CASE STUDY

Use of bilevel positive airway pressure ventilatory support for pathological flail chest complicating multiple myeloma

J. Abisheganaden, C.B.E. Chee, Y.T. Wang


ABSTRACT: Multiple myeloma is a common disease that universally involves the skeletal system. Although rib involvement may occur, the development of pathological flail chest is rare. We describe the treatment and course of this condition in an elderly female, and the use of the bilevel positive airway pressure (BiPAP) ventilatory system in providing pneumatic stabilization, while definitive chemotherapy was given to heal the pathological fractures.

Our experience with this patient suggests that, despite its dramatic clinical manifestation, the association of flail chest with multiple myeloma may not predict a poor prognosis. We have also found that pneumatic stabilization can be achieved by using the bilevel positive airway pressure ventilatory support through a tracheostomy.


Flail chest usually occurs secondary to traumatic multiple rib or sternal fractures. Patients with such injuries, who are in acute respiratory failure, have often required positive pressure ventilation and prolonged mechanical ventilation to achieve internal pneumatic stabilization of the chest wall [1, 2]. This has been the traditional form of treatment. Surgical stabilization has also been suggested for severe flail chest, but its use has been questioned due to complications [3]. Other forms of achieving pneumatic stabilization have developed in recent years, including the use of mini-invasive [4] and noninvasive techniques [5]. These methods of achieving pneumatic stabilization are reliable and are now widely practised [4–6]. Hence, there has been a change in attitude and management practice over the years from the traditional invasive methods to the current less invasive or noninvasive techniques.

Although rib involvement may occur in multiple myeloma, the development of pathological flail chest is rare. We describe a female with multiple myeloma presenting as flail chest with respiratory failure, and the use of the bilevel positive airway pressure (BiPAP) ventilatory system in providing pneumatic stabilization, whilst definitive chemotherapy was given to heal her pathological fractures.

Case report

An 84 year old, previously healthy female first presented elsewhere with complaints of bone pains and progressive dyspnoea over a 6 month period. She was found to have flail chest due to multiple bilateral lower rib fractures. There was no history of trauma prior to these symptoms. She was in respiratory distress and required urgent intubation and ventilatory support. She underwent tracheostomy on day 20 of hospitalization. Her clinical course was further complicated by recurrent nosocomial pneumonia and failure to wean from the ventilator. She was subsequently transferred to our intensive care unit (ICU) for further management on day 68.

At the ICU, paradoxical inward movements of the lower thoracic segments were evident bilaterally on inspiration. The patient was maintained on mechanical ventilation via the tracheostomy, which stabilized the flail segments. On pressure support ventilation of 15 cmH₂O and positive end-expiratory pressure (PEEP) of 5 cmH₂O, the arterial pH was 7.48, the arterial oxygen tension (PₐO₂) 10.4 kPa (78 mmHg), and the arterial carbon dioxide tension (PₐCO₂) 6.4 kPa (48 mmHg), whilst breathing 28% oxygen (fraction of inspired oxygen (FiO₂) 0.28). Plain chest radiographs showed generalized osteopenia, with multiple pathological rib fractures bilaterally. Multiple myeloma was established on the basis of multiple lytic bone lesions and bone marrow biopsy, which showed increased plasma cells with 80% myeloma cells, and serum protein electrophoresis confirming an immunoglobulin A (IgA) myeloma (IgA 63.3 g·L⁻¹, monoclonal band with anti-IgA and anti-kappa in serum samples). Urine was negative for Bence Jones proteins and there was no acute renal failure or renal impairment. The serum β₂-microglobulin level was elevated at 9,732 µg·L⁻¹ (normal range (NR) 878–2,000 µg·L⁻¹). There was an associated hypercalcaemia (serum calcium 2.85 mmol·dL⁻¹) and anaemia (blood haemoglobin 6.7 g·dL⁻¹). A radiographic bone survey of all the major bones revealed, in addition to the multiple rib fractures, the presence of a sternal fracture. There were no fractures outside the chest.

