รายงานผู้ป่วย : Hughes-Stovin Syndrome

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A case report : Hughes-Stovin Syndrome

Abstract

Hughes-Stovin Syndrome (HSS) is a very rare clinical disorder characterized by thrombophlebitis and multiple pulmonary and/or bronchial aneurysms. The exact etiology and pathogenesis of HSS is unknown; possible causes include infections and angiodysplasia. HSS has also been considered as a variant of Behcet's disease.

A case report of young male patient, initially presented with cerebral venous sinus thrombosis. Later, he presented with massive hemoptysis. The computed tomography of the chest demonstrated multiple pulmonary aneurysms. Pathogenesis, computed tomography, surgical pathology and treatment of this syndrome are also briefly discussed.

In young men presenting with venous thrombosis as revealed on imaging examination, with platelet count and coagulation tests within normal and hemoptysis the eventuality of HSS is to be considered.

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Keywords : Hughes-Stovin syndrome, Behcet's disease, massive hemoptysis, thrombophlebitis, multiple pulmonary and/or bronchial aneurysms, computed tomography of the chest

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บทคัดย่อ

Hughes-Stovin syndrome (HSS) เป็นภาวะที่พบได้น้อยมาก ผู้ป่วยจะมีภาวะของหลอดเลือดดำอักเสบอุดตัน, การโป่งพองของหลอดเลือดแดงที่ไปยังปอด และ/หรือหลอดลมหลายตำแหน่ง ซึ่งยังไม่ทราบสาเหตุ และการเกิดโรคที่แท้จริง เป็นไปได้ว่าอาจจะเกิดจากการติดเชื้อ และความผิดปกติของหลอดเลือด HSS ถูกพิจารณาว่าเป็นตัวแปรหนึ่งของ Behcet's disease

รายงานผู้ป่วยวัยรุ่นชายรายนี้ได้รับการวินิจฉัยเป็น HSS เนื่องจากผู้ป่วยมีประวัติของหลอดเลือดดำสมองอุดตัน ต่อมามีอาการไอเป็นเลือดปริมาณมาก แพทย์จึงส่งตรวจเอ็กซเรย์คอมพิวเตอร์บริเวณทรวงอก พบการโป่งพองของหลอดเลือดแดงที่ไปยังปอดหลายตำแหน่ง ร่วมกับผลตรวจทางพยาธิวิทยาที่เข้าได้กับภาวะนี้ ซึ่งจะขอกล่าวถึงการเกิดภาวะนี้, การตรวจเอ็กซเรย์คอมพิวเตอร์, ผลตรวจทางพยาธิวิทยา และการรักษาอย่างคล่าวๆ ต่อไป

ฉะนั้นผู้ป่วยวัยรุ่นที่มีประวัติหลอดเลือดดำอุดตัน โดยผลการตรวจจำนวนเกล็ดเลือด และค่าการแข็งตัวของเลือดอยู่ในระดับปกติ ระยะต่อมามีอาการไอเป็นเลือดจำนวนมาก ควรนึกถึง HSS ด้วย

คำรหัส : ไอเป็นเลือดปริมาณมาก, ภาวะของหลอดเลือดดำอักเสบอุดตัน, การโป่งพองของหลอดเลือดแดงที่ไปยังปอด และ/หรือหลอดลมหลายตำแหน่ง, เอ็กซเรย์คอมพิวเตอร์บริเวณทรวงอก

Case Report รายงานผู้ป่วย

Introduction

Hughes-Stovin syndrome (HSS) was named after two British physicians, Drs. John Patterson Hughes and Peter George Ingle Stovin. They first described the findings of the syndrome (deep venous thrombosis and segmental pulmonary artery aneurysms) in a total of four male patients with pulmonary artery aneurysms in 1959. Typical symptoms are recurrent fever, chills, hemoptysis and coughs and it usually affects young men. The natural course of the illness is usually fatal because of massive hemoptysis1,2,3.

About 25% of patients with HSS develop thromboembolism, arterial aneurysms and vascular occlusions. The distribution of the vascular component of the syndrome is as follows: arterial (7%), venous (25%) or both (68%). The clinical paradigm of HSS can be divided into three phases:

a. Symptoms of thrombophlebitis

b. Formation of large pulmonary and/or bronchial aneurysms

c. Aneurysmal rupture leading to massive hemoptysis and death

The aetiology of HSS is still unknown; however it is supposed to be a clinical variant manifestation of Behçet disease1,4,5,8.

