

Diffuse Alveolar Hemorrhage as a complication of Churg Strauss

Lovitt C., Mermis J.

Background

Churg Strauss (CSS) is a multi-system disorder characterized by allergic rhinitis, asthma, and prominent peripheral blood eosinophilia. This disease is also classified as a vasculitis of small and medium sized arteries. CSS represents approximately 10% of diagnosed vasculitides. This disease is known to have multiple system effects including pulmonary infiltrates, mono-neuritis multiplex, pericarditis, GI manifestations and skin manifestations.

Case Report

A 31 yo female presented to pulmonary clinic after being referred for possible cystic fibrosis. She had been diagnosed with asthma in the past and had multiple pulmonary complaints with frequent exacerbations, as well as gastrointestinal complaints. She had frequent courses of steroids for asthma exacerbations. The work up for CF was negative but further work up revealed she had Churg Strauss. Following this diagnosis she began having pulmonary symptoms including cough, shortness of breath and hemoptysis. She was initially treated with high dose steroids as well as broad spectrum antibiotics and began to improve. However, she again decompensated and required intubation. When the ET tube was placed, pulmonary suction revealed frank blood, her hematocrit had dropped from 37 to 27, and chest X-ray showed dense bilateral alveolar opacities. Subsequent CT showed scattered bilateral pulmonary infiltrates and mild peribronchial thickening and were consistent with Diffuse alveolar Hemorrhage (DAH). Her treatment was escalated to cyclophosphamide along with steroids. Over the course of 4 weeks and treatment with cyclophosphamide, this young patient slowly recovered and was eventually discharged from the hospital.

December - February

Patient presented to clinic with uncontrolled asthma, she was on oral steroids and had recently been started on montelukast.

