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Recurrent Primary Spontaneous Pneumothorax Masquerading as a Congenital Pulmonary Airway Malformation in a Young Female

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Research Abstract Title: Case Report: Recurrent Primary Spontaneous Pneumothorax Masquerading as a Congenital Pulmonary Airway Malformation in a Young Female

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IRB Number: None – case report not requiring IRB approval

Describe role of Submitting/Presenting Trainee in this project (limit 150 words):
Chandra Swanson completed the literature review, chart review, and the initial first draft of this case report.

Background, Objectives/Goal, Methods/Design, Results, Conclusions limited to 500 words

Background: Spontaneous pneumothoraxes in children are uncommon, may be idiopathic or associated with underlying pulmonary disease, and can present management challenges.

Objectives/Goal: We present a 12-year-old female with recurrent right sided spontaneous pneumothorax in the setting of an asymptomatic SARS-Co-V2 (COVID) infection and imaging concerning for congenital lobar overinflation (CLO) versus congenital pulmonary airway malformation (CPAM), prompting surgical intervention.

Methods/Design: A literature review and chart review were completed for this case report.

Results: A 12-year-old pre-menstrual female with remote history of eczema, asthma, and environmental allergies presented from an outside facility with four-days of progressive chest pain and dyspnea on exertion and diagnosis of right-sided spontaneous pneumothorax, improving after pigtail chest tube placement. Physical exam was significant for tall thin body habitus; family history was significant for paternal spontaneous pneumothorax as an adolescent. She was incidentally found to be COVID positive. Chest plain films (CXRs) showed subcutaneous emphysema and persistent right-sided pneumothorax. Clamping trial failed, prompting removal of the pigtail and placement of 12F chest tube with resultant near complete re-expansion. On serial CXRs, a right hilar cystic lucency was newly identified. Chest CT confirmed the right upper lobe (RUL) air-filled

cystic structure and abrupt narrowing of the RUL posterior segmental bronchus, concerning for CLO versus CPAM. Chest tube was successfully removed on day 6, and she was discharged home with planned follow-up. Three months later, she was readmitted for recurrent right-sided spontaneous pneumothorax diagnosed after one day of chest pain, cough, and dyspnea. Laboratory testing revealed mild leukopenia and anemia; she was COVID negative. A chest tube was placed and set to wall suction. Due to persistent pneumothorax, this was replaced with a pigtail drain on day 5. CXRs demonstrated persistent cystic RUL lung mass. With her prior COVID infection now resolved, RUL wedge resection was completed via video-assisted thoracoscopic surgery on day 8. She tolerated the procedure well and was discharged on day 10 with resolving pneumothorax. Tissue for pathology results revealed pleural fibrosis and focal hemorrhage without malignancy, most consistent with a ruptured bleb. At one week follow up, she remained stable without complications.

Conclusions: Pneumothoraxes in tall, thin adolescents are often categorized as primary spontaneous. Most pneumothoraxes resolve with conservative management and often do not require surgical intervention. Congenital lung malformations are a rare secondary cause in children and may be detected on CXR. Chest imaging should be carefully reviewed for congenital malformations requiring specific surgical intervention. These findings, along with the patient's clinical course, may assist in determining management.