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Research Article

Bronchiectasis in Children: Etiological Diagnosis

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Abstract

Introduction: The Bronchiectasis is not an uncommon condition in pediatric population.

Aim: To describe the etiological characteristics of children with non CF bronchiectasis.

Materials and Methods: A retrospective descriptive study of 40 cases of bronchiectasis in children in pediatric unit A of Mohammed VI Hospital, Marrakesh.

Results: The average age was 8 years with no sex predominance. The consanguinity was found in 42%. The main signs were cough 75% bronchorrhea dyspnea recurrent bronchopneumonia in 78%. At the physical examination, we found a digital clubbing in 55%, crackles in 65%. Chest CT showed bilateral involvement in 60% of cases with a cylindrical shape in 50%. The etiologies were mainly an immune deficiency 27.5%, asthma in 17.5% of the cases, a sequela of tuberculosis in 3 cases, a primitive ciliary dyskinesia in 3 cases, a foreign body a kartagener syndrome and a cystic fibrosis in two cases each and a sequela of viruses in one case. They remained unknown in 20% of the cases. Management was based on antimicrobial therapy in 85% associated with postural physiotherapy in 42.5%, immunoglobulin substitution in one case and lobectomy in 3 cases.

Conclusion: This study highlights the prevalence of immune deficiency in bronchiectasis in children, so it should always kept in mind if we have repeated bronchopneumopathy.

Keywords: *Bronchiectasis; Child; Immunodeficiency; Asthma; Lobectomy*

Introduction

The bronchiectasis describes a permanent dilation of the bronchi and bronchioles as a result of destruction of the muscles and elastic connective tissues. The disorder mostly starts with a narrowing of the bronchial tree triggered by an infection, which may lead to destruction of the epithelium if it becomes chronic. In developed countries, cystic fibrosis is the most common cause of bronchiectasis. In our context, they have multiples causes and the affection is quite common despite the codification of respiratory infections management in children and the effectiveness of immunisation. Through this study, we present data on underlying causes of bronchiectasis in Moroccan children in pediatric pulmonology unit of a tertiary care hospital in Marrakesh.

Methods

A review of children diagnosed with bronchiectasis attending the pediatric pulmonology unit of a tertiary-care hospital over a period of 9 years (2010 - 2019) was carried out. The diagnosis was done if it was a history of consanguinity or similar cases of bronchiectasis in the family chronic cough respiratory infections recurrent bronchorrhea or hemoptysis and if clinical examination showed thoracic deformation, digital clubbing, cyanosis and failure to thrive the features of chest x ray and the especially the CT scan. A chest X-ray can show a thoracic distension atelectasis, when it was normal the diagnosis of bronchiectasis was not excluded. High-resolution chest CT scan is the gold standard for the diagnosis of bronchiectasis, it was performed to evaluate the type the severity and distribution of bronchiectasis, it was often performed outside of an exacerbation after antibiotic treatment although, It revealed three types cylindrical varicose and cystic and can show a bronchus of higher caliber than that of the adjacent vessel, a bronchoarterial ratio > 0.6, mucus plugging or impactions, visibility of the bronchi at the periphery under the pleura, mosaic perfusion defects, air trapping on expiration. Basic investigations of children with bronchiectasis are shown in table 1. Indetermined etiology was if the assessment did not detect any abnormalities. An impact assessment consisted of respiratory functional explorations to highlight bronchial hyperreactivity, an echocardiography to detect an impact on the right heart cavities, a chronic lung heart and pulmonary artery hypertension and a 24-hours proteinuria to identify amyloidosis. Our management was mainly an alternating antibiotic respiratory physiotherapy, inhaled corticosteroid therapy alone or bronchodilators, immunoglobulin substitution antiretroviral therapy and antibiotic prophylaxis. Surgery was required in case of localized bronchiectasis, before lightning hemoptysis or if repetitive infections were resistant to the usual antibiotics and in pulmonary abscess, It was lobectomy. The follow up was carried out every 3 months. A chest CT and echocardiography were performed once a year. Data was analyzed by using excel.

Basic investigations of children with bronchectasis
1. Full blood count
2. Serum immunoglobulins (Ig G Ig A Ig M Ig E)
3. Immunoglobulin subclasses
4. Protein electrophoresis, alpha -1 antitrypsin
5. Lymphocyt subset (count of the CD4 and CD8 and CD19 CD16 NK)
6. NBT (Nitrobluetetrazolium)
7. Human immunodeficiency virus testing
8. Microbiological analysis including acid resistant bacilli staining, genexpert and quantiferon
9. Sputum for culture if children could expectorate.
10. Sweat test
11. PH metry
12. Bronchoschopy
13. Spirometry in chidren aged ≥ 6 years.