Labs December
WBC 5.6 with 15 % eosinophils
Hgb/Hct: 13/39

Labs Early February
P-ANCA:160
Myeloperoxidase AB: 7.6

PFTs December

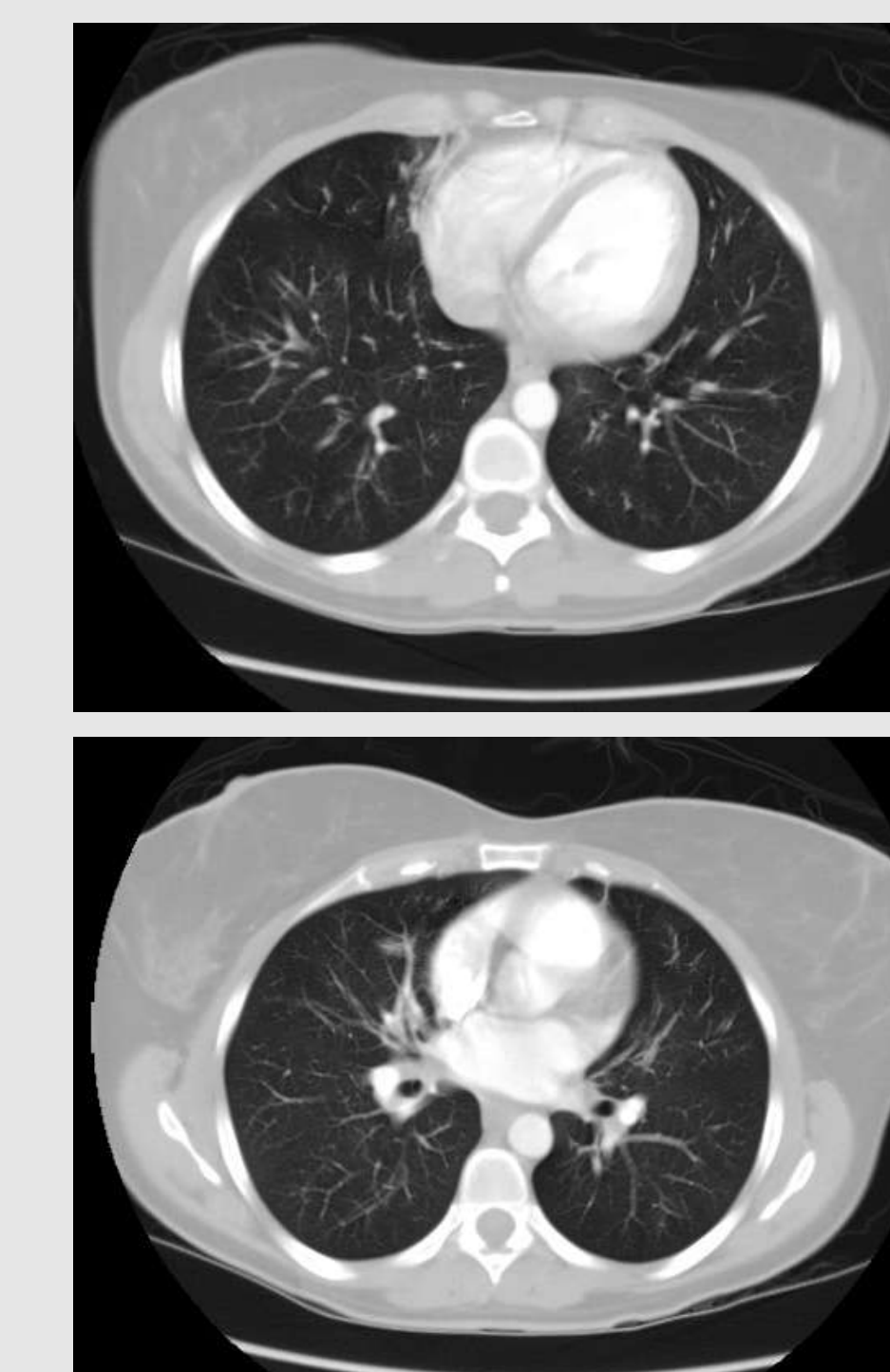
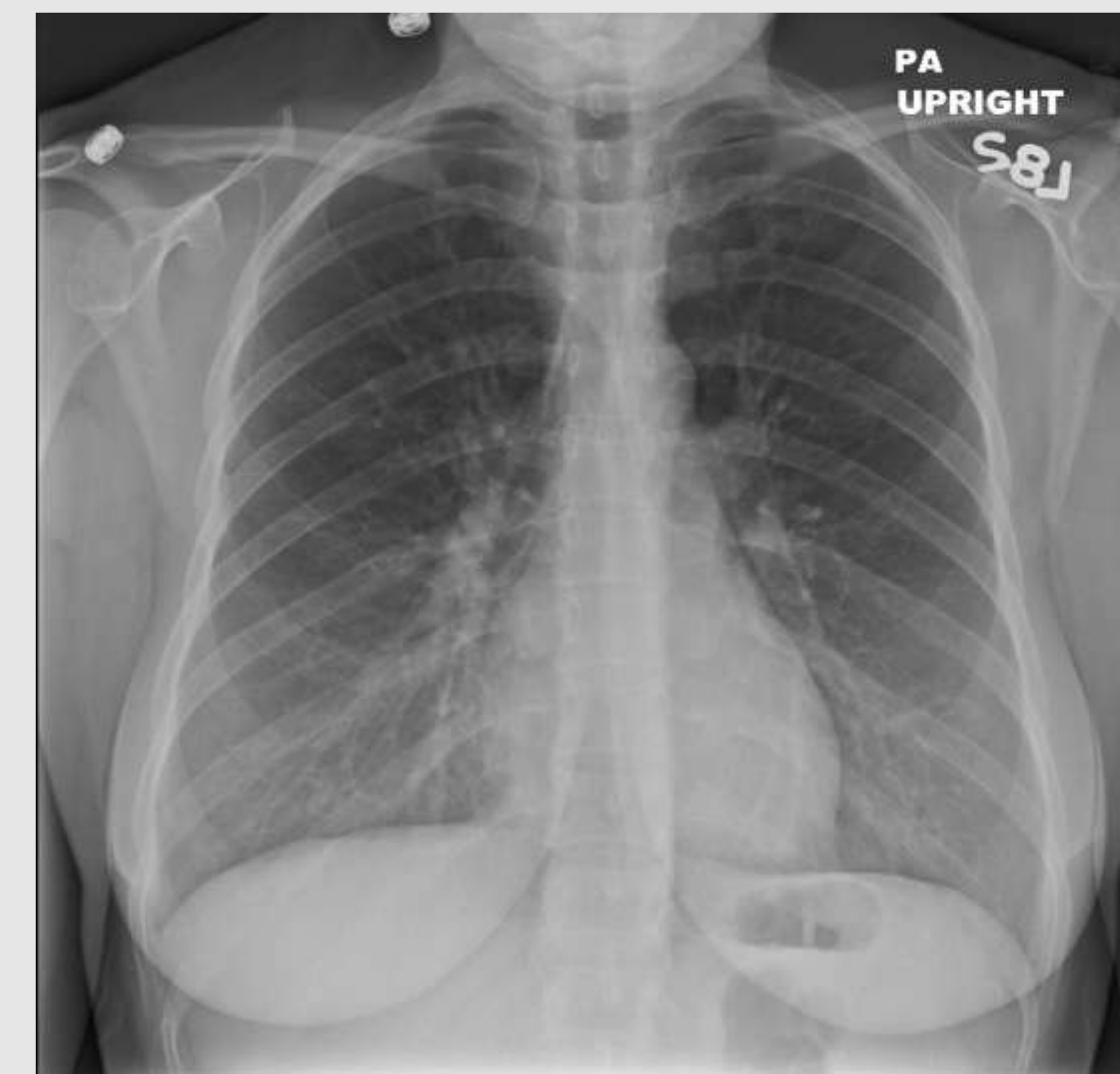
@SPIROMETRY@	PRE-BRONCH		%Prd	POST-BRONCH	
	Meas	Prd		Meas	%Prd
FVC (L)	2.31	3.19	72		
FEV1 (L)	1.64	2.82	58		
FEV1/FVC (%)	71	88	81		
FEF 25% (L/sec)	3.35	5.23	64		
FEF 50% (L/sec)	1.55	4.47	35		
FEF 75% (L/sec)	1.13	1.88	60		
FEF 25-75% (L/sec)	0.90	3.59	25		
FEF Max (L/sec)	4.26	5.73	74		
FVC (L)	1.62				
FIF 50% (L/sec)	2.72	3.82	71		
FIF Max (L/sec)	2.76				