Case report

A 18 years old, Thai male patient presented with massive hemoptysis. He has previous history of cerebral venous sinus thrombosis and old pulmonary tuberculosis, after treatment 1-2 years ago. His vital signs including temperature, arterial pressure and heart rate were normal. Clinical examination was negative. His platelet count and coagulation tests were within normal limits. There was no leukocytosis, and the erythrocyte sedimentation rate was normal. Skin examination was normal.

Chest postero-anterior plain film shows mass-liked lesions at left lower lung zone and reticulonodulopatchy opacity at right upper-middle lung zone. No cardiomegaly and/or pleural effusion is detected (Figure 1).

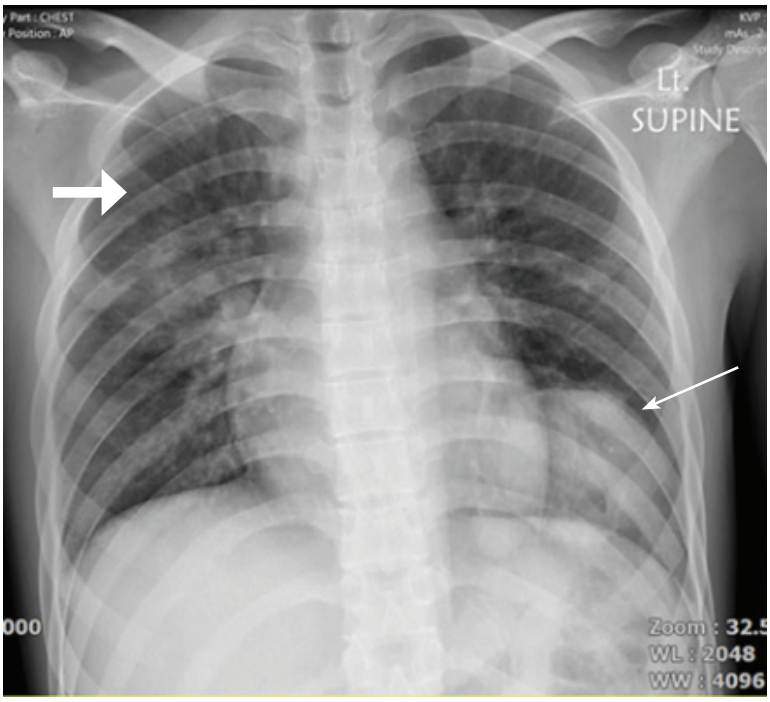

Figure 1 Chest postero-anterior plain film shows mass-liked lesions at left lower lung zone (thin white arrow) and reticulonodulopatchy opacity at right upper-middle lung zone (thick white arrow). No cardiomegaly and/or pleural effusion is detected.

The computed tomography of the chest reveals pulmonary aneurysms of right upper and lower lobe pulmonary arteries and partially thrombosed aneurysm of the left lower lobe pulmonary artery (Figure 2).

