Respiratory failure in a patient with hypophosphatemic rickets: can an endobronchial stent make the difference?

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Abnormalities associated with phosphate metabolism can lead to thoracic deformities that result in respiratory failure, which is conventionally managed by means of supplemental oxygenation, positive airway pressure and physiotherapy. However, when these measures fail, the clinician faces a dilemma, since many patients cannot tolerate a major surgical procedure. A minimally invasive technique, insertion of an endobronchial stent, might offer a solution.

KEYWORDS: respiratory failure, rickets, endobronchial stent

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Introduction

Although reports of osteomalacia-related respiratory complications are abundant in the paediatric literature, reports in adults are scarce. Abnormalities associated with phosphate metabolism can lead to respiratory failure by different pathogenetic mechanisms including skeletal thoracic deformities and myopathy. Patients are conventionally managed by means of supplemental oxygenation, positive airway pressure and physiotherapy, in additional to specific medical therapies targeting their metabolic abnormalities. If these measures fail, options are limited, as surgical interventions can pose significant risks to the patient. We report, for the first time to our knowledge, successful use of minimally invasive procedure, bronchoscopic stenting, with relief of respiratory failure secondary to lobar collapse in a patient with hypophosphotemic rickets and thoracic deformities.

Case presentation

The patient, a 21-year-old man, was under follow-up for hypophosphatemic rickets, which had been diagnosed in early childhood. It was complicated by recurrent varus tibial deformities that required multiple corrective surgeries. He had also undergone sleeve surgery for obesity. He was being treated with alpha calcidol, calcium, phosphate and later growth hormone. Despite treatment, his disease had progressed, with the development of hyperparathyroidism, and he was using a wheelchair and dependent on his family for activities of daily living.

The patient was admitted with a complaint of progressive shortness of breath over a month, which in the last 10 days prior to admission was associated with fever and cough productive of whitish sputum. He was initially managed in his local hospital with physiotherapy, antibiotics, supplemental oxygen and positive airway pressure with little improvement. Eventually, he was discharged home on oxygen 5 L/minute. A week later, he presented to the emergency department at our hospital. On examination, he was afebrile, in respiratory distress and using his accessory muscles with subcostal retractions with a respiratory rate of 34 per minute, with blood pressure of 120/72, pulse rate of 120 per minute and oxygen saturation of 88% on 5 L by nasal canula. The patient had gross limb and thoracic deformities, with collapse of the upper region of the thorax on the right side and scoliosis. Upon auscultation, lung sounds were diminished bilaterally, but more so on the right side. Systemic and cardiac examination was unremarkable. Chest radiography showed predominantly ‘Z’ shaped scoliosis with areas of bilateral atelectasis. Computed tomographic scan of the chest with angiogram (CT) showed complete right upper collapse and partial right lower lobe collapse and subsegmental atelectasis in the left lower lobe. There was no evidence of pulmonary embolism. Furthermore, when compared with last CT image performed few months earlier, the new CT scan showed progression of thoracic deformities because of worsening of the scoliosis and rib fracture. Accordingly, the patient was started on high-flow nasal oxygen alternating with bi-level positive airway pressure (BiPAP) and broad-spectrum antibiotics. However, despite these interventions, there was no clinical improvement or reduction in his oxygen requirement.

Thereafter, his case was discussed in a multidisciplinary meeting. As his imaging (Fig 1) showed compression of the right main bronchus between the ribs and spine resulting in total collapse of the right upper lobe and partial collapse of the right lower lobe, bronchoscopic stenting was recommended. It was felt that any surgical intervention would carry a high risk to the patient.
Stenting for lobar collapse in rickets

12 days after admission, bronchoscopic stenting was performed in the operating room with the fluoroscopic guidance under general anesthesia using endotracheal tube (size 7). A fiberoptic therapeutic scope was used to insert 30 × 15 mm Boston metallic uncovered bronchial stent. The procedure was uneventful. Following the procedure, the patient had gradual improvement with oxygen saturation over 90% on room air. Repeat CT scan chest showed re-expansion of the right lung, particularly right upper lobe (Fig 1). He was discharged 3 days after the stenting procedure with supplemental oxygen as needed and nocturnal BiPAP. He was followed up in the clinic as an outpatient and remained stable other than one mild infective episode that resolved after few days with antibiotics. His oxygenation normalised on room air and he did not use oxygen or BiPAP when he was seen in his last follow up 14 months after stenting. Imaging also showed stability of the pulmonary condition and the stent remained in position.

Discussion

Thoracic deformities resulting from rib fractures secondary to osteomalacia are an unusual cause of respiratory failure, with very few cases reported in adults. Expert medical management is necessary to halt progression of the pathophysiologic processes that leads to spinal deformities and myopathy and eventually respiratory failure. However, sometimes that is not possible and the physician maybe confronted with a patient with respiratory failure, as with the case described in this report. Our patient failed to respond to conventional medical management such as oxygen and non-invasive ventilation. Spinal surgery might be another option; a successful outcome was reported in a few cases. However, our patient was considered at high risk for anaesthesia and a major surgical procedure.

Stent placement in our patient led to reopening of the collapsed lobes and recovery of respiratory function to the baseline condition. We hypothesise that the stent restored patency of the airway leading to better aeration and gas exchange. The response was evident both clinically and radiologically. To the best of our knowledge, this is the first case in which such an intervention has been reported.

In conclusion, bronchoscopic stent placement offers a relatively non-invasive intervention, which might be of potential value in the management of respiratory failure associated with thoracic deformities secondary to osteomalacia and rickets.

References


Fig 1. CT Chest before (a,b) and after (c,d) the stenting procedure, showing reopening and reaeration of the collapsed right upper lobe.
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