Interpretation:
Spirometry demonstrates moderately severe obstructive defect with a mild reduction in vital capacity.

She then presented with hypoxia and hemoptysis.

Labs February 18th
Hgb/Hct: 13.9/40
ABG 7.37/34/61

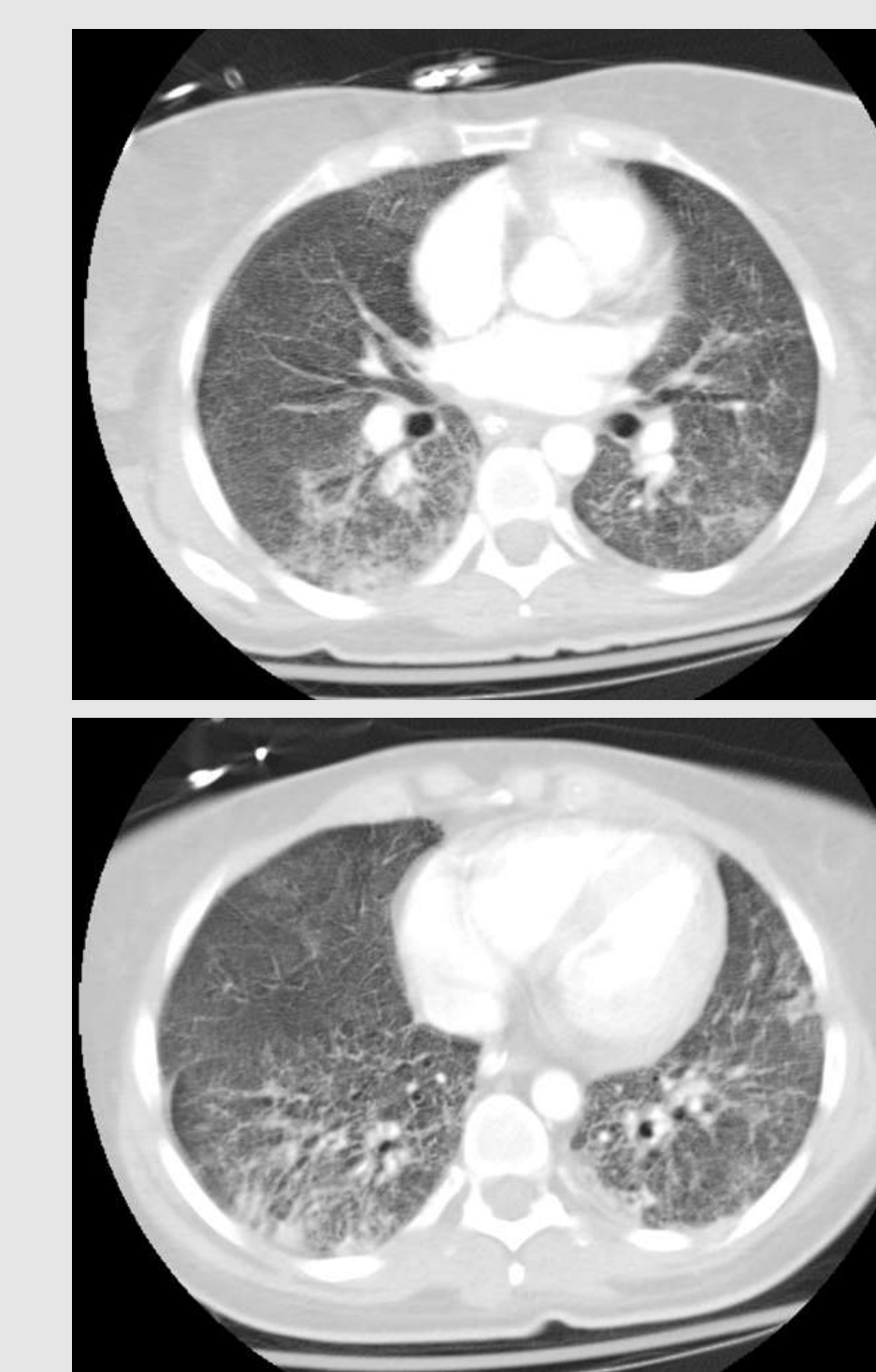
Chest X ray February 18th



Within four days she rapidly declined with respiratory distress requiring intubation.

Labs February 22nd
Hgb/Hct: 10/27

Chest X ray February 22nd



Discussion

DAH is characterized by hemoptysis (though this is not infrequently absent), fevers, chest pain, cough, dyspnea and hypoxic respiratory failure. Laboratory examination shows falling hematocrit. Imaging reveals a patchy, focal or diffuse alveolar filling process, and CT confirms this alveolar filling process and is more detailed as to the extent of the disease process. For the diagnosis of this disease, flexible bronchoscopy should be done and will show progressively hemorrhagic BAL in serial samples. DAH is known to be associated with IPH, ANCA-Associated Granulomatous Vasculitis, MPA, SLE, mixed connective tissue disorders, and Goodpasture Syndrome, per Schwarz and Lara in May 2010. A case report from 1998 examined a patient with CSS who presented with respiratory symptoms and transbronchial biopsy revealed necrotizing vasculitis. On autopsy they discovered this patient had DAH, and their conclusion emphasized that CSS should be considered in patients who are found to have ANCA positive vasculitis and DAH. This case also supports that a patient with known CSS is at risk for DAH, and the early recognition of this will aid in rapid diagnosis and treatment of this frequently fatal complication.

References

1. Guillevin, L., Pagnoux, C., Mouthon. Churg Strauss. *Semin Respir Crit Care Med* 2004; 25(5): 535-545
2. Lai, R., Lin, S., Lai N., Lee P. Churg-Strauss Syndrome Presenting with Pulmonary Capillaritis and Diffuse Alveolar Hemorrhage: Case report. 1998 *Vol 27, No. 3, 230-232*
3. Lara, A., Schwarz, M. Diffuse Alveolar Hemorrhage. *Chest* May 2010 vol 137 no. 5 1164-1171