A serious fit of cough: A 66-year-old patient with myasthenia gravis presenting with spontaneous intercostal lung herniation after coughing. Case report and pathophysiological discussion

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Abstract

We present a case of a 66-year-old man with history of myasthenia gravis, severe obesity and osteopenia self-presented to our Emergency Department (ED) with severe pain in his left hemithorax, occurred after an episode of cough three days before. No history of trauma was reported. The patient underwent a chest-XR showing uncomplicated spontaneous fractures of the 5th and 6th left ribs. He was therefore discharged with appropriate analgesic treatment. Five days later, the patient came back to our ED for a wide left abdominal hematoma, though hemodynamically stable and eupneic. The only symptom he reported was some tenderness in the site of the rib fractures occurred on the same day. He denied once again any traumas. The physical examination was inconclusive: a wide hematoma was found in the left side of the abdomen; the left rib cage was tender at superficial palpation and the respiratory sounds were reduced in the inferior left lung field.

Ordinary laboratory tests were taken, showing normal hemoglobin levels, leukocytosis (WBC: 18.2 x 10³ μL), increased C-reactive protein (CRP: 281.3 mg/L) and procalcitonin (PCT: 0.34 ng/mL, normal value <0.05). The Arterial Blood Gas (A.B.G.) demonstrated a moderate respiratory insufficiency with chronic compensated respiratory acidosis (pH 7.4; pCO₂ 48.4 mmHg; pO₂ 89 mmHg; HCO₃⁻ 29; PaO₂/FiO₂=287). Coagulation and other blood tests were all in the normal range normal.

A plain CXR was performed, showing a displacement of the rib fragments, possible bibasal consolidations and a left pleural effusion (Figure 2). The lung ultrasound pattern was unclear, showing a diffuse ultrasonographic barrier effect.

In the suspicion of a hemothorax, a complete chest and abdominal CT-scans with contrast were performed, describing an unexpected picture of moderate intercostal lung herniation (2.3 cm) in the 5th space, a diffuse subcutaneous emphysema from the neck to the abdomen, a minimal left-sided pneumothorax and mild-to-moderate bilateral pleural effusion, partially loculated in the left side. The CT-scan also confirmed a non-refurnished abdominal wall hematoma and the known 5th and 6th ribs detached fractures (Figures 3-5).

Our patient underwent an urgent surgical consult, suggesting a conservative approach in a medical ward. The patient did not require non-invasive ventilatory supports, but we administered oxygen via face mask and nasal canulae. A prophylactic antibiotic regimen was started first with amoxicillin/clavulanic acid then switched to piperacillin/tazobactam on the 4th day. During the hospitalization, Hb values decreased to a minimum of 12.0 g/dL. No blood transfusions were prescribed. The patient was discharged after 15 days, with complete normalization of the clinical picture. Further CXRs in the following months described a complete spontaneous reduction of the hernia.

Case presentation

A 66-year-old man self-presented to our Emergency Department (ED) with severe and increasing pain in his left hemithorax, occurred after an episode of cough three days before. No history of trauma was reported. The patient had medical history of myasthenia gravis, severe obesity, ulcerative colitis and type-2-diabetes in chronic treatment with prednisone 12.5 mg/day, repaglinide and metformin. In 2006, he was diagnosed with osteopenia and a spontaneous vertebral compression fracture. The patient was also an active smoker.

He underwent a chest-RX (CXR) which demonstrated compound fractures of the 5th and 6th left ribs (Figure 1), classified as uncomplicated and spontaneous. The patient was therefore discharged with appropriate analgesic treatment.

Five days later, the patient came back to our ED for a wide left abdominal hematoma. The patient was hemodynamically stable and eupneic. The only symptom he reported was some tenderness in the site of the rib fractures occurred on the same day. He denied once again any traumas. The physical examination was inconclusive: a wide hematoma was found in the left side of the abdomen; the left rib cage was tender at superficial palpation and the respiratory sounds were reduced in the inferior left lung field.

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Discussion

Spontaneous rib fractures are a common complication in osteopenic patients affected by respiratory infections, particularly in those patients treated chronically with corticosteroids. The initial evaluation and management of spontaneous stress fractures of the ribs involve the ruling out of main complications such as pneumothorax and thoracic bleeding. Further, a correct pain management is required: an association of paracetamol, NSAIDs/COX-2 inhibitors and as needed opioids (i.e. codeine, tramadol, etc.) is suggested, with an increasing role of ultrasound-guided locoregional anesthesia. Since the surgical fixation is rarely needed, the usual approach considers early discharge and conservative therapy as first choice.

The late development of a thoracic and abdominal wall hematoma suggested a delayed intercostal hemorrhage. The patient never reported the typical signs or symptoms of intercostal lung herniation such as a palpable soft mass or worsening dyspnea. Though a thoracic wall hematoma is frequently associated with lung hernias, it’s unspecific for this condition. The initial physical examination was possibly misled by the severe obesity of the patient. Despite the attested sensitivity and specificity of chest ultrasound in the ED for pleural effusions, rib fractures and pneumothorax.

