

CASE REPORT

Bilateral Antrochoanal Polyp with Diabetes: A Rare Presentation of Cystic Fibrosis

Manish Gupta, Monica Gupta

Assistant Professor, Department of Otolaryngology, Gian Sagar Medical College and Hospital, Ramnagar, Banur, Punjab, India

Correspondence: Manish Gupta, Assistant Professor, 1217, Government Medical College and Hospital Campus, Sector 32-B Chandigarh-160030, India, e-mail: manishgupta1217@gmail.com

Abstract

Bilateral antrochoanal (AC) polyps have been rarely reported in english literature. Here we report the first case of an adolescent male with bilateral AC polyps, cystic fibrosis and diabetes. The case was managed successfully by functional endoscopic sinus surgery (FESS). The patient has been under follow-up for the last six months with no signs of recurrence.

Keywords: Nasal polyps, Antrochoanal, Cystic fibrosis.

INTRODUCTION

Antrochoanal (AC) polyps are usually unilateral and occur more commonly in children. There have been only four case reports of bilateral AC polyps in english literature.¹⁻⁴ None of them have been associated with cystic fibrosis so far. Here we report a case of adolescent male with bilateral AC polyps with cystic fibrosis and diabetes. The case was managed successfully by functional endoscopic sinus surgery (FESS). The patient has been under close follow-up and there has been no recurrence in last six months.

Cystic fibrosis (CF) is characterized by chronic bronchopulmonary infection, often accompanied by airflow obstruction, malabsorption, failure to thrive and a high sweat sodium concentration. The signs and symptoms typically occur in childhood, but about 5% of patients are first diagnosed in adulthood. CF has also been implicated in the etiology of ethmoidal nasal polyps in children. Paranasal sinus disease is present in almost every patient with cystic fibrosis. However, symptoms are rarely reported.⁵ Stern et al⁶ recommend that children less than 10 years of age presenting with nasal polyps should be screened for CF.

CASE REPORT

A 15-year-old boy presented to the out patient department complaining of gradual onset, bilateral, progressive nasal obstruction and difficulty in breathing for one year. He also had recurrent nasal discharge and occasional headaches. There was no history of bleeding from nose.

On anterior rhinoscopy right side single, large, pale polyp was seen coming into the vestibule with mucoid discharge. On posterior rhinoscopy large polypoidal mass coming from



Fig. 1: Clinical photo showing polyp protruding into the right nostril

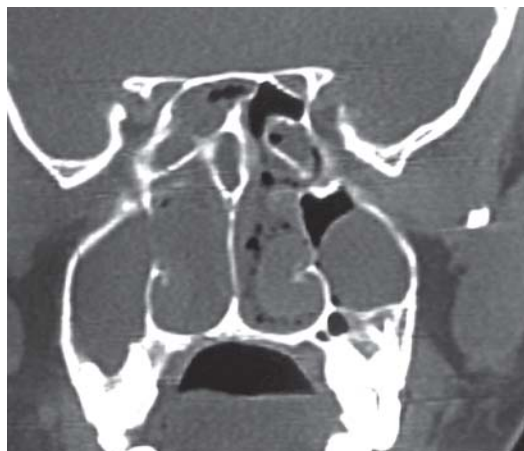


Fig. 2: Coronal cut of contrast enhanced computed tomogram (CECT) of the patient showing opacification of bilateral maxillary sinus by polyp and its protrusion into nasal cavity through the widened ostium

right choana could be seen (Fig. 1) occupying whole nasopharynx along with small polyp from left choana.

Noncontrast computerized tomogram of the nose and paranasal sinus was done with coronal cuts. It revealed near total opacification of bilateral maxillary sinuses (Fig. 2) with masses extending into the nasal cavities through both the osteomeatal complexes (which were slightly widened) and then protruding into choana.

All routine blood and urine investigations were done. The patient's random blood glucose was 233 mg/dl. Overnight fasting and postprandial blood glucose values were 130 mg/dl and 208 respectively. His glycosylated hemoglobin was 7.5 gm/dl. Patient was referred to the physician for diabetes management and was started on insulin therapy. The pilocarpine-induced sweat test revealed sweat chloride concentrations to be 65 mmol/l and 64 mmol/l, in two consecutive samples. As the genotyping was not available in our set-up, hence diagnosis of cystic fibrosis was based on clinical and laboratory values. The patient was evaluated for the microvascular complications of diabetes, and revealed no abnormality. There was no evidence of steatorrhea, or bronchopulmonary infection.

