Pediatric bronchiectasis: An orphan disease ending in pneumonectomy: A case report

Esubalew Taddesse Mindaye a,*, Goytom Knfe Tesfay b, Maru Gama Erge b

a Department of Surgery, St. Paul’s Hospital Millennium Medical College, Swaziland Street 1271, Addis Ababa, Ethiopia
b Department of Surgery, St. Paul’s Hospital Millennium Medical College, Addis Ababa, Ethiopia

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A B S T R A C T

INTRODUCTION: Bronchiectasis is a progressive infectious and inflammatory disease characterized by chronic airway inflammation resulting in irreversibly dilated, thick walled bronchi and progressive decline in lung function [1]. It commonly affects proximal and medium sized bronchi [1]. Although it is regarded as an orphan disease in the developed world, it is still major cause of respiratory morbidity for children of developing nations as most are undiagnosed and untreated leading to early decline of respiratory function [2]. The presentation of pediatric bronchiectasis is different from the adult and vary depending on age and severity of the disease [3]. Chronic wet cough is the most common and consistent presentation in most children [3].

1. Introduction

Bronchiectasis is a progressive respiratory disease characterized by chronic airway inflammation resulting in irreversibly dilated, thick walled bronchi and progressive decline in lung function [1]. It commonly affects proximal and medium sized bronchi [1]. Although it is regarded as an orphan disease in the developed world, it is still major cause of respiratory morbidity for children of developing nations as most are undiagnosed and untreated leading to early decline of respiratory function [2]. The presentation of pediatric bronchiectasis is different from the adult and vary depending on age and severity of the disease [3]. Chronic wet cough is the most common and consistent presentation in most children [3].

High resolution computerized tomography of the chest (HRCT) showing increased broncho-arterial ratio (signet ring sign), bronchial wall thickening and lack of bronchial tapering is the gold standard diagnostic tool [3]. Spirometry may show functional information about the severity of the disease [4]. The main goals of treatment are control of symptoms, decrease exacerbation and preserve lung function, and this requires multidisciplinary approach [3]. Surgical intervention is reserved for patients with localized bronchiectasis with persistent symptoms and recurrent infections despite maximal medical therapy [2]. The case report has been reported in line with the SCARE 2020 criteria [5].

2. Case presentation

Ten years old female presented with a complaint of intermittent wet cough of 5 weeks’ duration associated with low grade intermittent fever, shortness of breath, easy fatigability and loss of appetite. She had frequent similar complaints for the last 4 years for which she visited different hospitals and was given antibiotics for the diagnosis of pneumonia. She had 6–8 acute exacerbations annually but...
wasn’t admitted to hospital. She had no contact history with known pulmonary tuberculosis patients and she is fully vaccinated. She has no history of drug allergy, self or family history of relevant medical or surgical illness. On presentation her vital signs were; PR = 96 beat/minute, RR = 28 breath/minute and oxygen saturation was 90% with 3 L of intranasal oxygen support. She had decreased air entry over her left posterior lower third lung field and digital clubbing. Otherwise, there was no remarkable finding on other systems evaluation.

Her complete blood count, renal function test and serum electrolytes were in the normal range. Chest computerized tomography (Chest CT) showed collapsed left lung with fluid filled lobar and segmental bronchi (Fig. 1). There was significant cardio-mediastinal shift to the left and compensatory hyperinflation of the right lung with herniation of the right upper and middle lobe in to the left chest. Bronchoscopic evaluation revealed destroyed left bronchial wall starting from the carina and filled with extensive purulent secretion. Bronchoscopic suctioning of the purulent fluid was attempted but it was very thick and was difficult for further evacuation. Culture from the bronchial aspirate revealed staphylococcus aureus growth which was resistant for penicillin, cloxacillin and cephalosporin’s, and sensitive for gentamicin.

With an impression of left lung collapse secondary to extensive diffuse bronchiectasis with recurrent infection, the patient was started on Intravenous antibiotics and continued on intranasal oxygen. The surgical team decided for surgical intervention and the patient was operated through left posterolateral thoracotomy after getting informed written consent from the patient’s family. The intraoperative finding was hyper inflamed right lung filling two-third of the left hemithorax and completely collapsed, bronchiectatic left lung (Fig. 2). With these findings left pneumonectomy was done, left tube thoracostomy inserted and thoracotomy wound was closed in layers.

