Hughes-Stovin Syndrome

Michael Kindermann, MD; Heinrike Wilkens, MD; Wilfried Hartmann, MD; Hans-Joachim Schäfers, MD; Michael Böhm, MD

A 50-year-old woman suffering from shortness of breath and unproductive coughing for 6 months was admitted to the hospital because of recurrent hemoptysis. The patient had a history of deep venous thrombosis of both legs and the pelvic veins. A thrombophilic state was assumed because of a factor V Leiden mutation.

Angiography showed occlusion of both iliac vein systems and of the vena cava inferior (Figure 1). Venous access to the central circulation was possible through the superior caval vein. Although pulmonary angiography of the left lung showed some enlargement at the origin of the left upper-lobe pulmonary artery, right-sided pulmonary angiography revealed two large aneurysms originating from the right upper-lobe artery and the intermediate branch of the pulmonary artery (Figure 2).

The intermediate artery aneurysm was resected, and the artery was reconstructed using a saphenous vein graft. The patient also underwent right upper lobectomy because the right upper-lobe aneurysm reached too far into the periphery to allow reconstruction. Histological examination of the specimen showed vasculitis, and immunosuppressive therapy was started.

The Hughes-Stovin syndrome consists of deep venous thrombosis often involving the caval vein accompanied by single or multiple pulmonary arterial aneurysms. Massive pulmonary hemorrhage caused by rupture of an aneurysm is a frequent terminal event. Patients affected by this syndrome are often young adult men in the second to fourth decade of life. Case reports describing women with Hughes-Stovin syndrome are rare.
Hughes-Stovin Syndrome
Michael Kindermann, Heinrike Wilkens, Wilfried Hartmann, Hans-Joachim Schäfers and Michael Böhm

Circulation. 2003;108:e156
doi: 10.1161/01.CIR.0000106680.35316.16
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2003 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/108/23/e156

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2003/12/08/108.23.e156.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/