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Talc pleurodesis in a case of Duchenne Muscular Dystrophy with recurrent pneumothorax

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Introduction

Recurrent pneumothorax is a rather uncommon finding in patients with neuromuscular diseases. For cases that happen to patients with neuromuscular diseases while on non-invasive positive pressure ventilation, pneumothorax could be fatal. Medical staff must be vigilant for this complication. Recurrent pneumothorax can be treated with either medical or surgical pleurodesis, depending on the patient's physical conditions. Each method has its own advantages and drawbacks. We present a young adult with history of Duchenne Muscular Dystrophy complicated with recurrent pneumothorax.

Case presentation

A 20-year-old boy with history of respiratory insufficiency secondary to Duchenne Muscular Dystrophy was admitted to our paediatric intensive care unit due to pneumonia.

He had been followed up regularly in our department for many years. He was bedbound and had severe scoliosis with Cobb's angle of 90° and chest wall deformity. His respiratory function was slowly deteriorating and he was put on noninvasive positive pressure ventilation with oxygen supplement at home. He had gastrostomy and tracheostomy performed a few months before this admission. There was history of repeated episodes of pneumothorax. The first episode happened in February 2010 involving his right lung and he had talc pleurodesis performed. Then he suffered bilateral pneumothorax and was treated with chest drain insertion in November 2010. In February 2011, he was incidentally found to have left pneumothorax with a chronic loculated right pneumothorax while having a CT scan for preoperative assessment for gastrostomy. Ultrasound guided chest drain insertion was performed with minocycline pleurodesis performed subsequently. Apart from transient left sided pleuritic chest pain which was managed with simple analgesia the procedure was well tolerated and there was no major complication. Post-procedural chest X-ray showed resolution of left sided pneumothorax.

Five months later, he was admitted to another hospital with dyspnoea, fever and impaired conscious state requiring mechanical ventilation through tracheostomy. Antibiotics were started. The first arterial blood gas showed $PaCO_2$ of 6.9kPa. He improved with treatment and regained consciousness. He was transferred to our department for further management. Physical examination on arrival showed symmetrical but fair air entry bilaterally. Chest X-ray showed no definite consolidation but there was a small rim of pneumothorax over left lung and also loculated pneumothorax over right lung similar to previous films. There was severe scoliosis with chest wall deformities. Later bronchoscopy showed extensive tracheitis, which was likely to be tracheostomy tube related and this was treated with antibiotics.

He had repeated episodes of lung collapse and persistent left sided pneumothorax with desaturation during his stay in intensive care unit. There was also hypercapnia with highest $PaCO_2$ up to 25.3 kPa and pH 7.08. Chest drain was inserted. Chest X-ray after chest drain insertion showed satisfactory re-expansion. In view of recurrent nature of pneumothorax, surgical pleurodesis was considered but the patient was physically unfit.

As a result, medical pleurodesis was performed using 3 grams of talc powder mixed with 10 ml of 2% lidocaine and 100 ml normal saline. He experienced pleuritic chest pain immediately after pleurodesis but vital signs remained stable. Morphine and paracetamol were given for pain control.

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He was noticed to have respiratory distress on day one following pleurodesis with desaturation again. Physical examination showed decrease air entry over left side but bedside cold light examination did not reveal any apparent air leak. Nevertheless, urgent needle aspiration was attempted and no air was aspirated out. Chest X-ray did not show any pneumothorax but there was increasing haziness over left lung field. He required a high ventilator setting to maintain satisfactory oxygenation with maximal inspiratory pressure up to 34 cm H_2O and FiO_2 of 90%. Though we were able to step down the ventilator support gradually over the following few days, he had an acute deterioration on day 3 after pleurodesis with desaturation and bradycardia despite manual bagging. This progressed to cardiac arrest and cardiopulmonary resuscitation was needed. Totally 3 doses of adrenaline were given. He developed ventricular fibrillation and two shocks of defibrillation were given. Sinus rhythm returned after 12 minutes. Large amount of think sputum was aspirated out from the tracheostomy tube. Emergency bronchoscopy was performed which showed persistence of tracheitis but was much improved comparing with previously. Vital signs were stable afterward and the patient regained full consciousness. C-reactive protein raised from normal to 148 mg/L on D3 and peripheral white cell count increased to 38.6 x 10^9/L with neutrophilia of 30.8 x 10^9/L. Bronchoalveolar lavage showed large amount of pus cell with many macrophages mixed with extracellular polygonal foreign bodies with green birefringence under polarizing microscopy, which was compatible with talc powder. Bacterial culture did not grow any organism. There was no further episode of pneumothorax till the time of writing.

