Hughes-Stovin syndrome is an extremely rare disease known to cause multiple pulmonary artery aneurysms (PAAs) and venous thrombosis (1, 2). The patients can develop massive hemoptysis and the disorder can result in death. Although the use of systemic corticosteroids with a combination of immunosuppressants has been the mainstay of treatment, interventional treatments are required for a life-threatening hemoptysis. However, a thrombosis in the central veins can impede catheter passage. In this report, we describe the use of percutaneous transhepatic venous embolization through the hepatic vein due to occluded common vascular pathways to the pulmonary artery.

**Index words:** Pulmonary artery
Embolization
Aneurysm
Computed tomography (CT)
Cardiovascular

Hughes-Stovin syndrome is an extremely rare disease known to cause multiple pulmonary artery aneurysms (PAAs) and venous thrombosis (1, 2). The patient developed deep vein and inferior vena cava (IVC) thrombosis, repeated internal bleeding and pulmonary artery aneurysms (PAAs). The patient presented with massive hemoptysis and with PAAs of a 2.5 cm maximum diameter. We describe the successful percutaneous transhepatic venous embolization of the PAAs due to occluded common vascular pathways to the pulmonary artery.

**Case Report**

The institutional review board approved the study reported here. A 42-year-old male patient was admitted with acute abdominal pain. The patient had a history of deep vein thrombosis on the left lower extremity nine months prior to presentation and had been taking warfarin. The patient had had a recurrent oral ulcer for 10 years, but a genital ulcer, eye lesion or skin lesion was not revealed. The blood pressure was 120/80 mmHg and the heart rate was regular at 96 beats per minute. On physical examination, the abdomen was rigid and flat and direct tenderness was felt in the right upper quadrant area. Laboratory investigations revealed a hemoglobin value of 14.3 g/dL that decreased to 9.3 g/dL the next day, and a white blood cell count of 9,800/μL. The prothrombin time was prolonged 30.8 sec (normal, 9.5-13.2 sec) due to the warfarin therapy. The amylase level was increased to 3263 IU/L (normal, 25-115 IU/L) from acute pancreatitis probably due to a mass effect of a hematoma. A pathergy test of an intradermal injection of 0.1 ml normal saline into a forearm and the develop-
ment of a skin eruption (papule, nodule, pustule) was also negative.

Computed tomography (CT) of the abdomen revealed a hematoma and active bleeding in anterior pararenal space around the duodenal second portion with a focal, localized eccentric thrombosis of the inferior vena cava (IVC) at the level of renal vein confluence. Arteriography demonstrated multiple microaneurysms at the transverse pancreatic artery and the pancreatocoduodenal artery; they were embolized with three 6-mm Tornado platinum microcoils (Cook, Bloomington, IN U.S.A.).

The patient had to stop anticoagulation therapy due to bleeding, and a Gunther Tulip IVC filter (Cook) was temporarily placed via the right internal jugular vein.

Recovery was uneventful, and the patient was discharged in good condition after two weeks with anticoagulation therapy, again with warfarin for IVC thrombosis and the IVC filter was retrieved via the right internal jugular vein. However, after another 4 weeks, the patient was readmitted to a local hospital because of massive hemoptysis. When the patient was referred to our hospital, a contrast-enhanced CT scan revealed well demarcated, two ovoid shaped vascular masses on the right lower lobe compatible with PAAs (maximum diameter 1.5 cm and 2.5 cm, respectively). The diagnosis of Hughes-Stovin syndrome was made only after finding the PAAs. The IVC was completely occluded and suprarenal placement of a Gunther Tulip IVC filter (Cook) was performed. The left internal jugular vein was used for placement of the IVC filter since the right internal jugular vein was occluded. The only medical treatment of immunosuppressive therapy was done without considering an interventional procedure since the patient had been stable when referred and had refused to undergo an intervention.