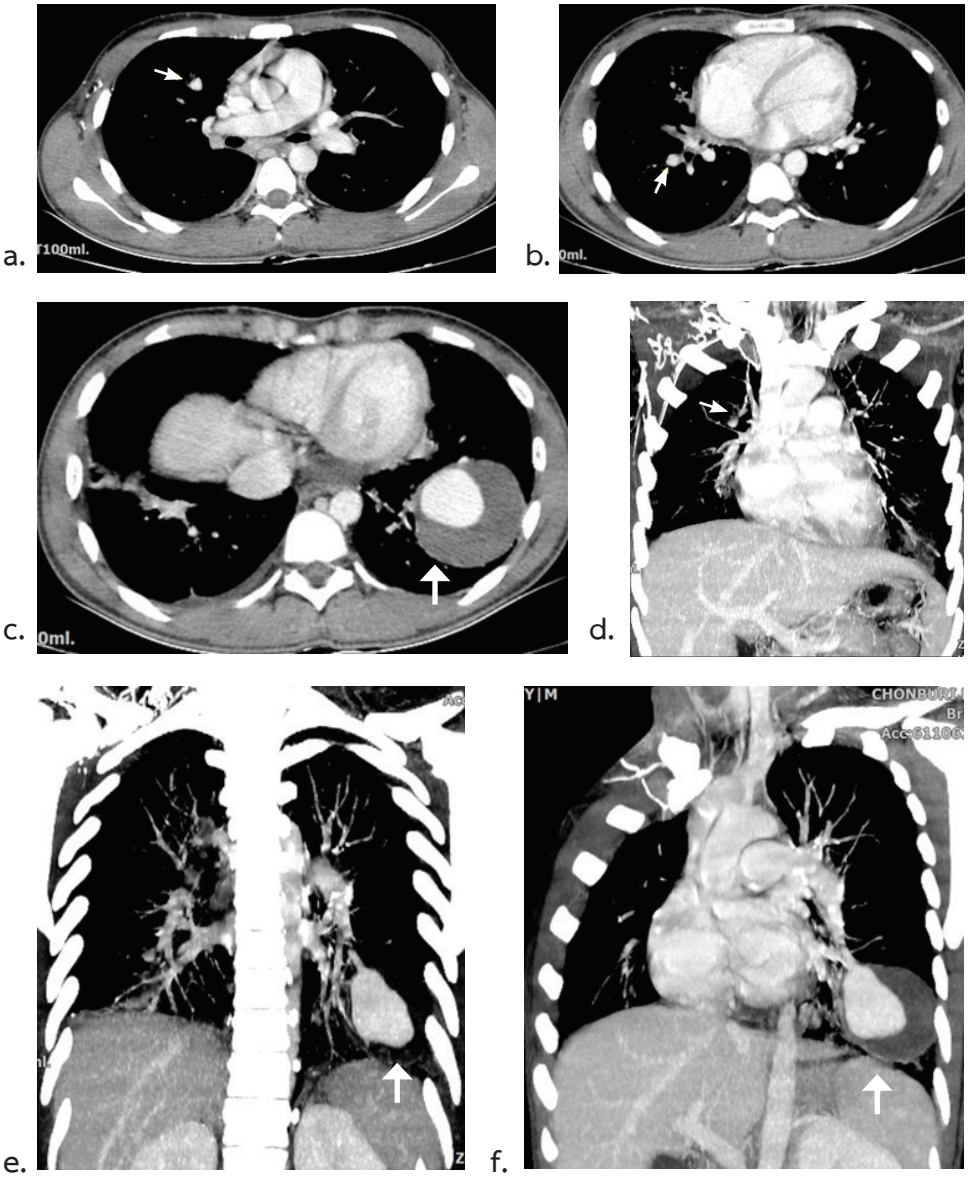

Figure 2 The computed tomography of the chest.

a. to c. shows pulmonary aneurysms of right upper and lower lobe pulmonary arteries (thin white arrows) and partially thrombosed aneurysm of the left lower lobe pulmonary artery (thick white arrow)

d. to f. maximum intensity projection (MIP) of pulmonary artery shows pulmonary aneurysms of right upper lobe pulmonary artery (thin white arrows) and partially thrombosed aneurysm of the left lower lobe pulmonary artery (thick white arrow)

The previous computed tomography of the brain shows thrombus in bilateral transverse to sigmoid sinuses (Figure 3).