After adequate diabetes control, patient was taken up for surgery. Functional endoscopic sinus surgery was performed using 30° Hopkins rod endoscope and polyps were removed from bilateral nasal cavities and maxillary antrum. Bilateral middle meatus were packed with merocele which was removed after 48 hours. The histopathological examination revealed polypoid mass lined by pseudostratified columnar epithelium, loose stroma with inflammatory cells like plasma cells, lymphocytes and polymorphs. The postoperative period was uneventful and there is no recurrence in last 6 months follow-up.

DISCUSSION

Cystic fibrosis is the most common lethal autosomal recessive disease.⁷ Cystic Fibrosis (CF) results from mutations in the gene that encodes the CF transmembrane conductance regulator (CFTR) protein located on chromosome 7.⁸ This gene encodes for a protein that functions as a cyclic adenosine monophosphate—regulated chloride channel. Abnormal function of the channel, results in aberrant conductance across the apical membrane of epithelial cells of ducts in a variety of organs (lung, pancreas, sweat gland, liver, nasal mucosa, salivary glands and colon).

The mutations in the CFTR gene fall into four major classes,⁹ classes I-III mutations are considered “severe,” as indexed by pancreatic insufficiency and high sweat sodium chloride (NaCl) values. Class IV mutations are “mild,” i.e. associated with pancreatic sufficiency and intermediate/normal sweat NaCl values.

The central hypothesis of CF airways pathophysiology is that the faulty regulation of Na⁺ absorption and inability to secrete Cl⁻ reduce the volume of liquid on airway surfaces; i.e, they are “dehydrated.” Both the thickening of mucus and the depletion of the periciliary liquid lead to adhesion of mucus to the airway surface. Mucus adhesion leads to a failure to clear mucus from the airways both by ciliary and airflow-dependent (cough) mechanisms.

The clinical manifestations of the disease include pancreatic enzyme deficiency with malabsorption, chronic progressive obstructive pulmonary disease, chronic pulmonary infection with *Staphylococcus aureus*, *Pseudomonas aeruginosa*, or both.⁷ The features suggestive of a diagnosis of cystic fibrosis have been tabulated (Table 1) according to various major and minor criterias.¹⁰ The otolaryngologic manifestations of CF include chronic sinusitis and nasal polyposis.⁷

The diagnosis of CF is based on repeated pilocarpine-induced sweat test (> 60 mmol/L of chloride in an adequate sample of sweat)⁷ In adolescents and adults sweat sodium is higher and the test less reliable.¹⁰ In case of repeated borderline measurements (40 to 60 mmol/L), clinical correlation or DNA genotyping is required for diagnosis.⁷ The differential diagnosis⁸ of raised electrolyte level in sweat are metabolic causes like cystic fibrosis, fucosidosis, mucopolysaccharidosis, hypothyroidism, vasopressin resistant diabetes insipidus, adrenal insufficiency, familial cholestasis and familial hypoparathyroidism. Raised levels may also be falsely reported due to partial evaporation of sample.

Nasal potential difference measurements (including responses to amiloride, chloride free solution and isoproterenol) may demonstrate abnormal CFTR function more reliably than the sweat test. Measuring the potential difference across the respiratory epithelium may be helpful, but invalid in the presence of upper respiratory tract infections and nasal polyps.¹⁰

Table 1: Features suggestive of a diagnosis of cystic fibrosis

Major criteria	Minor criteria
Bronchopulmonary infection	Nasal polyps*
Failure to thrive	Meconium ileus equivalent*
Malabsorption	Rectal prolapse
Meconium ileus	Intussusception
Family history	Biliary cirrhosis*
Azoospermia*	Pancreatitis*
	Peptic ulcer*
	Diabetes mellitus*
	Pneumothorax*

*More common in adolescents and adults than children

In difficult cases, genetic analysis can be performed. Although it can add important evidence, genotyping alone cannot establish the diagnosis or rule it out. At least 500 CFTR mutations associated with cystic fibrosis are known; the commercially available probes test for only 70.⁸ Although these 70 mutations can be used to identify more than 90% of all cystic fibrosis genes, failure to find two abnormal genes does not rule out the disease. In approximately 1% of those with the disease no abnormal gene can be found, and in about 18% more, only one abnormal gene will be identified.

