Hughes-Stovin Syndrome : A Case Report and Review of the Literature

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Abstract

A young man presented with prolonged pyrexia, recurrent optic neuritis, thrombophlebitis and bilateral pulmonary artery aneurysms with thrombus formation. The life-threatening hemoptysis necessitated mechanical ventilatory support and emergency left lower lobectomy. Systemic corticosteroids conferred clinical improvement and reduction of the remaining right pulmonary artery aneurysm. The patient eventually succumbed to sudden massive hemoptysis. This report underscores the unpredictable nature of this syndrome and emphasises the need for aggressive surgical intervention of pulmonary artery aneurysms in Hughes-Stovin syndrome.

Key word : Hughes-Stovin Syndrome, Pulmonary Artery Aneurysm, Optic Neuritis

Hughes-Stovin syndrome, originally described in 1959, is characterised by multiple, segmental pulmonary artery aneurysms and venous thrombosis[1]. Fever and signs of intracranial hypertension are recognised associated features [2,3]. There have been 15 reported cases in the English literature as of 1993. Most patients (14 of 15) were young males. The disorder carries a grave prognosis with death from recurrent, massive hemoptysis almost as a rule.

Described herein is the first report in Thailand of Hughes-Stovin syndrome presenting with bilateral pulmonary artery aneurysms, preceded by recurrent optic neuritis, thrombophlebitis and prolonged fever. Although the eventual outcome was fatal, this case adds further evidence of tangible benefits of systemic corticosteroids in this condition.

CASE REPORT

A 19-yr-old student was referred to the Central Chest Hospital in November 1995 with a 5-month history of unremitting fever and chills. Hemoptysis commenced in early August and gra-

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dually progressed over the ensuing 4 months. The patient also described stabbing left sided chest pain, and increasing dyspnea spanning 2 months.

Prior to this referral, he had required three admissions from July to November at a local hospital for the above symptoms. He developed recurrent lower limb thrombophlebitis during the second admission. He also experienced blurred vision, for which he received 40 mg/day of prednisolone with some relief. It transpired that a similar episode of visual disturbance occurred in January, for which he was diagnosed as having optic neuritis. Systemic enquiry disclosed no oral or genital ulcers. Family history was unremarkable. There was no history of illicit drug usage. Various investigations and different courses of antimicrobials were given to no avail. The symptoms continued unabated, necessitating this referral.

On examination, he was febrile (temperature 38.9°C) and tachypnic (respiration 26/min). Fundoscopy showed a pale right optic disc but no retinal hemorrhages or exudates. He had no cervical lymphadenopathy or oral ulcers. Bronchial breathing was audible at the left posterobasal region with no pleural rubs. Apart from tachycardia (heart rate 112/min), cardiovascular system was unremarkable. There was no digital clubbing, rash or edema. He had no calf swelling or tenderness but there were signs of previous superficial thrombophlebitis of both legs.

Chest radiograph on arrival displayed well-defined, homogeneous bilateral juxtahilar densities (Fig. 1). Computed tomography of the chest (Fig. 2) and spiral CT angiogram with 3D-reconstruction of the pulmonary artery (Fig. 3) showed bilateral pulmonary artery aneurysms with thrombus formation, the diameters being 10 cm on the left and 3 cm on the right. His poor clinical condition rendered a pulmonary angiography too risky.

Hemograms showed Hb 11.2 g/dl, WBC 18,400 (N 93, L 5, M 2%), platelets 280,000/mm^3. ESR was 126 mm/1st hour. Prothrombin and partial thromboplastin times were normal. Renal indices, liver function tests, and electrolytes were normal. Urinalysis showed no cells, no glycosuria or proteinuria. Blood anti HIV was negative. Blood cultures grew no organisms on 2 sets, 3 specimens each. Mycoplasma, Melioid, Widal, Leptospira, VDRL, Weil-Felix titres were all unyielding. RF, ANA, anti DNA, anticardiolipin antibody, cANCA were all non-contributory. Echocardiogram showed normal cardiac chambers and valvular structures, and particularly no vegetations.