Table 1: *Investigations applied in determinig the etiology of non CF- Bronchectasis.*

Results

The median age at the time of diagnosis of non CF bronchiectasis was 8 years (range 1 - 15) without any sex predominance. Consanguinity was in 42% and similar cases of bronchiectasis in the family in 18%. The average time between onset of symptoms and consultation was 1 year. The main signs were cough 75% bronchorrhea 78%. Clinical manifestations are shows in table 2. On the physical examination, we had digital clubbing in 55%, chest deformation in 15% cyanosis and ataxia telangiectasia in 10% of cases, crackling in 65% and wheezing in 38% dextrocardia in 5%. A chest X-ray showed bilateral involvement in 58% of unilateral in 42% of atelectasis in 28% and rail images in 30% of cases. Thoracic CT was performed in all patients. This showed bilateral involvement in 60% with a cylindrical form in 50% a cystic form in 22.5% and a varicose form in 10%. The details of etiologic factors are shown in figure 1. Among immune deficiency, we had ataxia telangiactasia10% HIV 10% combine immune deficiency hyper IgM syndrome and Buckley syndrome in 2.5% each one. Management was based on alternating antibiotic therapy in 85% of cases associated with respiratory drainage physiotherapy in 42.5%, inhaled treatment and bronchodilators in 62.5% of cases. An immunoglobulin substitution in one case an antiretroviral treatment in 4 cases associated with antibioprophylaxis and a lobectomy in 3 cases. The evolution is characterized by stabilization (77.5%) extension of lesions (7.5%) complications (12.5%) and death in one case.

Symptoms	N (%)
Fever	27 (67.5%)
Dyspnea	26 (65%)
Chronic cough	30 (75%)
Bronchorrhea	31 (78%)
Hemoptysis	8 (20%)
Recurrent bronchopneumonia	26 (65%)
Failure to thrive	26 (65%)

Table 2: *Symptoms at the time of diagnosis in children with non CF- bronchiectasis in our study.*

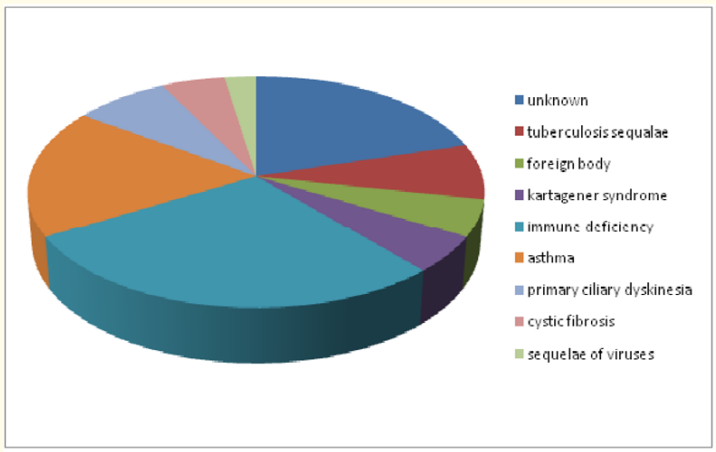

Figure 1: *Underlying causes of bronchiectasis in children with non CF- bronchiectasis.*

