Hughes-Stovin Syndrome

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Introduction

Hughes-Stovin Syndrome (HSS) is an autoimmune disorder, that presents with Deep Venous Thrombosis (DVT) and pulmonary and or bronchial aneurysms.

Case Presentation

49-year-old man was referred to outpatient clinic with dyspnea that started 4 years ago, but his symptoms got worse in the previous year [functional class II of New York Heart Association (NYHA)]. He reported no chest pain, hemoptysis or sudden onset of dyspnea. He complained of intermittent fever since 3 months ago. His medical history revealed that he was a heavy smoker (40 packs/year) and had a deep vein thrombosis in left leg two months ago. He received warfarin 5 mg/d and his INR (International Normalized Ratio) was in ideal range.

His viral signs were stable except oxygen saturation of 89% on room air and decreased breath sounds in both lungs. Another exam was intact. There was obstructive pattern in spirometry. His chest computed tomography scan showed large central pulmonary arteries (Figure 1) with multiple aneurysmal dilatation of...
right and left pulmonary arteries (Figures 2 and 3) The final diagnosis was Hughes-Stovin syndrome.

Discussion

Hughes-Stovin Syndrome (HSS) is a rare autoimmune disorder, characterized by Deep Venous Thrombosis (DVT) and pulmonary and or bronchial aneurysms. The pathogenesis is not clear, but it is assumed to be a consequence of angiodysplasia and vasculitis similar to those in Behcet’s Disease (BD) [1]. If a patient presents with this set of findings (aneurysms and thrombosis) and the clinician is able to rule out other causes, then the patient either has HSS or BD [2]. The management of HSS is medical (steroid and cytotoxic agent) and not surgical.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles were considered in this article. The participant was informed about the purpose of the research and its implementation stages.

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Conflict of interest

The authors declare no conflict of interest.

References