Post procedure, she was transferred to pediatric intensive care unit; and put on oxygen support and continued on intravenous antibiotics and analgesics. Subsequently, she had smooth recovery and was transferred to surgical ward. The chest tube output was insignificant and control chest x ray taken on her 4th post op day showed no evidence of collection for which the chest tube was removed.

Histopathologic study of the excised lung showed prominently dilated bronchial airways lined by flattened respiratory epithelial cells. There was extensive peribronchial inflammatory infiltrates and, extensive fibrosis and hyalinization of blood vessels with perivascular inflammatory infiltrates, the final impression being diffuse bronchiectasis (Fig. 3).

Subsequently, the patient showed a remarkable improvement and was discharged from the hospital in a stable condition.

3. Discussion

It is difficult to know the true burden of bronchiectasis in children as the diagnosis is often delayed and depends on the living condition of a society, physicians awareness and availability of advanced imaging with pediatric protocol [6]. It is common in socioeconomically disadvantaged communities across the globe where there is poor hygiene, overcrowding and access to basic health care is very limited [6]. Recurrent lower tract airway infections are the leading causes of pediatric bronchiectasis followed by primary immune deficiency, primary ciliary dyskinesia, foreign body aspiration and airway structural abnormalities [2]. Although we did not work up her for other causes, our patient is from low...
socioeconomic family, has moderate malnutrition and was being treated for pneumonia frequently which looks to be the likely cause for her disease.

Like our case chronic wet cough is the predominant presentation of pediatric bronchiectasis accompanied by exertional dyspnea, hemoptysis, chest wall deformity, failure to thrive and reduced quality of life [2]. Although chest x-ray has poor diagnostic value in the absence of compatible clinical history and additional investigations, it can be used as the first imaging test in the work up of suspected patients [2]. HRCT is the gold standard modality to diagnose and stratify severity of bronchiectasis based on: extent of bronchiectasis (involved lung segment), peribronchial thickening, extent of mucus plug and lung collapse/consolidation [7,8]. In this regard, our patient had severe disease with diffuse bronchiectatic changes involving the left lobe which is collapsed and filled with excessive purulent secretion. Considering all these facts, the surgical and pediatric team decided to manage her surgically as the success of medical therapy is limited. Although surgery is the last option of management for bronchiectasis especially for pediatric patients, it is very difficult to salvage the lung once destroyed irreversibly. This alarms the importance of early prompt diagnosis and intervention with multidisciplinary approach. It is also crucial to equip health care professionals with adequate knowledge about the disease as most pediatric patients like our case may not have productive cough like adults leading to misdiagnosis or significant delay in diagnosis.
Surgical treatment like segmentectomy and lobectomy is reserved for patients with localized bronchiectasis with persistent symptoms, recurrent infections despite maximal medical therapy and hemoptysis [2]. We are following our patient post operatively with clinical evaluation, chest x-ray every 3 months and chest CT scan annually for the coming 2 years as involvement of the contralateral lung is associated with higher morbidity and mortality. Our patient’s symptoms have resolved completely and she has no evidence of recurrent infection. Her family and she is also happy with her treatment.

4. Conclusion

Neglected pediatric bronchiectasis is associated with significant morbidity and mortality. So, it should be considered as differential diagnosis in children with recurrent respiratory symptoms as timely and prompt diagnosis is crucial for early intervention. Although surgical resection is the last option of treatment for patients with bronchiectasis, it is inevitable once the lung is irreversibly destroyed. Post-operatively patients require cautious follow up as remaining lung involvement is associated higher morbidity and mortality.

Declaration of Competing Interest

All authors declare that they have no conflict of interest.

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Ethical approval

Ethical Clearance was obtained from the Institutional Research and Ethics Review Committee (IRB) of SPHMMC for the publication of the case report and accompanying images.

Consent

Written informed consent was obtained from the patient’s family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author’s contribution

1. Esubalew Taddese Mindaye, MD

Conceived and conducted the study, did literature search and Critical revision of the manuscript, involved in the management of the case

2. Goytom knfe tesfaye, MD

Conducted over all supervision and critical revision of the manuscript

3. Maru Gama, MD

Involved in the surgery, conducted over all supervision and critical review of the manuscript

Registration of research studies

Not applicable.

Guarantor

Esubalew Taddese Mindaye, MD.

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