The life expectancy of individuals with CF has dramatically increased over last few decades due to improvements in respiratory and nutritional therapy.¹¹ More than 41% patients of CF are now adults (>18 years) and 13% are past the age of 30. Adult diagnosed CF patients are less likely to present with gastrointestinal and respiratory symptoms than childhood diagnosed CF patients. Accordingly these patients have less CF related diabetes, low frequency of pancreatic insufficiency and quite subtle bronchiectasis.¹²

Diabetes mellitus occurs in approximately 10% cases of CF with onset generally after the age of 15 years.¹³ Thus it is seen in 9% of CF children, 26% of adolescents, 35% of adults age 20-29, and 43% of adults age 30 and older. Although cystic fibrosis related diabetes (CFRD) is a distinct entity, it shares features of both Type 1 and Type 2 diabetes mellitus. Microvascular complications (diabetic retinopathy, neuropathy, and nephropathy) have been reported in CFRD.¹⁴ However, unlike Type 1 and Type 2 diabetes, macrovascular complications like ischemic heart disease and stroke are not widely seen in CFRD, probably because of shorter life span.

Screening for CFRD with annual, fasting glucose monitoring should begin by age 14 for all patients with cystic fibrosis and pancreatic insufficiency.¹⁴ If the fasting blood glucose level is high (> 126 mg/dl) at two readings diabetes should be diagnosed. Hemoglobin A1c (HbA1c) is not recommended to screen for CFRD, as it may be normal in them due to increased erythrocyte turnover and insufficient hyperglycemia in CFRD.¹³ Insulin is the only medical therapy recommended for CFRD with fasting hyperglycemia. Calorie restriction is never appropriate in CF as these individuals are already underweight.

Nasal polyposis usually ethmoidal occurs in 6-36% of patients with cystic fibrosis most frequently between the age of 4 and 12 years.¹⁵ They are frequently multiple and bilateral and recur frequently. The histology of these polyps is the same as in patients without cystic fibrosis. In a study by Henriksson⁷ (2002) forty-four of the 111 CF patients

(39%) revealed ethmoidal nasal polyposis by the endoscopic examination. The majority showed polyposis of grade 1 (23%) and grade 2 (45%), i.e. polyps concealed in the middle meatus. Only one-third of the polyps were visible in the nasal cavity. No large polyps were found.

Killian¹⁰ in 1906 gave the first detailed description of antrochoanal polyp. AC polyps are benign lesions that arise from the mucosa of the maxillary sinus, grow into the nasal cavity and reach the choana, nasal obstruction being their main symptom.¹⁶ Most of them are unilateral in origin.¹⁷ Macroscopically, they have a cystic intramaxillary portion and a solid intranasal portion. Microscopically, they are similar to a maxillary cyst of the mucosa. Nasal endoscopy, computed tomography and magnetic resonance are the main diagnostic techniques.¹⁶ Radiographs of the paranasal sinuses may show completely opaque antrum. The lateral view may show the polyp in the postnasal space with a crescent of air shadow behind the mass.

Till date there have been only four case reports of bilateral AC polyps in english literature.¹⁻⁴ However, none of them have been associated with cystic fibrosis so far. Moreover we could not come across any case of bilateral antrochoanal polyps in cystic fibrosis on extensive search of all the electronic medical english databases. The present case is in all probability the first case report of bilateral Antrochoanal polyp in cystic fibrosis in english literature.

Medical or topical treatment is unsuccessful in AC polyps. Surgery is the treatment of choice. The simple intranasal polypectomy results in symptomatic recurrence in about 60% of patients within 18 months. It is necessary to remove both parts of the polyp. Caldwell-Luc antrostomy was advised to reduce the recurrence, but is associated with damage of the maxillary and dental growth center. Functional endoscopic sinus surgery (FESS) decreases the risk of polyp recurrence and allows removal of diseased sinus mucosa and polyp with minimal disruption of the normal anatomy. FESS has been well-tolerated by CF patients and provides good symptomatic relief.

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