Hughes-Stovin Syndrome with Pulmonary Angiitis and Focal Glomerulonephritis

A Case Report With Necropsy Study

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A clinicopathologic case of Hughes-Stovin syndrome with pulmonary eosinophilic angiitis and focal prolifeering extracapillary glomerulonephritis is reported.

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In 1959 Hughes and Stovin reported the occurrence of hemoptysis due to ruptures of aneurysms of segmental pulmonary arteries in four patients (including two from the literature) that had previously had recurrent peripheral venous thrombosis. These authors noticed, moreover, that symptoms of increased intracranial pressure were prominent features of the early stages of the illness and proposed that the aneurysms of the pulmonary arteries might be due either to degenerative changes of the bronchial arteries or to infected emboli. The etiopathogenesis of the so-called Hughes-Stovin syndrome remains obscure, however, since the findings in most of the cases reported after the original description (only ten cases have been collected in a recent review of the literature) do not seem to confirm any of the aforementioned hypotheses. We report another case of Hughes-Stovin syndrome in which pulmonary angiitis and focal glomerulonephritis were observed.

CASE REPORT

A 33-year-old man was admitted to the Hospital S. João in May 1974 because of abrupt onset of diplopia and severe headaches. He had bilateral papilledema and right external rectus paresis. The WBC was 12,300/cu mm, with 63.5 percent neutrophils, and the ESR was 35 mm/hr. Apart from slightly raised pressure, the CSF was normal. Skull roentgenograms, EEG, echoencephalography, and cerebral arteriography did not show any abnormality. He was discharged two months later without diplopia. No diagnosis was established.

During the ensuing 18 months, the patient noted frequent bouts of headache that responded only to ergot preparations. In March 1976 he began to have recurrent episodes of thrombophlebitis in the lower limbs.

He was readmitted in December 1976 with left saphenous vein thrombophlebitis and acute pyelonephritis of the left kidney. Chest roentgenogram was normal. Superficial thrombophlebitis recurred twice during hospitalization. Migrating muscular pain, three episodes of conjunctivitis, and an acute pneumonia of the right lung also occurred during this hospitalization. Repeated blood cultures were negative. No histologic abnormalities were found in skin and muscle biopsies, and tests for antinuclear antibodies were negative. Electrophoresis and immuno-electrophoresis of serum proteins did not show any abnormality. He was discharged...
three months later without diagnosis.

In February 1978 he was readmitted because of hemoptysis. Chest roentgenograms and tomograms revealed a large, round opacity in the right parahilar area suggesting a lung tumor (Fig 1). Neoplastic cells were not found in repeated sputum specimens. Bronchoscopy showed that the left common basilar bronchus was narrowed by external pressure and the biopsy revealed only inflammatory changes. There was too much bleeding in the right bronchus, and no biopsy was done. Three months after the admission the patient died suddenly of a massive hemoptysis.

**Main Autopsy Findings**

Two sacciform aneurysms were found: one in a segmental branch of the inferior lobar right pulmonary artery and the other, which had perforated into the adjacent bronchus, in the inferior lobar branch of the left pulmonary artery. There was partially organized intra-aneurysmatic thrombosis. In some areas of both aneurysms destruction of most of the media of the artery and inflammatory exudate also were seen.

The pulmonary arteries showed widespread lesions of marked medial hypertrophy and severe, often obliterating, post-thrombotic intimal fibrosis in which there was a very dense infiltrate of eosinophil leukocytes (Fig 2). Similar eosinophil infiltrates were seen in the intimal fibrosis of some pulmonary veins as well as diffusely dispersed throughout the pulmonary interstitium, but were not observed in the segmental branches of the pulmonary arteries. A peripheral infarction was found in the lower lobe of the left lung; there was neither noticeable lesions of bronchial arteries nor right ventricular hypertrophy.

The examination of dural sinus and jugular veins did not reveal any thrombosis.

Several foci of lymphocytic infiltration and focal proliferative extracapillary glomerulonephritis with fibro-epithelial crescents (Fig 3) were observed in both kidneys. There was no thrombosis of the renal veins.

