SELFISH LUNG SYNDROME

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WHAT IS CONGENITAL LOBE EMPHYSEMA (CLE)?

- Rare congenital developmental anomaly characterized by hyperinflation of one or more pulmonary lobes.
- Prevalence: I out of 20000/30000. Higher incidence in males.
- Etiology is unknown.
- Most frequently identified cause is obstruction of developing airway, occurs in 25%.
- Obstruction can be intrinsic or extrinsic.
- Intrinsic causes: bronchomalacia or absent cartilage, causing alveolar distention.
 - This creates compression atelectasis \rightarrow ventilation perfusion mismatch
 - Air trapping on expiration due to a ball-valve effect.
- Extrinsic compression can be caused by vascular anomalies or intrathoracic masses.
- Left upper lobe affected most often, 40-50% of the cases.

CASE REPORT

• Patient is a 55 year old male, with a hx of CLE and tobacco abuse, who comes to the ED with dyspnea and cough. He was in mild distress, tachypneic, and oxygen saturations were in the 70s. He had a barrel-shaped chest wall and some expiratory wheezes were noted as well. He was not on oxygen at home and did not follow up with a pulmonologist. Pt denied having any surgical intervention for his congenital malformation other than bronchoscopy as a child.

IMAGING











IMAGING

CT chest with contrast:

- Massive congenital lobar overinflation/emphysema involving the entirety of the left lung with mass effect on the right lung and deviation of the anterior junction line by approximately 8 cm to the right.
- Additional airspace opacities within the left lower lobe with fluid/fluid level within some of the left-sided bullae, raising the possibility of pneumonia.

CASE REPORT

- Pulmonology consulted.
- Cardiothoracic surgical team consulted
 - Recommended left pneumonectomy
- Patient chose to pursue outpatient surgery.
- At the time of discharge: hemodynamically stable, saturating above 90%, at baseline.
- Unfortunately, patient was lost to follow up.

CONCLUSION

- Patient was mostly asymptomatic throughout his life.
- Lungs continue to deteriorate.
- Perhaps if he had regular follow up with a pulmonologist, the damages to his lung could have been limited.
- Early surgical intervention has been found to significantly improve the patient's condition. However, number of lobes affected can also determine the outcome of surgery.
- Remind patients the importance of monitoring and follow up regardless of symptom status.

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