Discussion

Duchenne Muscular Dystrophy is known to have deterioration in pulmonary function during the late adolescence. Their vital capacity usually peaks between 9-16 year old and decreases by 5-10% per year until ventilatory support is instigated.[1] Long term non- invasive ventilation has improved survival as well as quality of life. Usually for Duchenne Muscular Dystrophy, there would be reduction in vital capacity during initiation of wheelchair use and respiratory insufficiency occurs between 18 and 20 year-old. Five year survival rate of 8% was quoted if assisted ventilation was not initiated.[2] Several large series suggested excellent long term survival rates for cases receiving non-invasive ventilatory support with less rapidly progressive muscular dystrophies.[2] Potential problems of positive pressure ventilation include barotrauma and pneumothorax.[3] The incidence of barotrauma for mechanically ventilated patients was quoted at 0.5-40% for heterogeneous groups of patients receiving via endotracheal tube or tracheostomy.[4] Scoliosis can cause unequal ventilation in the lungs with the distortion of the bronchial tree causing air trapping and thus increased risk of air leaking syndrome. However a Japanese series of Duchenne Muscular Dystrophy had suggested that DMD itself had increased risk of spontaneous pneumothorax even during absence of positive airway pressure, and there was no additional risk posed by the use of mechanical ventilation or the presence of thoracic deformities.[5] Positive pressure ventilation may contribute to the air leak but not necessarily was the main cause. Also in our case, thoracic deformities have persisted for a long time. Further studies are necessary to define its relationship.

Different strategies for recurrent pneumothorax have been studied including both surgical and medical pleurodesis. Table 1 shows the comparison between the 2 management options. The main target is to prevent further episodes of pneumothorax. For secondary pneumothorax, different series have quoted mortality ranging from 1 to 17%, which is much higher than primary spontaneous pneumothorax, which is 0.09% in male and 0.06% in women.[6] Prevention of this fatal complication is therefore an important issue. The British Thoracic Society recommends surgical treatment, using either an open or video-assisted thorascopic surgery (VATS) approach, for recurrent pneumothorax as recurrence rate after surgical pleurodesis is far less than medical pleurodesis.[7] However, for patients who are unfit for surgery, like in our case, medical pleurodesis has to be used.

There are wide range of agents that can be used for medical pleurodesis, including talc, tetracycline derivatives like minocycline or doxycycline and silver nitrate. Other agents like bleomycin, mitomycin C, cisplatin or OK-432 (Streptococcus pyogenes A3) are also used in pleurodesis but mainly for recurrent pleural effusion.

In our case, talc and minocycline were used. Talc is one of the most effective agents for medical pleurodesis for patient with secondary spontaneous pneumothorax. It was first described by Bethune in

Table 1. Comparison of surgical and medical options for recurrent pneumothorax

Surgical strategies	Medical strategies
Open thoracotomy	Medical pleurodesis with chemical agents
Video-assisted thoracoscopic surgery	
Surgical chemical pleurodesis	
Recurrence rate	Recurrence rate
Open thoracotomy with pleurectomy: 1%	Medical pleurodesis: 10-20%
VATS with pleurectomy and pleura abrasion: 5%	
General anaesthesia	Local anaesthesia
For all surgically fit cases	For cases not fit for surgery or refuse

1935 as a tri-layered magnesium sheet silicate.[8] It causes extensive inflammation over the pleura and induces cytokines and adhesion molecules including IL-8, VEGF and TGF-beta, which will promote adhesion of visceral and parietal pleura. It can be used in treatment of benign and malignant effusions, as well as recurrence of pneumothoraces.[9,10] Use of talc pleurodesis for treatment of pneumothorax had been shown to have a successful rate of 91% in a series of studies from 1947 to 1994.[8] However, high potency means it would induce a stronger inflammatory response in the pleura and causes more adverse effects.

Fever is common after talc pleurodesis as the inflammatory cytokines can enter the systemic circulation and cause inflammatory response. This usually occurs in the first 12 hours and may last till 72 hours.[8] It can also be caused by systemic absorption of talc particles. A study had suggested that talc containing mixed particles with diameter smaller than 5 micrometer would cause more marked acute systemic inflammatory response and suggested using particles with diameter larger than 20 micrometer.[10] Post-talc respiratory failure had been documented. The most severe form would be acute respiratory distress syndrome (ARDS). A large series showed the incidence of ARDS was 1.3% and the mortality rate was 0.55%.[11] Respiratory insufficiency was also noticed in cases after pleurodesis, which could be either explained by the limitation by pain and also the talc induced pneumonitis.[11] In our case with the combination of underlying musculoskeletal problem, it may worsen the pre-existing respiratory insufficiency, causing increasing hypoxemia and hypercapnia. Empyema might occur after instilling contaminated talc in the pleural cavity therefore aseptic technique must be observed during the procedure of medical pleurodesis.[8]

Other non pulmonary side effects including arrhythmia, myocardial infarction, hypotension or even cardiac arrest have been reported.[8] Thus close monitoring of the vital signs of patients after talc pleurodesis is necessary. Talc has been associated with development of lung cancer, which is probably related to the asbestos in the talc.[12] The medical grade talc however is free from asbestos.

Conclusion

Recurrent pneumothorax has a higher mortality and morbidity especially in secondary spontaneous pneumothorax. Inpatient management is advised. In Duchenne Muscular Dystrophy, the disease itself, as well as other factors may be associated with the increased risk of recurrent pneumothorax. Prevention by pleurodesis would be an important management strategy for decreasing the mortality. However, pleurodesis has its own adverse effects including life threatening complications in short term. Therefore it is important that we closely monitor the patient after commencing agents for pleurodesis.

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