



**Figure 1:** Image of the nasosinus CT scan showing a filling of the anteroposterior sinus complexes, with agenesis of the frontal sinus.



**Figure 2:** Lung CT image showing bilateral diffuse bronchial dilation lesions.

Given the combination of symptoms, a specialist opinion in pulmonology was requested or a nitrogen concentration dosage was requested to detect possible mucoviscidosis or primary ciliary dyskinesia and which returned significantly lowered.

The patient was operated on for his polyposis by endoscopic route or multiple histological cross-sections of the nasal mucosa were performed showing a greatly reduced ciliary density, and the diagnosis of primary ciliary dyskinesia was retained. The evolution was good with close multidisciplinary follow-up, and antibiotics and corticosteroid coverage were introduced.

### Discussion:

In the early years, patients with PCD present with otologic and rhinological sinus signs that are often prominent to such an extent that the absence of upper airway involvement makes the diagnosis of PCD very unlikely. Patients with PCD present also with early, constant rhinitis, sometimes noted as early as the neonatal period [5,7]. In children, otitis media is chronic, almost constant, improves with age, and can disappear in adults. Secondary

infections with perforation of the eardrum and otorrhea are rare after the age of three to four years. Also, the occurrence of otorrhea on trans-tympanic aerators is almost constant and should suggest the diagnosis of PCD.

Chronic sinusitis from PCD is often present in infancy, purulent, diffuse, often debilitating, and difficult to treat, it progresses to nasal polyposis in at least half of cases [6]. On computed tomography, there are diffuse and bilateral sinus opacities confirming, with endoscopy, the existence of general pathology of the lining of the nose and sinuses. In older children and adults, this edema-purulent pan sinusitis can be associated with hypo or agenesis of the frontal sinuses, as in the case of our patient where the frontal sinuses were completely absent.

The diagnosis will also be evoked by the presence of a reverse rotation of the viscera (situs inversus), present in approximately 50% of cases, thus achieving Kartagener's syndrome (incidence 1/32000), defined by the triad bronchiectasis, chronic sinusitis, and situs inversus [8]. As it is a rare disease that is transmitted classically in an autosomal recessive mode and following this mode of transmission, the frequency of affected individuals is much higher in populations where there is a high degree of consanguinity, such as the case of our patient who came from a co-blood marriage (first cousins first degree). It is, therefore, the combination of upper and lower respiratory infections that will suggest the diagnosis of ciliary dyskinesia, especially as there is consanguinity, a family history, a history of respiratory distress, and/or a situs inversus [5].

### Conclusion:

The respiratory mucosa must be considered as a whole, both from a diagnostic and therapeutic point of view. Indeed, the management of double involvement, ENT, and bronchial, is generally possible without increasing the prescription and the treatment of sinus involvement improves bronchial hyperreactivity. The close collaboration between the ENT doctor and the pulmonologist makes it possible to adopt a concerted global treatment strategy in the face of an inflammatory disease which often affects all of the respiratory tracts.

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