Pulmonary Manifestations of Ankylosing Spondylitis

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ETIOLOGY

- AS is a chronic multisystem disease characterized by inflammation of the spine, sacroiliac and peripheral joints as well as involvement of extra-articular organs including the lungs and the heart.

- Chronic inflammation of the spinal structures, costovertebral, apophyseal, and sacroiliac joints leads to fibrosis and ossification of these structures.

- Consequently, spinal mobility is reduced and there is considerable limitation of rib cage expansion.

- AS affects approximately 0.1% of the general population and is more common in men than in women.

- Its etiology is unclear.

- There is a genetic predisposition for AS, as 95% of whites with AS have the HLA-B27 antigen.
**CLINICAL FEATURES**

- Presents in patients in late adolescence or early adulthood, present with back pain or morning stiffness due to involvement of sacroiliac joints. Onset of symptoms after the age of 45 is rare.
- Typically, the symptoms are worse in the morning or after prolonged rest.
- About 30% of individuals with AS have involvement of a peripheral joint or nongranulomatous anterior uveitis. Aortitis and dilatation of the aortic root may occur in up to 25% of patients.
- On physical examination, there may be tenderness of the anterior chest wall, the costochondral region, or the manubriosternal junction.
- Upper airway obstruction due to cricoarytenoid cartilage involvement is a rare complication.
Inflammatory lesions of the anterior chest wall displayed by whole-body MRI in an ankylosing spondylitis patient complaining of anterior chest wall pain. Arrows point to bone marrow edema in both parts of the manubriosternal joint.
Individuals with advanced AS have very limited rib cage motion and are more dependent on diaphragmatic breathing.

Prominent abdominal excursion during inspiration may be apparent in supine or upright individuals.

The degree of rib cage immobility can be assessed by measuring the change in rib cage circumference at the level of the fourth intercostal space between a full inspiration and full expiration.

Rib cage expansion less than 2.5 cm should raise the possibility of AS in young patients with chronic low back pain.
Limited rib cage expansion is the pathophysiological hallmark of AS. This limitation results from fusion of the costovertebral and sternoclavicular joints and possibly leads to intercostal muscle atrophy.

Rib cage motion is similar to that in healthy individuals in terms of direction, but the extent of movement is diminished.

Rib cage immobility leads to a reduction in chest wall and total respiratory system compliance but lung compliance is generally normal.

Only mild reductions in VC or TLC (75%-80% of predicted) may be present despite moderately severe reductions in rib cage mobility.

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Diaphragmatic strength is typically intact in these patients.

Chest wall inflation is primarily accomplished through diaphragmatic displacement of the abdomen.

The increased diaphragm shortening and the relatively greater transdiaphragmatic pressure required to inflate a stiff rib cage may potentially provide a training stimulus to the diaphragm.
Changes in anteroposterior dimensions of the rib cage (RC) and abdomen (Ab) in a healthy individual and one with ankylosing spondylitis (AS). There is limited mobility of the rib cage in all positions resulting in greater motion of the abdomen relative to the rib cage.
In the absence of parenchymal lung disease, PaO₂ is either in the normal range or mildly reduced. Exercise capacity may be mildly decreased in patients with AS, especially in those with marked rib cage restriction.

The mechanism of limited exercise is most likely related to peripheral deconditioning rather than ventilatory constraints.
PLEUROPULMONARY ABNORMALITIES

- Upper lobe fibrobullous disease occurs in a small percentage up to 4%.
- It is more common in males with long-standing disease and may manifest as interstitial infiltrates, fibrosis with honeycombing, or cavititation that may mimic tuberculosis.
- The pathogenesis of upper lobe fibrobullous disease is unknown.
- Possible mechanisms include decreased upper lobe ventilation, mechanical stress due to rib cage rigidity, and recurrent lung infection due to impaired cough.
- Individuals with fibrobullous disease have an increased propensity for spontaneous pneumothorax and infections with Aspergillus or atypical mycobacteria.
Fibrobullous disease tends to be progressive and its course is not altered by steroid treatment.

Resection of the lung segment with fibrobullous disease can be complicated by bronchopleural fistula in 50% to 60% of patients.

Surgery is reserved for treatment of massive hemoptysis.

Additional pleuropulmonary abnormalities that can be detected by chest CT include interstitial lung disease, pleural thickening, parenchymal bands, and bronchial wall thickening.

These abnormalities are subtle and do not correlate with clinical or functional impairment.
SLEEP DISORDERED BREATHING

- An increased prevalence of obstructive sleep apnea in AS has been reported, especially in patients with long-standing disease (>5 years).
- Because fatigue is a common complaint in AS, clinicians should have a high index of suspicion for concomitant sleep disordered breathing contributing to excessive daytime sleepiness and screen these patients with polysomnography.
Treatment of individuals with AS should incorporate pain relief, physical therapy, and measures to maintain posture.

Physical therapy exercises that promote rib cage expansion have beneficial effects on pain and spinal mobility. Smoking should be avoided and baseline chest radiographs and spirometry obtained.

The use of antagonists of tumor necrosis factor (TNF) has had a dramatic effect on pain control, fatigue, spinal flexibility, and quality of life in these patients.

Rib cage expansion is also improved following treatment with biological agents.
An adverse effect of the anti-TNF therapy is reactivation of tuberculosis.

AS patients who are candidates for anti-TNF therapy should be screened for latent tuberculosis and receive prophylactic treatment with isoniazid before starting treatment.

Additional rare adverse pulmonary complications of anti-TNF blockade include interstitial lung disease and development of a sarcoid-like disorder.