The Macklin Effect in Recurrent Spontaneous Pneumomediastinum in Systemic Lupus Erythematosus & COVID19

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Introduction

- Spontaneous pneumomediastinum (SPM) is defined as the presence of free air in the mediastinum, with no clear traumatic etiology.
- Interestingly, pneumomediastinum was found in 1% of Covid related cases (Chen et al 2020).
- The Macklin effect, described in 1939, refers to centripetal alveolar rupture causing free air to track along the broncho-vascular sheaths towards the hilum of the lung and into the mediastinum (Murayama, 2014).
- With SPM seen in the setting of COVID, there are a few case reports correlated with immunosuppression.
- We describe the first case of the Macklin effect in a patient with systemic lupus erythematosus (SLE) and prior COVID-19 infection.

Case Report

- A 38-year-old male with a history of SLE and COVID-19 was admitted acute hypoxic respiratory failure.
- CT imaging displayed pneumomediastinum with subcutaneous emphysema tracking up to the orbital regions (Figure 1).
- His eyes were shut closed due to the extensive subcutaneous emphysema.

Figure 1: CT Torso revealed air around the orbits (A), as well as air around the left hilar region, around the pulmonary artery (B).

- The patient was discharged on steroids and antibiotics and improved briefly.
- After 1 month, he returned with worsening dyspnea, cough, and pneumomediastinum (Figure 2).
- EGD was performed and was unremarkable for an esophageal perforation.
- Bronchoscopy revealed severe tracheomalacia, but no airway defects.
- Bronchoscopic cultures were positive for MSSA, and he was started on broad spectrum antibiotics given the dense bilateral pulmonary infiltrates.

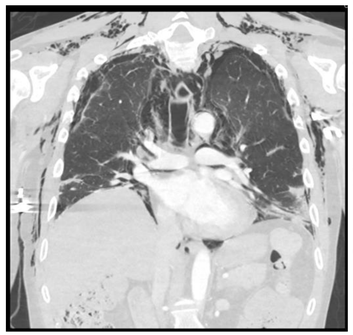


Figure 2: Repeat CT imaging was significant for developing significant and persistent subcutaneous emphysema, with air migrating along the broncho-vascular bundles around both hila, with air tracking into the mediastinum. Note the peripheral air bronchograms down the LLL.

- The patient eventually developed bilateral pneumothoraces (Figure 3), requiring chest tube placement, and eventual intubation with low tidal volume, low PEEP, and high FiO2.
- He was subsequently transferred to another center for VV ECMO, in which he eventually was discharged in stable condition after 1 month. No surgical repair was needed.

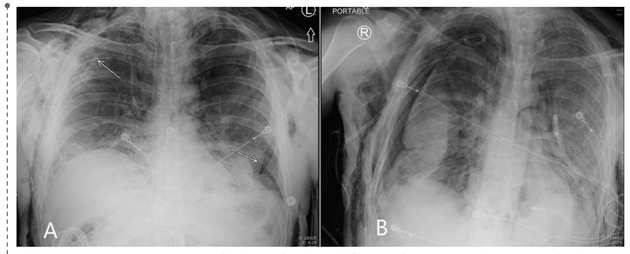
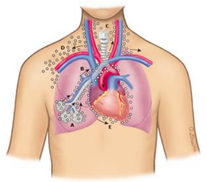


Figure 3: Image A reveals developing bilateral pneumothoraces (refer to the arrows). Image B was three days later, s/p bilateral chest tube placement.

Discussion

- This case illustrates a clinical scenario in which an immunosuppressed SLE patient with recent COVID infection presented with SPM, bilateral pneumothoraces, subcutaneous emphysema, and basilar superimposed pneumonias creating consolidation, air bronchograms, and suspected rupture of adjacent alveoli, triggering retrograde leakage of air that tracked into the mediastinum, neck, and face, consistent with the “Macklin effect”.
- This patient was likely susceptible to developing the Macklin effect due to his underlying immunosuppression predisposing him to pneumonia and his subsequent cough causing increased intrathoracic pressure.
- Furthermore, SLE may make the lung parenchyma more susceptible to damage possibly through an aberrant inflammatory response and alveolar injury from immune-mediated inflammation.
- Most patients are managed conservatively with bedrest, antibiotics, bronchodilators, steroids, high concentration oxygen, and/or careful watching.



- The treatment in this patient consisted of addressing the underlying infection and using high oxygen concentration, while avoiding positive pressure allowing time for the lungs to heal.
- In severe cases, air may dissect into other spaces leading to complications such as pneumothorax, pneumopericardium, and pneumorrhachis (free air in the spinal canal).
- Management for these complications may include VATS, thoracotomy, chest tubes, and/or skin incisions for decompression.

Conclusion

- This case illustrates the importance of being aware of complications such as SPM in post-covid patients, who are immunocompromised, particularly with SLE.
- Complications can range from rare, such as pneumorrhachis or pneumoperitoneum to more common, such as pneumothorax.
- As the Macklin effect is not commonly identified, this is a case to bring to the forefront of our clinical minds.

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