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Pulmonary vasculitis in Hughes Stovin syndrome: Current status and future perspectives

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Hughes-Stovin syndrome (HSS) is a severe systemic vasculitis characterized by widespread venous/arterial thrombosis and pulmonary artery aneurysms (PAAs). All fatalities reported in the HSS were the result of unpredictability fatal suffocating hemoptysis. As a result, pulmonary complications must be identified at an early stage of the disease.

The HSS International Study Group reference atlas categorizes pulmonary vasculitis in HSS into six stages and defines the various radiological patterns of pulmonary vasculitis, most notably pulmonary artery aneurysms detected by computed tomography pulmonary angiography (CTPA). HSS International Study Group reference atlas describes the CTPA images that best define the broad range of pulmonary vasculitis seen in HSS. Pulmonary aneurysms were classified into six radiographic patterns: true stable PAA with adherent in-situ thrombosis, unstable leaking PAA, BAA, and/or PAP with loss of aneurysmal wall definition (most prone to rupture), CTPA images demonstrating right ventricular strain and intracardiac thrombosis.

The classification's main goal is to provide physicians with information about this rare syndrome. Since Hughes and Stovin first description of the syndrome in 1959, no such scheme has been proposed. This classification will serve as the foundation for future recommendations on the diagnosis and treatment of this syndrome.

The presentation will go over the most important CTPA findings that can occur in HSS-related pulmonary vasculitis and included very illustrative CTPA images of various pulmonary aneurysmal lesions.

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