A Rheumatologic Disorder Mimicking Asthma

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Background

- Relapsing polychondritis (RP) is rare and in some cases fatal autoimmune disease characterized by inflammation of cartilaginous structures and end organs (i.e. ears, nose, joints, eye, vestibular system, heart, kidneys and tracheobronchial tree).
- The diagnosis is difficult and often overlooked because of the fleeting nature of symptoms and the lack of serological markers.
- We present a case of RP mimicking symptoms of asthma.

Clinical Presentation

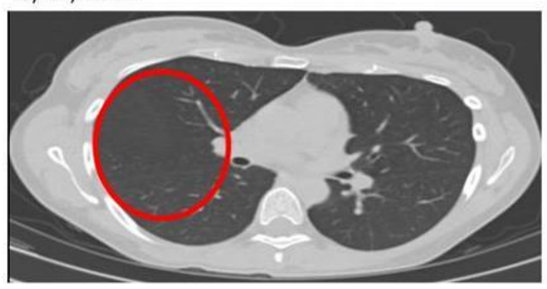
- 36-year-old woman with no past medical history developed a progressive dry cough over 3 months following an upper respiratory tract infection.
- Initial diagnosis was presumed to be post viral reactive airways disease. She was treated with a five-day course of prednisone and albuterol inhaler.
- Her cough initially improved but she experienced worsening dyspnea. Fluticasone with albuterol therapy for an additional 2 months did not improve her symptoms. She was then started on fluticasone/salmeterol and later montelukast without clinical improvement.
- The patient developed fevers, myalgias, worsening cough and dyspnea. Prednisone (60 mg) was empirically started resulting in symptomatic improvement.
- Attempts to taper the prednisone resulted in reemergence of significant symptoms with new pleuritic chest pain and chest wall tenderness.
- Review of systems revealed episodes over several years of voice loss lasting 3-5 days, hand arthralgias, and more recently a red nose and severe self-limited ear pinnae pain.
- Bronchoscopy did not demonstrate inflammation or tracheomalacia.
- Patient required high doses of steroids to control her symptoms after the bronchoscopy. She was started on tofacitinib and methotrexate with excellent clinical response.

Pulmonary Function Test

Date	FEV1/FVC	TLC	RV/TLC	FEV1 pre-BD	FEV1 Post-BD	FVC Pre-BD	FVC Post-BD
7.8.2015	96 (111% of predicted)	4.3L (96% of predicted)	45% (145% of predicted)	2.26L (85% or predicted)	1.71L (64% or predicted)	3.71 L (75% of predicted)	3.71 L (75% of predicted)
7.29.2015	83 % (97 % of predicted)	4.2L (94% of predicted)	50% (159% of predicted)	1.76L (66% of predicted)	1.23L (46% of predicted)	2.12L (75% of predicted)	1.49 (47%% of predicted)

CT Scan of the Chest

6/24/2015



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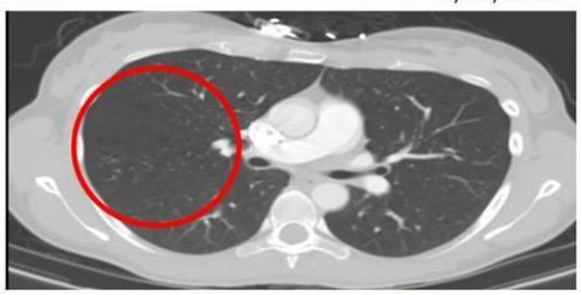

Summary of Published RP Cases Presenting as Asthma (n=11)

Age	Sex	Symptoms	Time from Asthma dx to RP dx
31-83 years old	54% Female	Cough, SOB, voice changes, anterior neck pain	6 months – 12 months

Diagnosis Criteria for RP

Clinical Symptoms (CS)	McAdam's Criteria	Damiani's Criteria
• Bilateral auricular chondritis • Nasal cartilage inflammation Respiratory tract chondritis • Non-erosive seronegative polyarthritis • Ocular inflammation • Audiovestibular involvement	3 of 6 symptoms	• 3 CS • 1CS + histologic confirmation • 2 CS + positive response to treatment

Clinical Investigations

Physical examination

- Pulmonary auscultation revealed a prolonged expiratory phase.
- An aortic regurgitant murmur was not heard.
- The musculoskeletal evaluation confirmed a tender left knee effusion by way of the bulge sign, a right knee effusion by way of ballottement, and costochondral and bilateral wrist tenderness.
- There was no saddle nose deformity, no cauliflower ear and no evidence for mononeuritis multiplex.

Laboratory evaluation

- The ESR was 8 mm/hr and renal studies were normal.
- Eosinophilia was not evident on the peripheral blood smear.
- The following tests were all negative or normal: ANA, ANCA (with myeloperoxidase and proteinase-3 specificity), anti-dsDNA, complement studies, anti-ENA, cryoglobulins, anti-glomerular basement membrane, anti-CCP, rheumatoid factor, anticardiolipin antibodies, lupus anticoagulant, hepatitis B S Ag, anti-hepatitis C, anti-HIV, RPR and the lymphocyte stimulation assay for TB.

Conclusions

- Pulmonary manifestations of RP are common and can include obstructive lung disease and air trapping.
- Airway compromise due to tracheomalacia or epiglottic stenosis can be a fatal complication of RP.
- Early recognition of lung involvement in RP can help improve morbidity and mortality due to complications of unchecked inflammation.
- This case represents the importance of including RP in the differential of patients with atypical asthma or new onset obstructive lung disease resistant to conventional therapy.

***References available upon request