RESEARCH PAPER

Non-Cystic Fibrosis Bronchiectasis in Children: Clinical Profile, Etiology and Outcome

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Objective: To describe clinical profile, etiology and outcome in children with non-cystic fibrosis bronchiectasis. **Methods:** A chart review of children diagnosed with non-cystic fibrosis bronchiectasis, attending pediatric chest clinic of tertiary care hospital. **Results:** The underlying cause was identified in 51 (63.8%) out of 80 children (mean age, 9.6 y). Common causes were post-infectious in 19 (23.8%), suspected primary ciliary dyskinesia in 12 (15%), and allergic bronchopulmonary aspergillosis in 6 (7.5%). One or more complications were observed in 76 (95%) patients; 14 (17.5%) children required surgery and 5 (11.1%) children died. **Conclusions:** Common causes of non-cystic fibrosis bronchiectasis are post infectious and primary ciliary dyskinesia. There is a need to create awareness about early diagnosis of bronchiectasis as it is often delayed.

Keywords: *Allergic bronchopulmonary aspergillosis, Chronic cough, Pneumonia, Suppurative lung disease.*

Bronchiectasis is not an uncommon condition in pediatric population. Cystic fibrosis (CF) is the most common cause of bronchiectasis in developed countries while in developing countries non-CF etiologies are more common [1,2]. Information on etiology of bronchiectasis from developing countries is scanty [3]. We present data on underlying causes, clinical features and outcome of non-CF bronchiectasis in Indian children diagnosed in pediatric chest clinic of a tertiary care hospital in Northern India.

METHODS

A chart review (from 2006 to 2013) of children diagnosed with non-CF bronchiectasis attending the pediatric chest clinic of a tertiary-care hospital was carried out.

Cases of bronchiectasis were identified by screening the clinical records and clinical details, investigations and course of illness were recorded in predesigned forms. Diagnosis of bronchiectasis was based on findings in high resolution computerized tomography (HRCT) of chest.

Post-infective cause was suspected when there was a history of measles, varicella, bronchiolitis or pneumonia. Post-tubercular bronchiectasis was diagnosed if a child had suffered from pulmonary tuberculosis (TB) in the past. Evidence of TB included some of the following: history of contact with adult TB patient in family, abnormal *X*-ray film (miliary shadows, cavity, mediastinal or hilar adenopathy), documentation of acid-fast bacilli (AFB) on sputum/gastric aspirates/broncho-alveolar lavage, positive Mantoux test and response to anti-tubercular treatment. Immune-deficiency was considered if there was multi-site recurrent infection along with supportive investigations like HIV ELISA, immunoglobulin profile, CD19, or NBT (Nitro blue tetrazolium) test. Diagnosis of hypogammaglobinemia was based on low levels of immunoglobulins. If CD19 were low along with hypogammaglobulinemia, a diagnosis of Bruton disease was made; else, a diagnosis of common variable immune deficiency was considered. A diagnosis of Allergic bronchopulmonary aspergillosis (ABPA) was made according to criteria laid down by International Society of Human and Animal Mycology (ISHAM) working group [4]. Primary ciliary dyskinesia (PCD) was suspected when patients who presented with history of recurrent sinusitis, otitis, and recurrent pneumonia and had Fractional exhaled nitric oxide (FENO) less than 10 ppb.

Diagnosis of repeated aspirations was made on the basis of clinical features (*e.g*. coughing/choking during feeding) with abnormal Technetium scan or barium swallow. Bronchiectasis due to foreign body was

diagnosed with history of foreign body aspiration in the past and documentation by imaging or bronchoscopy. With history of Steven Johnson syndrome (SJS) preceding onset of recurrent/persistent pulmonary symptoms and evidence of bronchiectasis on HRCT, a diagnosis of post-SJS bronchiectasis was considered. Diagnosis of asthma was made on the basis of clinical signs/symptoms, family history, and response to bronchodilator therapy. Diagnosis of malformation of airways was based on clinical features, imaging and bronchoscopic findings.

All children were followed up every 3-6 months. On follow-up visits, all children were examined, adherence to treatment including physiotherapy was checked, and spirometry and sputum examination (culture and sensitivity) were performed, whenever indicated. Complications and associated problems were also noted. Outcome was determined by course of illness during follow-up in terms of weight, height, need for surgery and death. During follow-up, cases were managed with medical treatment including antibiotic use in exacerbations, bronchodilators, steroids (oral/inhaled), antihistamines and chest physiotherapy. Patients who were not attending the clinic were contacted telephonically.

