Paediatric Respiratory Reviews 20S (2016) 30–33

Contents lists available at ScienceDirect

Paediatric Respiratory Reviews

Review

Pneumothorax in cystic fibrosis: beyond the guidelines

Robert W. Lord [1,2], Andrew M. Jones [1,2], A. Kevin Webb [1,2], Peter J. Barry [1,2,*]

[1] Manchester Adult Cystic Fibrosis Centre, University Hospital of South Manchester, Southmoor Road, Wythenshawe, M23 9LT, United Kingdom
[2] Institute of Inflammation and Repair, University of Manchester, United Kingdom

ARTICLE INFO

Keywords:
Cystic Fibrosis
Pneumothorax
Persistent air leak
Pleural intervention
Pleurodesis

SUMMARY

Pneumothorax is a serious but common complication in patients with cystic fibrosis (CF). It has adverse prognostic implications as well as associations with subsequent reduction in lung function and significant risk of recurrence. Management dilemmas frequently occur that are beyond current guidelines. We review the evidence and highlight management difficulties in pneumothoraces in CF.

INTRODUCTION

Pneumothorax represents one of the most feared complications of respiratory disease in cystic fibrosis (CF) for patients. Its annual incidence is estimated at 0.64% according to US registry data, with a lifetime risk of 3.4% [1]. In CF although pneumothoraces may occasionally be asymptomatic, the majority present symptomatically with chest pain or breathlessness and on rare occasions respiratory failure. The gravity of this complication in CF is emphasised by the repeated identification as pnemothorax as an important prognostic factor [1,2].

There are identifiable anatomical risk factors for pneumothoraces in CF. Cysts, blebs and bullae are all commonly identified in the lungs of CF patients [3]. These vulnerable areas are prone to distension as a result of gas trapping due to small airway obstruction. This is supported by the finding of high residual volumes in CF patients with pneumothorax [4]. In addition, registry data has shown clear associations between pneumothorax and both severe airflow obstruction, and factors known to result in airway obstruction such as allergic bronchopulmonary aspergillosis (ABPA) [1].

Guidelines on the management of pneumothoraces in patients with CF exist, however any such guidelines are unable to cover all aspects of the condition in a complex patient population [5,6]. The purpose of this review is to highlight management difficulties beyond the scope of the guidelines as illustrated by the following case.

CASE

An 18 year old male with CF (genotype Phe508del/Phe508del), presented for the first time to the adult CF services during a period of spirometric decline which had been ongoing for 3 years. He was chronically colonised with *Pseudomonas aeruginosa* and *Mycobacterium avium*. Despite courses of intravenous antibiotics and treatment for *M. avium*, he remained symptomatic. His aspergillus titres which were previously unremarkable became consistent with ABPA and he was started on therapy with subjective improvement.

Within weeks he re-presented as an emergency with sudden onset of a large right-sided pneumothorax. Despite insertion of a large-bore intercostal drain the lung failed to re-inflate, and a considerable air leak remained present for weeks. A CT scan of his thorax demonstrated a bronchopleural fistula. Additional management strategies including surgical repair of the air leak, insertion of an endobronchial valve and lung transplantation were considered. Each approach however was limited by the development of an aspergillus pleural infection, likely as a consequence of direct seeding via a bronchopleural fistula. He was treated conservatively with a Heimlich valve and ambulatory bag, and antifungal therapy.

The sequela of this approach was a chronic pneumothorax with significant pleural thickening. Due to these relatively fixed anatomical changes removal of the chest drain was possible without further clinical deterioration. Unusually the patient described the ability to expectorate foul watery fluid, which we suspected was pleural fluid, when he adopted certain positions. He was discharged home and remains independent and mobile. As alluded to previously, the difficulties of management of chronic broncho-pleural fistulae, pleural infection and chronic pneumothoraces are areas of limited evidence and require careful multi-disciplinary consideration.

* Corresponding author. Manchester Adult Cystic Fibrosis Centre, University Hospital of South Manchester, Southmoor Road, Wythenshawe, M23 9LT, United Kingdom, Tel.: +44 0 1612912016.
E-mail address: peter.barry@uhsm.nhs.uk (P.J. Barry).

http://dx.doi.org/10.1016/j.prrv.2016.06.012
1526-0542/

DILEMMAS IN DIAGNOSIS

Role of imaging beyond the chest radiograph

Diagnosing pneumothorax can be challenging in patients with advanced CF lung disease (see Fig. 1). Pleural adhesions are common and thus associated lung collapse is commonly non-uniform. The presence of lung cysts that often can be large and extensive further increase difficulties in assessing the extent of a pneumothorax or, in some instances, falsely suggest the appearance of a pneumothorax. Therefore a low threshold is justified for the use of a chest CT scan to aid evaluation and management. They allow a more accurate assessment of size of a pneumothorax, identify a site for the chest drain insertion and help confirm resolution following treatment [7].