Discussion

Bronchiectasis are a fairly common pathology in pediatrics in undeveloped countries. In fact, its incidence is estimated in Great Britain at 1/5800 excluding cystic fibrosis [1], however in Morocco, its incidence remains unknown. The average age was 8 years in our series which is similar to the literature. We had no gender predominance contrasting with authors [2-5] and joining others [4]. The consanguinity and similar cases of bronchiectasis in the family were superimposed on data from other series [5]. The study shows that the average time between the onset of the first symptoms and diagnosis was 1 year, which is similar to the Eastman series [1], it was 3.8 years in Algeria, indicating a early diagnosis in our cohort. It is commonly accepted that productive cough is the main symptom of bronchiectasis in children [6]. In our series, the main clinical manifestations were a cough a recurrent bronchorrhea a hemoptysis which is similar with other series [2,4,5,6,7]. Physical examination showed a digital clubbing chest deformation and cyanosis like the authors [4]. For radiological features, the lower and middle lobes were the most affected which is similar to satire., *et al.* [2] and could be explained by the high frequency of immune deficiency and asthma in our study. Meanwhile, Chest CT showed bilateral involvement in 60% of cases and the predominance of the cylindrical form in half of cases, in another series the varicose form was the most frequent [5]. There is no correlation between the affected territory and the underlying disease, bronchiectasis very often affects several territories and or both lungs [8]. Cytobacteriological examination of sputum allows to isolate *Haemophilus influenzae, Streptococcus pneumoniae, Moraxella catarralis staphylococcus aureus* and *Pseudomonas aeruginosa* [9]. The most identified bacteria were *Streptococcus pneumoniae* and *Pseudomonas aeruginosa* in our unit. The etiologies were the immune deficiency 27.5% and asthma 17.5% which joins a Moroccan series where the immune deficiency represented 28% of the etiologies and contrast with an American series where pneumonia was noted in 19% of cases [10] and a Turkish series or primitive ciliary dyskinesia was found in half of the cases [2]. Among the identified immune deficiency, we had a predominance of ataxia telangiectasia and retroviral infection followed by humoral immune deficiency and finally combined immune deficiency which contrasts with an American series where the immune deficiency is predominant 74% especially the IgG deficiency (48%) and IgG subclass (18%) followed by combined immune deficiency (10%) and cellular immune deficiency (7%) and finally telangiectasia ataxia (2%) [10]. In Turkish series, a humoral immune deficiency was identified in (72%) with hypogammaglobulinemia (9.6%) and a cellular deficit and combined in 14% of cases each [2]. This difference could be explained by an under diagnosis of humorous immune deficiency in our structure that could be attributed to an evolution of the child's immune status during the first years of life. Therefore, we suggest to repeat the assay of immunoglobulins even if the levels are normal for age. Agammaglobulinemia may be complicated by bronchiectasis despite the substitution of immunoglobulins [11]. Asthma is the second etiology which contrasts with our American series where asthma was the last etiology [10]. This could be attributed to the parents lack of the observance of treatment and environmental control and to the low socioeconomic level of our populations. Among the infectious etiologies, we had two cases of sequelae of tuberculosis and two cases of sequelae of viruses. In contrast, pneumonia 61% measles 14% tuberculosis 11% and whooping cough 5% were found in another series [10]. It seems that whooping cough measles and tuberculosis are no longer providing bronchiectasis in our context, this could be related to the effectiveness of the immunization program in our country. Cystic fibrosis was in 3 cases in our series and 7 cases in Algeria and 33 cases in Tunisia [12]. Its frequency seems to be underestimated in the Maghreb countries. We have developed tools of diagnosis and nowadays we search genetic mutation. According to Munck, we must develop neonatal screening [13]. Primary ciliary dyskinesia with respiratory manifestations sinus and extra respiratory otorhino, it is confirmed by the existence of an abnormality of the ciliary beat or ciliary ultrastructure with a flow of collapsed nasal NO. It was detected in 3 cases in our series and in half of the cases in a Turkish series [2]. This rather high prevalence in Turkey would be explained by the high consanguinity rate. This low rate found in our series be related to an underdiagnosis. The foreign body represents a significant cause of bronchiectasis in children, it must be kept in mind if we had respiratory distress pneumonia that recur in the same territory or an unusual asthma and make an emergency bronchoscopy. The prevention

is mandatory. Our management of children's bronchiectasis was respiratory physiotherapy for improving the quality of life and the extension of lesions. Inhalation of hypertonic saline 3% appears to be useful in children with bronchiolitis and atelectasis [14], that of saline 7% at 4 ml twice a week for 2 years would improve respiratory function [15]. Antibiotic therapy can be administered orally injectable and inhaled in case of cystic fibrosis, it can be continuous or alternating to reduce the risk of resistance based on amoxicillin clavulanic acid or ciprofloxacin if it is a *Pseudomonas aeruginosa* for 14 days for at least 3 months. Studies report that using an anti-inflammatory macrolide at 5 mg/kg/day three times a day per week for 6 months would have a similar effect [15,16]. Azithromycin is more tolerated than erythromycin. It is thought to decrease airway inflammation by inhibiting neutrophil migration to the respiratory epithelium and by blocking proinflammatory cytokines and mediators. Inhalation of corticosteroid inhaled alone or associated with a bronchodilator is of interest in children with allergic asthma or in cases of aspergillosis [17]. The surgery consisted of a lobectomy which joins other series [18]. Influenza and pneumococcal vaccination and treatment of otitis and gastroesophageal reflux are mandatory. For immune deficiency, we used immunoglobulin substitution antiretroviral therapy and antibiotic prophylaxis. Moreover, the management of cystic fibrosis consisted of an inhalation of colimycin associated with quinolone, nebulization with hypertonic saline a daily self-drainage of fat-soluble vitamins and pancreatic extracts if pancreatic insufficiency. In fine, surgery was required in case of localized bronchiectasis, before lightning hemoptysis or if repetitive infections were resistant to the usual antibiotics and in pulmonary abscess.

Conclusion

The bronchiectasis in children represent a diagnostic and therapeutic challenge for the pulmonary pediatrician. Etiologies were immune deficiency followed then by asthma. The management is still little codified, it must be part of the multidisciplinary and focus mainly on prevention. Improved diagnostic and therapeutic tools could help the management of children's bronchiectasis in our country.

Conflict of Interest

None.

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Author Contributions

EF K wrote the manuscript. RN, AB, DG supervised the findings of this work, BM approved the findings of this work. All the authors have read the final version of the manuscript.

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