**Discussion**

The clinical features and the aneurysmatic lesions of our case are similar to those reviewed by Kinjo et al and almost identical to those described in case 1 of Hughes and Stovin, in which diplopia and other symptoms of increased intracranial pressure were also the presenting complaints. These symptoms are thought to be related to thrombosis in dural sinus or jugular veins or both. \(^1\) The lysis of such thrombi during the evolution of the illness may explain the disappearance of the diplopia as well as the absence of intracranial venous thrombosis at autopsy. The pulmonary aneurysms of Hughes-Stovin syndrome have almost always been diagnosed at autopsy, except the Hughes-Stovin first case \(^1\) and Kopp and Green's case, \(^2\) where lobectomies have been done. But the radiologic suspicions were of neoplastic lesions.

The etiopathogenesis of Hughes-Stovin syndrome remains unexplained. In our case, as in the others, the blood cultures were negative, and we did not find morphologic evidence to support the advanced causative role of infected emboli. \(^3\) The lack of histologic abnormalities in the bronchial arteries also does not support the suggested interference of such vessels in the pathogenesis of the syndrome. \(^1\)

We have observed a widespread pulmonary eosinophil angiitis with very intense arterial lesions. This angiitis is completely different from the arteritis reported so far in this syndrome, \(^4,5\) and we do not know whether it is related to the aneurysmatic lesions, since no eosinophil infiltrates were detected in the segmental pulmonary arteries.

It is tempting, however, to invoke the existence of an immunologic linkage between the pulmonary angiitis and the renal lesions observed in our case.

Our case reinforces the assumption that Hughes-Stovin syndrome is not a definite pathologic entity but a complex of clinical manifestations and pathologic...
findings eventually caused by different factors; it suggests, moreover, that if one is aware of the clinical presentation and x-ray findings of such a syndrome, the diagnosis of pulmonary aneurysms can be made at the onset of hemoptysis.

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Retrograde Embolization of a Detached Polyethylene Catheter*

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The increasing use of intravenous polyethylene catheters has led to a growing incidence of accidental breakage of catheters and migration to the heart. In this communication, we describe a patient in whom the broken fragment of catheter migrated retrogradely from the subclavian vein to the inferior vena cava and left iliac vein. The possible mechanism for such retrograde migration is postulated.

Embolization of polyethylene catheters is a particularly distressing complication of therapy with intravenous fluids and central venous pressure monitoring. A polyethylene catheter which was inserted into the left subclavian vein for hyperalimentation of a 65-year-old patient suffering from an active ulcerative colitis was sheared off during insertion and migrated via the superior vena cava to the right atrium and then retrogradely to the inferior vena cava and the left iliac vein. The fragment of catheter was removed from the left iliac vein by laparotomy. The possible mechanism of this rare and unique retrograde migration, as well as the importance of the immediate preoperative chest x-ray film and fluoroscopic examination for localization of the fragment of catheter, are emphasized.

CASE REPORT

The patient, a 65-year-old retired worker, was admitted to another hospital because of numerous bloody discharges (of six days' duration), dehydration, and fatigue. History revealed that he was known to suffer from ulcerative colitis and had been under treatment and supervision by a gastroenterologic clinic for 13 years. The patient also had undergone left femoropopliteal bypass six years ago.

Because of the patient's poor nutritional status, a radiopaque polyethylene catheter was inserted through the attached needle into the left subclavian vein for hyperalimentation. While pulling the catheter backwards through the introducing needle, it was instantly noticed that the cannula was severed and sheared off by the introducing needle. X-ray films of the chest showed embolization into the superior vena cava and right atrium.

The patient was transferred to the Beilinson Medical Center, Tel Aviv, for removal of the embolized catheter, where two unsuccessful trials to remove the fragment percutaneously were made. After a subsequent chest x-ray film confirmed the presence of the catheter in the right atrium, the patient was scheduled for surgical removal of the fragment lying in the region of the left iliac vein.

Figure 1. Plain abdominal x-ray film shows polyethylene fragment lying in region of left iliac vein.

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