DILEMMAS IN MANAGEMENT

Management strategies for pneumothorax in CF

Aspiration is recommended in the BTS guidelines however there is little evidence to support this in CF patients. The current authors suggest that given the high risk of recurrence in CF and the potential risks associated with this, aspiration should be avoided and the placement of a chest drain should be considered to facilitate definitive management strategies such as pleurodesis. A caveat exists in that some small asymptomatic pneumothoraces may be managed conservatively and resolve spontaneously.

Despite the insertion of an intercostal drain, risk remains for marked deterioration in respiratory status due to a number of factors including pleural pain, relative immobility and the underlying lung collapse. A number of measures should be implemented immediately to minimise risk of deterioration, including intravenous antibiotics, airway clearance and early mobilisation under the supervision of experienced CF physiotherapists, and a review of all nebulised therapy by the CF team. Also pivotal is appropriate analgesia to allow the above measures to be delivered.

The downstream effects for other systems affected by CF, such as the gastrointestinal tract, should be considered–for example adequate hydration and prophylactic use of laxatives to prevent constipation and bowel obstruction predisposed by relative immobility and use of some analgesics.

Strategies to prevent recurrence

Pneumothorax recurrence is common in CF with one study estimating recurrence rates as high as 63% when treated with intercostal drainage alone [4]. Given the associated morbidity of recurrence, strong consideration should be given to medical pleurodesis following lung re-expansion in all patients treated with an intercostal drain including those with a first pneumothorax. This recommendation is in contrast to current guidelines, but we would argue that this approach permits greater rehabilitation of the individual and less limitation of lifestyle over the immediate to medium term.

Choice of sclerosing agent

Previously pleural interventions, such as talc pleurodesis, were seen as a contraindication to lung transplantation surgery. Evidence now exists that it should not influence decisions and lung transplantation is possible following talc pleurodesis but may necessitate longer surgical times [8]. Similarly blood pleurodesis is feasible and does not present a contraindication for lung transplant surgery. With a chest drain in situ the instillation of either agent is performed easily. Talc pleurodesis has been reported to have high success rates [9].

Management strategies for a persistent air leak

A persistent air leak (PAL) is defined as continued bubbling through the chest drain with or without re-expansion of the collapsed lung at 48 hours. Initial management is covered in the BTS guidelines. Despite not being specific to CF, the general principles of application of suction to the chest drain, wider bore chest drains and allowing time are appropriate. This combination in a series of secondary pneumothorax, with any underlying lung disease, has been shown to be successful in 79% of patients at 14 days [10].

Potential management strategies for PAL that continue following the above measures include surgery through thoracotomy, video-assisted thoracoscopic surgery (VATs), medical pleurodesis with talc or other sclerosing agents, or conservative approaches using Heimlich valves as required in the above case [11]. A single case report exists of the successful use of endobronchial valves in CF for the treatment of pneumothorax [12]. The potential risk of abscess formation and consolidation distal to the valve should be considered in the setting of the CF lung. Persistent pneumothorax remains an indication for lung

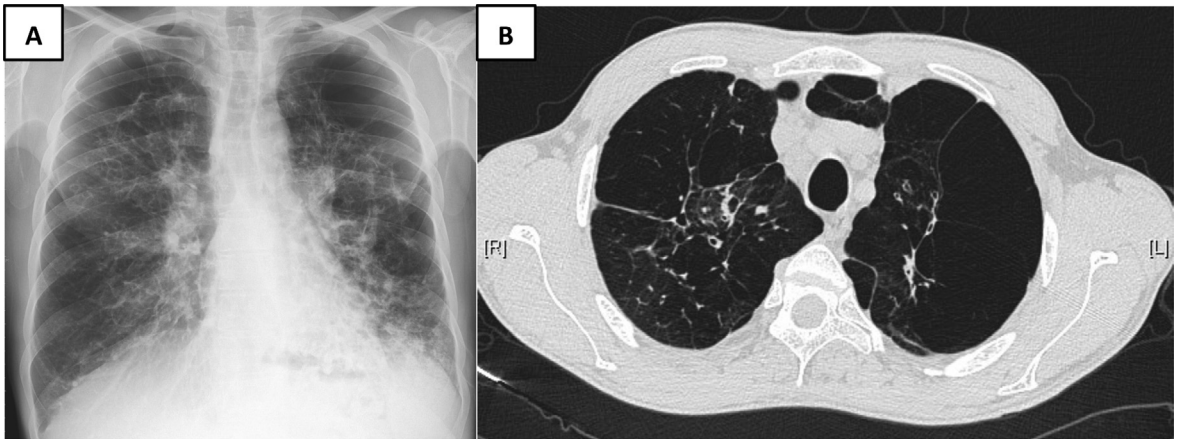

Fig. 1. Demonstrates imaging from a CF patient with a suspected left apical pneumothorax on chest radiograph (A). On CT chest this was shown to be a large bulla (B).