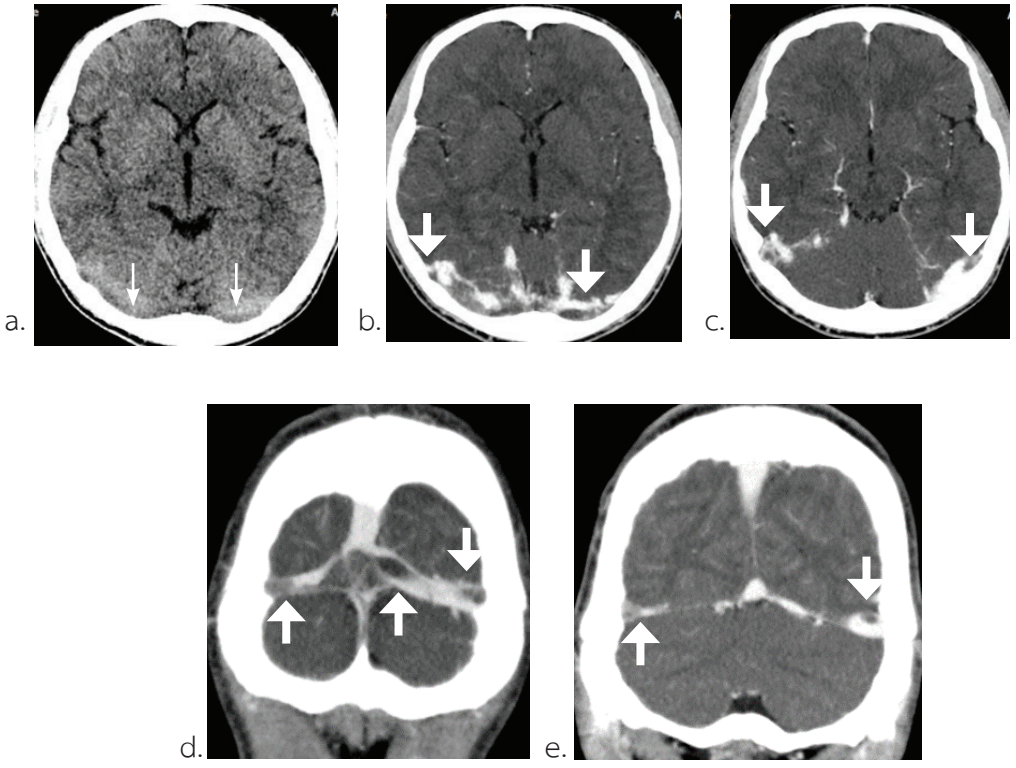

Figure 3 The computed tomography (CT) of the brain

a. pre-contrast CT of the brain shows hyperdense lesions along bilateral transverse sinuses (thin white arrows)

b. and c. post-contrast CT scan of the brain shows filling defect or thrombus in bilateral transverse to sigmoid sinuses (thick white arrows)

d. and e. coronal post-contrast CT scan of the brain shows filling defect or thrombus in bilateral transverse to sigmoid sinuses (thick white arrows)

The physician also decided to combine pulse therapy with methylprednisolone and cyclophosphamide for initial management and consulted the cardiothoracic surgery team for surgical management (left lower lobe lobectomy and/or aneurysmectomy).

The surgical pathology reveals a large pulmonary aneurysm at sections of left lower lobe of lung with containing recurrent hemorrhage and organizing thrombosis. The aneurysmal wall shows smooth muscle layer and mixed inflammatory cells infiltrate (Figure 4).

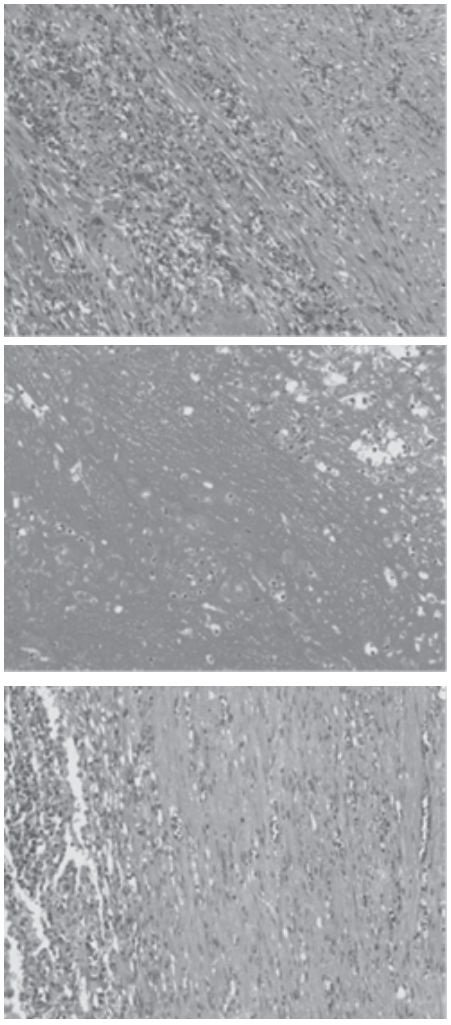

Figure 4 The surgical pathology report reveals a large pulmonary aneurysm at sections of left lower lobe of lung with pulmonary hematoma, remaining pulmonary tissue edema and congestion. The aneurysmal wall shows smooth muscle layer and mixed inflammatory cells infiltrate. The surrounding and remaining lung parenchyma are unremarkable.

Finally, a diagnosis of Hughes-Stovin syndrome, was made on the basis of pulmonary artery aneurysms and venous thromboses in such a young patient, without clinical findings consistent with Behçet disease. Even though our patient lacked a history of fever and chills, however dry cough, followed by hemoptysis due to pulmonary aneurysm.

Discussion

HSS is a rare clinical condition characterized by multiple pulmonary artery aneurysms and peripheral venous thrombosis, first described in 1959. Patients, mostly men aged 12–40 years may present with hemoptysis, cough, dyspnea, chest pain, and signs of pulmonary hypertension. Other associated features reported in HSS include fever and elevated intracranial hypertension. Since its first description, there have been only a few reports of its occurrence in the English medical literature 1,2,3,9.