On a follow-up chest CT scan 4 months later, one of two PAAs in the superior segment of the right lower lobe had spontaneously regressed. The second aneurysm in the posterior basal segment of the right lower lobe decreased in size, and was partially thrombosed. The patient was readmitted with pain in the left inguinal and left lower quadrant 4 months later. CT revealed a hematoma of the left psoas muscle area and contrast extravasation. Arteriography revealed an aneurysm of the third left lumbar artery, and endovascular embolization with glue was performed successfully.

Three months later, the patient returned to the hospita...
Hughes and Stovin first described a syndrome consisting of multiple pulmonary aneurysms and peripheral venous thrombosis in 1959 (1). Patients can show diverse symptoms from mild symptoms such as cough, dyspnea, headache and intermittent fever to severe symptom such as papilledema or massive hemoptysis (3, 4). The etiology is still unknown. Hughes and Stovin postulated that a congenital defect of the bronchial arterial wall results in inadequate nutrition to pulmonary arteries (1, 4, 5). In the setting of pulmonary embolic disease, inflammation and vessel wall destruction occurs and aneurysms are formed. Another theory is that infected emboli with organisms of low-grade virulence will cause mycotic aneurysms (1, 4, 5).

Hughes-Stovin Syndrome can be diagnosed in the case of PAAs combined with deep vein thrombosis without evidence of Behcet’s disease.

Until now, fewer than 30 cases have been published in the English language literature.

Several cases revealed that massive hemoptysis due to rupture of a PAA was the predisposing condition followed by death (6, 7).

In Hughes-Stovin syndrome, the pulmonary artery is usually involved with hemoptysis but there has been a report of hemoptysis caused by rupture of a bronchial artery aneurysm [8]. In another case, deep vein thrombosis and combined left hepatic artery aneurysms were

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Fig. 2. A. Percutaneous transhepatic venousvenous access was attempted. An IVC filter is demonstrated on the suprarenal portion for IVC thrombosis.

B. A pulmonary artery angiogram reveals a 2.5 cm sized pulmonary artery aneurysm on the right lower lobe.

C. Angiography shows complete exclusion of the pulmonary artery aneurysm following embolization with twelve Nester coils.
This case showed pancreaticoduodenal and lumbar artery aneurysms that were ruptured as well as PAA with progressive deep vein and IVC thrombosis. The frequency of pulmonary involvement by Behcet’s disease is about 1-8% (9). When a pulmonary artery aneurysm is revealed, usually Behcet’s disease is suspected. Nevertheless, if clinical symptoms such as a skin lesion or genital ulcer are not consistent with Behcet’s disease, the next step is to consider Hughes-Stovin syndrome. The use of a pathergy test that has been accepted as one of the major criteria in Behcet’s disease shows a negative finding for Hughes-Stovin syndrome.

Treatment for the PAA is mainly the use of immunosuppressive agents such as steroids, cyclophosphamide or azathioprine. Acican et al. (10) reported complete regression of a pulmonary aneurysm when azathioprine-steroid combination therapy was used in a patient with Behcet’s disease. This case also demonstrated that medical treatment could induce regression of a PAA though the partially regressed PAA later increased in size. If rapid resolution of a pulmonary aneurysm causing clinical symptoms is required, endovascular embolization or surgery can be considered.

Lobectomy is major method for surgical treatment of a pulmonary aneurysm. However, a lobectomy has significant limitations. Pulmonary involvement in Hughes-Stovin syndrome shows a tendency of multiple pulmonary aneurysms in a few lobes, and close follow-up should be mandatory. Endovascular embolization with coils or other embolic agents is the ideal method for PAA treatment. This method can lead to rapid occlusion of an aneurysm and the procedures can be performed several times. The general condition of a patient will more quickly recover than for surgery. However, a thrombosis in central veins can impede catheter passage. To the best of our knowledge, this is the first report of the use of transhepatic venous embolization of PAA in Hughes-Stovin Syndrome.

We conclude that the percutaneous transhepatic venous approach is a good alternative access route to treat PAA in patients whose normal pathways are occluded due to a venous thrombosis.

References