Updates on Diffuse Lung Disease

This first presentation by Dr. Bellina was a chart review of patients with idiopathic inflammatory myopathies and overlapping interstitial lung disease seen at a single center. They had 32 patients whose charts they reviewed and really presented characteristics of the patients' demographics as well as their treatment and their disease progression.

What they found was that the overwhelming majority, 70% were female and about 48% of them presented with their interstitial lung disease prior to the development of the myositis. They were treated with a variety of immunosuppressive medications including azathioprine and mycophenolate. And about 30% of those patients progressed to the use of rituximab for either failure of one of the other agents or disease progression or inability to tolerate an oral agent. The good thing about this is that only about a third of those patients had clinical worsening over the follow-up period. The key takeaway here is that we need to have an index of suspicion.

In this cohort, almost 50% of patients presented with just interstitial lung disease before the development of myositis. And so we have to maintain that index of clinical suspicion even after a diagnosis is made for other entities that can develop.

This next session was presented by Dr. Zeba and was a retrospective cohort study of 82 patients at a single center. She looked at patients with combined pulmonary fibrosis and emphysema versus those patients with idiopathic pulmonary fibrosis and compared their echocardiographic and radiologic findings that predicted the existence of pulmonary hypertension, as well as outcomes with that pulmonary hypertension. What they found there was that the right inferior pulmonary vein diameter and the left ventricular short axis diameter were predictive of development of pulmonary hypertension in combined pulmonary fibrosis and emphysema. In addition, the pulmonary artery to ascending aorta diameter ratio greater than one was associated with decreased survival in both cohorts of CPFE and IPF.

The take home point here is that there are non-invasive measures that may help us with prognostic information for both patients with idiopathic pulmonary fibrosis and combined pulmonary fibrosis and emphysema, as these patient populations are likely to develop pulmonary hypertension.