The patient was taken off the ventilator on day 75 (a week after arrival at the ICU) and started on BiPAP support (initial inspiratory positive airway pressure (IPAP) of 16 cmH₂O, and expiratory positive airway pressure (EPAP) of 5 cmH₂O) via the tracheostomy. Monitoring of the rib cage and abdominal movements demonstrated that the asynchronous paradoxical movements were abolishe d by BiPAP ventilatory support (fig. 1). At this stage, attempts to wean the patient caused the return of paradoxical move-
BiPAP in the high dependency ward for 2 months, during which period the myeloma was treated with three 4 day courses of melphalan-prednisolone chemotherapy given every 3 weeks for the rib fractures to heal. The BiPAP support was gradually tailed-off as her pathological fractures healed. At IPAP 8 cmH₂O and EPAP 5 cmH₂O and breathing room air, the pH was 7.47, $P_{\text{a,CO2}}$ 10.5 kPa (79 mmHg) and $P_{\text{a,CO2}}$ 5.5 kPa (41 mmHg).

The condition of the patient improved, as evidenced by callus formation on serial chest radiographs, improvements in her respiratory status, and a marked decrease in the IgA levels to less than half the original value. On day 128, she was weaned off BiPAP support and was able to breathe unaided. On breathing room air, the pH was 7.48, the $P_{\text{a,O2}}$ 8.7 kPa (65 mmHg) and the $P_{\text{a,CO2}}$ 5.3 kPa (40 mmHg). The flail segments healed and paradoxical motion was minimal. She was discharged on day 158, 3 months after arrival at our hospital, with further courses of chemotherapy given for her fractures to heal. The BiPAP was successful in reducing the work of breathing for the patient, whilst at the same time airway suctioning was possible through the tracheostomy. The patient was weaned by gradually diminishing the IFAP and EPAP levels, and, later, by taking her off the portable ventilator for longer and longer periods. All this was done out of the ICU setting, freeing up the ICU resources.

Gregoretti et al. [4] have recently reported the use of pressure control ventilation delivered through a minitracheostomy cannula as an alternative to conventional tracheal ventilation to treat severe flail chest. They found that this method could achieve a suitable gas exchange and provide stabilization in a flail chest condition, at the same time reducing the side-effects of tracheal intubation.

Our experience with this patient suggests that, despite its dramatic clinical manifestation, the association of flail chest with multiple myeloma may not predict a poor prognosis. In such cases, vigorous specific as well as supportive therapy is mandatory. We have also found that pneumatic stabilization can be achieved by using the bilevel positive airway pressure ventilatory support through a tracheostomy.

**Discussion**

Although skeletal involvement is common in multiple myeloma, spontaneous flail chest resulting from rib fractures is extremely rare. The occurrence of such a complication in a patient with multiple myeloma prompted this report, and a review of the literature. There have been only six previous reports of patients with pathological flail chest complicating myeloma [7–11]. The uniqueness of this patient was the fact that the myeloma, which was previously undiagnosed, first manifested as flail chest with respiratory failure.

Mild flail chest generally requires only supportive therapy. However, positive airway pressure, provided by continuous positive pressure ventilation, continuous positive airway pressure (CPAP), or a combination of intermittent mandatory ventilation and PEEP is necessary in patients with severe flail chest in order to diminish the paradoxical movements of the chest by internal stabilization [1, 2]. However, what was especially unique in this patient was the early use of BiPAP support, delivered through the tracheostomy, within a week of her transfer to the ICU, and its ability to provide a means of ventilatory support while definitive chemotherapy was given for her fractures to heal.

The BiPAP ventilatory support system is a lightweight, portable support ventilator developed for noninvasive use. It is based on a standard positive airway pressure, high-flow generator system and cycles in response to the patient’s respiratory flow, between preset levels of IPAP and EPAP. We believe that this is the first such case of the use of BiPAP ventilation through a tracheostomy for pathological flail chest complicating multiple myeloma. The BiPAP was successful in reducing the work of breathing for the patient, whilst at the same time airway suctioning was possible through the tracheostomy. The patient was weaned by gradually diminishing the IFAP and EPAP levels, and, later, by taking her off the portable ventilator for longer and longer periods. All this was done out of the ICU setting, freeing up the ICU resources.

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**References**