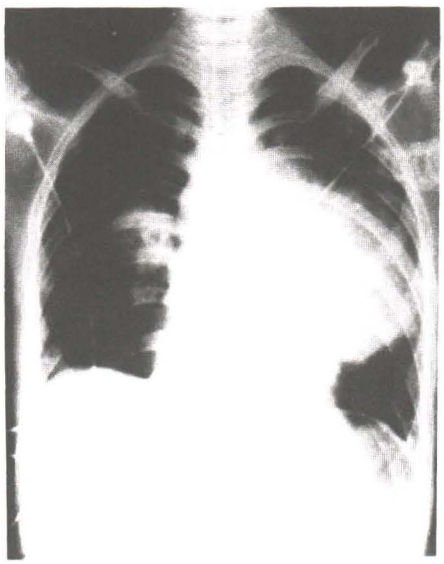

Fig. 1. Chest radiograph on arrival showing bilateral juxtahilar densities.

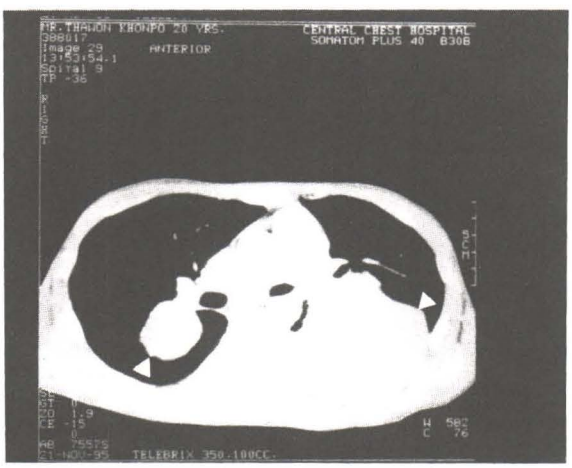

Fig. 2. CT scan of the chest showing bilateral pulmonary artery aneurysms with thrombus formation (arrowheads).

The preceding recurrent optic neuritis, prolonged pyrexia, recurrent thrombophlebitis and bilateral pulmonary artery aneurysms constitute the Hughes-Stovin syndrome. Shortly after referral, the

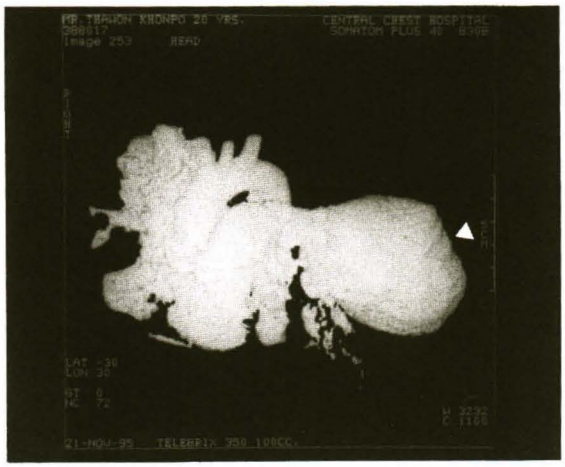

Fig. 3. Spiral CT angiogram with 3D-reconstruction showing the left pulmonary artery aneurysm (arrowhead).

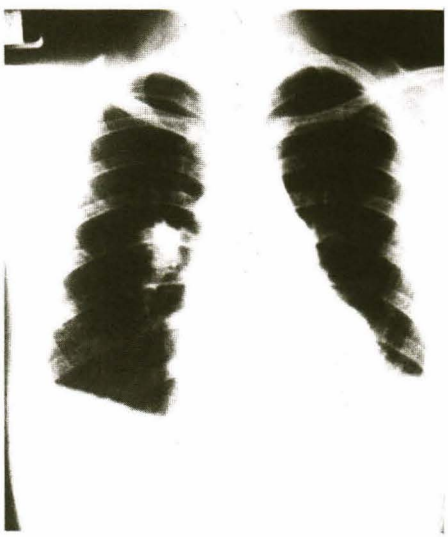

Fig. 4. Chest radiograph (3 months later) showing a reduction of the right pulmonary artery aneurysm.

patient deteriorated from life-threatening hemoptysis requiring mechanical ventilatory support. Fibreoptic bronchoscopy showed markedly compressed superior segmental bronchus of the left lower lobe with swollen, hemorrhagic mucosa. Milder compression was also noted at the orifice of the superior segmental bronchus of the right lower lobe. He underwent succesful emergency left lower lobectomy. It proved impossible to isolate the left pulmonary artery aneurysm in view of severe adhesions and inflammatory process of the left lower lobe. Histopathological examination showed atherosclerotic and aneurysmal changes with relatively little inflammatory cell infiltrations of the resected pulmonary arteries. Severe hemorrhagic infarction of the lung parenchyma was observed. Staining and cultures of the aneurysmal tissues revealed no bacteria, spirochetes or fungi.