Figure 1. laying AP CXR (panel A) and left hemithorax (panel B) showing a compound antero-lateral fracture of the 5th and 6th left ribs (in the circle) and an elevation of the left hemidiaphragm.

Figure 2. Standing AP CXR showing possible detachment of the fracture lines (a), subcutaneous emphysema (b), left pleural effusion (c), possible bibasal consolidations and a prominent elevation of the left hemidiaphragm (d).
mothsorax and, therefore, it’s possible role in the detection of a lung herniation, the concomitant presence of obesity and subcutaneous emphysema reduced the diagnostic power of lung ultrasound. The first standing CXR was inconclusive. A CT-scan with contrast was therefore indicated in this condition. The CT-scan with contrast showed a self-limiting intercostal hemorrhage with no active bleeding, not needing for interventional hemostasis or blood transfusions. The CT-scan interestingly reported a lung herniation and subcutaneous emphysema, too. Lung herniations are rare findings in non-traumatic fractures, mostly associated with corticosteroid chronic therapies, primitive pulmonary affections, or previous invasive procedures. In our patient, the predisposing factor is likely to be the prednisone therapy. Corticosteroid-induced osteopenia is a common side-effect in chronic corticosteroid treatments; (in these patients, the bone samples show a lower density in osteoblasts and an increased rate of osteocyte apoptosis, with a general reduction in bone strength. On the other hand, glucocorticoid effects include an increase in osteoclasts lifespan).

Anyway, the rib fractures alone can’t explain the lung herniation, thus a global thoracic wall weakness needed to be present.

![CT-scan coronal sections in lung window (panel A) and soft tissue window (panel B) showing the pulmonary herniation (a), the minimal pneumothorax (b) and the diffuse subcutaneous emphysema (c).](image1)

![CT-scan transverse sections (cranial, panel A, and caudal including the spleen in panel B) showing the pulmonary herniation (a), the left sided loculated pleural effusion (b), the right sided pleural effusion (c) and the diffuse subcutaneous emphysema (d).](image2)
Actually, corticosteroid activity over muscle catabolism is well known: glucocorticoids inhibit protein synthesis while stimulating protein degradation, possibly leading to a skeletal muscle atrophy in several acute and chronic condition. Glucocorticoid-induced myopathy is a common complication in Cushing-like-syndromes which induce diffuse atrophy and substitution of muscle fibres, affecting both limbs and respiratory muscles.16,17

Severe obesity can be involved in the pathogenesis of lung herniation, too, both due to its increasing effect on intrathoracic pressure, and to a diffuse respiratory muscle overstretching and related weakness.18

Myasthenia Gravis (MG) affects the neuromuscular junction, resulting in a localized or diffuse muscle weakness which can involve the respiratory system. Many clinical conditions can exacerbate MG, as infections. Even if the idea of MG possibly involved in the pathogenesis of lung herniation is appealing, our patient didn’t show any sign of myasthenic crisis as dyspnoea, fatigue or severe respiratory insufficiency; thus, a direct involvement of MG is unlikely.19,20 The patient was treated conservatively. Though some authors suggest an early interventional approach,21 in minor hernias, a watch-and-see conservative regimen, based on pain management and rest, could be sufficient to obtain a complete spontaneous resolution. Admission to hospital in case of acute complications is suggested. In case of large hernias or severe complications, as hernial incarceration or strangulation, with lung necrosis, a surgical reduction is required both with invasive or thoracoscopic approaches.22 Surgery is also the treatment of choice in combined lung and intestinal hernias and lung hernias involving the diaphragm. Our patient presented a moderate intercostal herniation with a risk for acute medical complications and therefore an in-hospital medical treatment was needed, with no surgical indications.23

A clinical diagnosis of pneumonia could be debated. WBC count, CRP and PCT were mildly increased, suggesting a possible bacterial infection24,25. The patient never developed fever, and the serial hemocultures were constantly negative. The finding of pleural effusion, absent in the first hospital presentation while moderate at the second admission, could be consistent either with infection or with an inflammatory response. PCT was later negative after one week of antibiotic therapy.

Conclusions

The lung herniation is a rare either congenital or acquired condition, mostly following thoracic surgical procedures or thoracic traumas. Few cases of spontaneous lung herniation are reported in literature, most of them associated with rib fractures and persistent coughing, mechanical ventilation with positive pressure, emphysema and COPD. The surgical reduction of the hernia is the usual therapeutic option in these reported cases.

Our case depicts a spontaneous lung herniation associated with pneumothorax and subcutaneous emphysema after atraumatic rib bone fractures in a patient in chronic corticosteroid therapy for his history of myasthenia gravis. We successfully treated this patient with a conservative approach. The pathophysiological mechanism of the lung herniation in this patient is due to the side effects of a long-term glucocorticoid therapy on the thoracic wall, muscles and bones. MG is unlikely to be a causal factor.
References