Aneurysms usually involve the pulmonary arteries and the bronchial arteries resulting in subsequent hemoptysis. However they can occur anywhere in systematic circulation. Recurrent phlebitis also commonly involves the large vessels resulting in thrombus formation. In general there is a thrombus formation predisposition affecting the peripheral veins10.

Radiological diagnosis, conventional angiography (Figure 5) has been regarded as a gold standard for the diagnosis of

pulmonary artery aneurysms. It also aids in assessment of angiodysplastic bronchial arteries in HSS. The characteristic picture seen is aneurysmal formation proximal to the occluded segments while distal to the interruption, signs of hypoperfusion are observed. However, it should be noted that selective pulmonary angiography can be hazardous as it carries the risk of aneurysm rupture7.

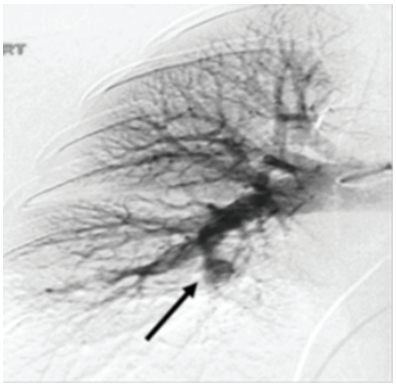

Figure 5 conventional angiography shows pulmonary aneurysm at right lower lobe pulmonary artery (back arrow)

The computed tomography of the chest or computed tomography pulmonary angiogram allows visualization of the vessel lumen, mural thrombus, vessel wall, and mediastinum (Figure 6). The volume-rendered image of our patient showed prominent and tortuous bronchial artery branches that apparently supplied a web of smaller vessels at the sites of pulmonary artery wall inflammation even before the actual aneurysm formation11.

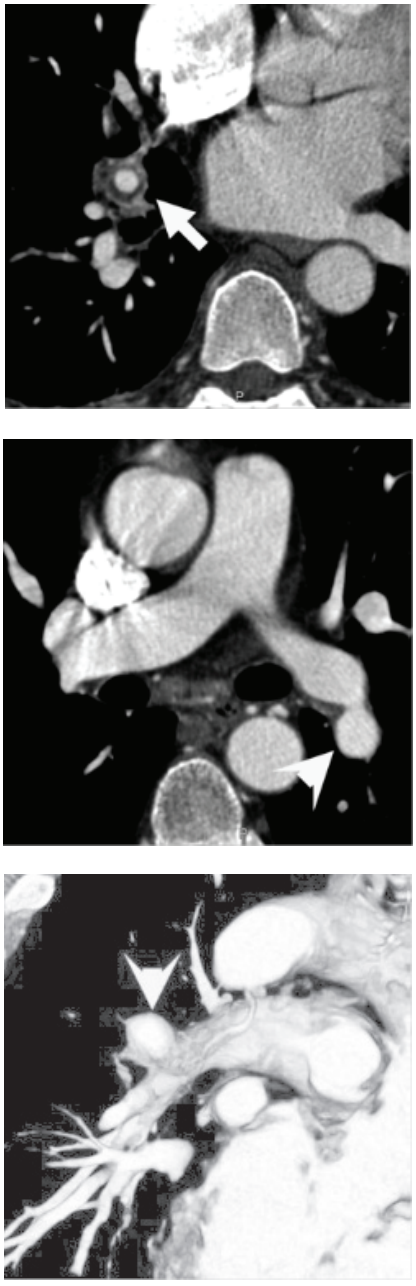

Figure 6 The computed tomography of the chest or computed tomography pulmonary angiogram and volume-rendered image reveal aneurysm of bilateral pulmonary arteries (white arrow and arrow head)

Histologic studies show destruction of the arterial wall and perivascular lymphomonocytic infiltration of capillaries and venules.

The pathogenesis of HSS is not known. The current consensus opinion is that HSS results from a vasculitis similar to that implicated in Behçet disease. Vasculitis in Behçet disease results in arterial occlusion, arterial aneurysm, venous occlusion, and varices, indicating systemic vessel involvement. Furthermore, it is very likely that HSS represents a cardiovascular manifestation of Behçet disease, since in both clinical entities pulmonary aneurysms are characteristic clinical features. However other clinical presentations can overlap significantly; both are more common in young men, the pulmonary manifestations of both can be identical, and the histology of the aneurysms in both entities can be similar1,6.