The postoperative period was largely uneventful. Methylprednisolone, one gram daily for three days, followed by daily maintenance dose of prednisolone (10 mg) was instituted. Defevescence was evident in 5 days and hemoptysis ceased.

Elective right pulmonary aneurysmectomy was strongly advised. Despite full explanation of the necessity, this was declined by the patient. He remained well throughout the following three months. Last follow-up radiograph (Fig. 4) showed a significant reduction of the right pulmonary artery aneurysm. Most unfortunately, the patient succumbed to a brisk episode of massive hemoptysis in March 1996.

DISCUSSIONS

This patient's massive hemoptysis resulted from bilateral, pulmonary artery aneurysms with thrombus formation. Recognised causes of a pulmonary artery aneurysm encompass congenital, mycotic, vasculitic and traumatic disorders[4]. Extensive investigations in this case did not reveal an underlying infection. Neither was his presentation compatible with disseminated giant cell arteritis. In view of the lack of oro-genital ulcerations, he did not fullfil criteria for the diagnosis of Behcet's disease[5].

The etiology of Hughes-Stovin syndrome remains obscure. The negative microbiological and autoimmune profiles in our case did not suggest an infection or autoimmunity. Furthermore, the time span of almost one year from the initial optic neuritis to the development of pulmonary artery aneurysms makes underlying infectious process unlikely.

Behcet's disease could produce identical pulmonary artery aneurysms[6]. It also shares other clinical manifestations with Hughes-Stovin syndrome, particularly prolonged fever and venous

thrombosis. The two conditions may be clinically indistinguishable, except for the lack of oro-genital ulcerations in the latter. It has thus been considered that Hughes-Stovin syndrome may be an incomplete (variant) form of Behcet's disease[7].

Vasculitic process appears to play an important role in the pathogenesis of Hughes-Stovin syndrome. However, the histopathological findings have been inconsistent. In contrast to eosinophilic infiltration of the pulmonary arteries as reported by Meireles et al[3], the inflammatory cells infiltration in the present case was scant and nonspecific.

Corticosteroids and cyclophosphamide have been tried with variable benefits[2]. Colchicine has purportedly been of some value[7]. The prompt clinical remission and reduction in the size of the remaining pulmonary artery aneurysm in the present case adds further evidence of a tangible benefit of corticosteroids in Hughes-Stovin syndrome.

This case report underlies the unpredictable nature of this syndrome and emphasises the necessity for aggressive surgical resection of the pulmonary artery aneurysms associated with Hughes-Stovin syndrome.

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กลุ่มอาการฮิวส์–สโตวิน : รายงานผู้ป่วยหนึ่งรายและทบทวนวารสาร

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รายงานผู้ป่วยชาย อายุ 19 ปีมารับการรักษาด้วยอาการต่าง ๆ คือ มีไข้เรื้อรัง, เส้นประสาทตาอักเสบเป็น ๆ หาย ๆ, เส้นเลือดดำอักเสบ, เส้นเลือดแดงที่ปอดทั้ง 2 ข้าง โป่งโตและมีก้อนเลือดดำอยู่ภายใน. ผู้ป่วยมีอาการไอเป็นเลือดมากขึ้น ๆ, และมีภาวะการหายใจวายรุนแรง จนต้องได้รับการช่วยหายใจด้วยเครื่องช่วยหายใจ, และต้องได้รับการผ่าตัดปอดด้านซ้ายกลีบล่างออกโดยด่วน. คอร์ติโคสเตียรอยด์ทำให้อาการต่าง ๆ ดีขึ้นมากและเส้นเลือดแดงที่ปอดขวามีขนาดเล็กลง. อย่างไรก็ตาม, ผู้ป่วยเสียชีวิต 3 เดือนต่อมาด้วยอาการไอเลือดออกมากเฉียบพลัน.

คำสำคัญ : กลุ่มอาการฮิวส์–สโตวิน, เส้นเลือดแดงที่ปอดโป่งพอง, เส้นประสาทตาอักเสบ

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