RESULTS

Eighty children (50 boys) with non-cystic fibrosis bronchiectasis were identified. The mean age of presentation was 9.6 (range 2-15) years; 62.5% were below 5 years of age. Clinical manifestations are shown in ***Table* I**. Clubbing was present in 58 (72.5%) children.

TABLE I SYMPTOMS AT THE TIME OF DIAGNOSIS IN CHILDREN WITH NON-CYSTIC FIBROSIS BRONCHIECTASIS (*N*=80)

Symptoms	*No. (%)*
Cough	77 (96.3)
Breathlessness	65 (81.3)
Expectoration	53 (66.3)
Fever	50 (62.5)
Wheezing	42 (52.5)
Repeated pneumonia	37 (46.3)
Chest pain	34 (42.5)
Sneezing	23 (28.8)
Repeated nasal discharge	19 (23.8)
Pain abdomen	13 (16.3)
Hemoptysis	13 (16.3)
Recurrent vomiting	08 (10.0)
Failure to thrive	08 (10.0)
History of ear discharge	04 (5.0)
Increase requirement of salt	01 (1.3)

An underlying etiology could be identified in 51 (63.8%) children. The commonest etiology was post-infectious (in 19 children). Details of underlying causes are given in ***Table* II.**

Sixty-two (77.5%) patients had at least 12 months of follow up with mean (SD) duration of follow-up as 21.1 (16.5) months. Five (11.1%) children died during follow-up. Significant proportion of children had morbidity such as poor growth in 61 (76.3%), pulmonary hemorrhage in 13 (16.2%), chronic hypoxemia in 8 (10%), and pulmonary hypertension in 3 (3.8%). Surgical intervention was needed in 14 (17.5%) children.

DISCUSSION

In our study, non-CF bronchiectasis was observed more commonly in boys and majority of children (62.5%) were below 5 years of age which was similar to other studies [2,5]. The clinical features were consistent with other studies [2,3,6-8]. We observed clubbing in 72.5% of children which is higher than earlier reports of 3-51%

TABLE II UNDERLYING CAUSES OF BRONCHIECTASIS IN CHILDREN WITH NON-CYSTIC FIBROSIS BRNCHIECTASIS (*N*=80)

Cause	*No. (%)*
Post-infective	19 (23.8)
Post tubercular*	8 (10.0)
Other infections	11 (13.8)
PCD (suspected)	12 (15)
ABPA	6 (7.5)
Malformations	3 (3.7)
Airway	2 (2.5)
Esophageal	1 (1.2)
Repeated aspiration	2 (2.5)
Primary immune-deficiency	5 (6.2)
CVID	2 (2.5)
Pan hypogammaglobulinemia	2 (2.5)
CGD	1 (1.2)
Foreign body	1 (1.2)
Steven Johnson's syndrome	1 (1.2)
HIV infection	1 (1.2)
Asthma	1 (1.2)
Cause not identified	29 (36.2)

**includes 3 children with reactivation of tuberculosis; CGD: Chronic granulomatous disease, CVID: Common variable immune deficiency; ABPA: Allergic bronchopulmonary aspergillosis; PCD: Primary ciliary dyskinesia.*

WHAT THIS STUDY ADDS?

- Common causes of non-cystic fibrosis bronchiectasis in Indian children are post-infectious, primary ciliary dyskinesia and allergic bronchopulmonary aspergillosis.

[2,3,5], indicating longer duration of illness or delayed diagnosis in our patients.

In our study, post-infectious causes were the commonest underlying illnesses, followed by suspected PCD which was consistent with other reports [2,5,7-9]. We could not identify underlying cause in about one-third of cases; earlier studies (5,7) also could not identify underlying cause in similar proportion of patients.

Majority of the patients did well with medical treatment alone. Only 14 (17.5%) patients required surgical intervention, suggesting that medical treatment is mainstay of treatment. Earlier reports suggest need for surgery in 6-23% [5,6-8]. Medical morbidity was observed in almost 95% of children in our study, and 5 children expired. An earlier study [10] reported death in 7% of children with bronchiectasis .

Limitations of our study include: retrospective design, hospital-based, and short follow-up. Also, FENO and not nasal Nitric oxide, was used for making a diagnosis of PCD. We also did not study immunoglobulin subclass deficiency. These may be some of the contributory factors for non- identification of underlying cause in one-third of our patients. Despite limitations, our study describes details of relatively large cohort from India. The study provides important information on non-CF bronchiectasis that may be applicable to developing countries.

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