Hughes and Stovin's original theory suggested that degenerative bronchial arteries lead to changes in the vasa vasorum of the pulmonary arteries and the development of aneurysms. HSS is often fatal as a result of massive hemoptysis due to pulmonary/bronchial arterial aneurysm rupture6.

The syndrome is most often treated by immunosuppressant, either systemic corticosteroids or cytotoxic agents (a combination of cyclophosphamide and glucocorticoids), in patients without or with small amount of hemoptysis in order to stabilize the pulmonary artery aneurysms or even make them disappear. Anticoagulation may prevent the progression of pulmonary embolism and resolve vein thrombi, but since it increases the risk and severity of hemoptysis, it is contraindicated. Surgical resection of the affected segments of the lung is to be considered in cases of high risk rupture aneurysms limited to one segment or one lung. However, the high morbidity and mortality associated with surgery, and the frequent bilaterality and multifocality of the pulmonary artery aneurysms at the time of diagnosis, makes transcatheter embolization an alternative to surgery in most cases1,7,8.

Early diagnosis and timely intervention is, therefore, crucial in improving the prognosis of patients with HSS. Appropriate treatment, if instituted promptly and early in the course of the disease, has the potential to induce remission.

Conclusion

The types of vasculitis involved in HSS and in Behçet disease seems to be very similar. Though most patients with HSS will present with hemoptysis and advanced disease, dysplastic bronchial arteries and

pathologic enhancement of pulmonary thromboemboli seen on computed tomography pulmonary

angiogram should alert the radiologist and raise suspicion of pulmonary vasculitis. Earlier diagnosis could be in some cases crucial, allowing the prevention of the development of potentially life-threatening pulmonary artery aneurysms.

Reference

1. Umair Khalid, Taimur Saleem. Hughes -Stovin Syndrome. Orphanet Journal of Rare Diseases 2011; 15 : 1-11.
2. Hughes JP, Stovin PG: Segmental pul monary artery aneurysms with periph eral venous thrombosis. Br J Dis Chest 1959 ; 53 : 19-27.
3. Kopp WL, Green RA: Pulmonary artery aneurysms with recurrent thrombo phlebitis: the “Hughes-Stovin syn drome”. Ann Intern Med 1962; 56 : 105-14.
4. Reimold WV, Emmrich J, Harmjanz D, Kochsiek K: Multiple aneurysms of the pulmonary artery following recur rent septic pulmonary embolism (Hughes-Stovin syndrome): report of 1 case. Arch Klin Med 1968; 215 : 1-18.
5. Khalil A, Parrot A, Fartoukh M, Marsault C, Carette MF: Images in cardiovas cular medicine. Large pulmonary artery aneurysm rupture in Hughes-Sto vin syndrome: multidetector computed tomography pattern and endovascular treatment. Circulation 2006; 114 : e380-1.
6. Al-Jahdali H: Massive hemoptysis and deep venous thrombosis presenting in a woman with Hughes-Stovin syn drome: a case report. J Med Case Reports 2010; 4 : 109.
7. Mahlo HR, Elsner K, Rieber A, Brambs HJ: New approach in the diagnosis of and therapy for Hughes-Stovin syn drome. AJR Am J Roentgenol 1996 ; 167 : 817-8.
8. Athanasios N Chalazonitis, Stefanos B Lachanis, Panagiotis Mitseas, Pa nagiotis Argyriou, Joannie Tzovara, Petros Porfyrides, Evangelia So tiropoulou, Nikolaos Ptohis. Hughes -Stovin Syndrome: a case report and review of the literature. Cases Journal 2009; 2 : 98.
9. Weintraub JL, DeMayo R, Haskal ZJ, Susman J. SCVIR Annual Meeting Film Panel Session: Diagnosis and Dis cussion of Case 1. J Vasc Interv Radiol 2001; 12 : 531-534.
10. Durieux P, Bletry O, Huchon G, et al. Multiple pulmonary arterial aneurysms in Behcet’s disease and Hughes-Sto vin syndrome. Am J Med 1981; 71 : 736-41.
11. Eric S. Ketchum, Roham T. Zama nian, Dominik Fleischmann. CT Angiography of Pulmonary Artery An eurysms in Hughes-Stovin Syndrome. AJR 2005 ; 185 : 330–332.