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A Rare but Fatal Behçet Variant: The Hughes–Stovin Syndrome—Successful Case Report and New Evidence from Literature Review

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Aorta (Stamford)

Abstract

Hughes–Stovin syndrome (HSS) is a rare potentially fatal vasculitis supposedly belonging to the spectrum of Behçet disease without ocular involvement. HSS tends to play by a temporal pattern, starting with thrombosis and followed by formation of pulmonary aneurysms. Since its mortality can reach 25% of cases, early recognition and appropriate therapy represent the major clinical challenges. We describe a rare case of HSS successfully treated via multidisciplinary management by an endovascular approach and immunosuppressive therapy.

Keywords

- Behçet disease
- Hughes–Stovin syndrome
- pulmonary artery aneurysm
- endovascular treatment
- genetic syndrome
- vena cava filter
- deep venous thrombosis
- CT scan

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Introduction

Hughes–Stovin syndrome (HSS) is a rare disorder characterized by multiple pulmonary aneurysms and thrombophlebitis: it is a life-threatening disease believed to be a cardiovascular clinical variant of Behçet disease (BD). It prefers the population between the second and fifth decades and the male sex. The evolution of the clinical course is characterized by the appearance of symptoms related to thrombophlebitis, the formation of aneurysms in pulmonary or bronchial arteries, and eventual hemoptysis, often fatal. In our case, the treatment of thrombophlebitis revealed the presence of hemoptysis. Endovascular therapy and successful pulmonary aneurysm exclusion was combined with immunosuppressive therapy.

Case Presentation

A 33-year-old male patient presented to our emergency room after an episode of hemoptysis the same morning, associated with shortness of breath and 5 days of pain in the left lower limb. Three days previously, he went to his general physician, who made diagnosis of deep venous thrombosis (DVT) and prescribed low-molecular-weight heparin (LMWH) 10,000 IU per day, which the patient took for 3 days until the hospitalization. In the emergency room, the left lower limb was edematous, and dorsalis pedis and popliteal arteries pulsation on the left side, although present, were thready. Homan's sign, dorsiflexion of the foot, and Bauer's sign, palpation of the calf following compression of the sural muscles against the interosseous membrane, were negative.

Arterial blood gases showed pH 7.45 (normal range [nr]: 7.35–7.45), pO_2 66 mm Hg (nr: 80–100 mm Hg), pCO_2 38 mm Hg (nr: 35–45 mm Hg), sO_2 94.7% (normal value [nv]: > 97%), lactate 0.9 mmol/L (nr: 0.5–1.6 mmol/l), partial pressure of oxygen in arterial blood (PaO_2)/inspired fraction of oxygen (P/F) ratio 314 mm Hg (> 350), FiO_2 21%.

He also had an history of recurrent oral ulcers, present at evaluation, and a lesion suggestive for a genital ulcer in the left scrotal region.

Initial laboratory investigation showed hemoglobin 9.5 g/dL (nv: 13–18 g/dL), white blood cell count 11.3 million/µL (nr: 4.300–10.000 million/µL), lactate dehydrogenase 507 U/L (nv: 80–300 U/L), C-reactive protein 29.9 mg/dL (nr: 0–5 mg/dL), D-dimer 2.740 ng/mL (nv: < 500 ng/mL).

An urgent chest X-ray detected three oval radiopacities with irregular margins, one in middle left length (40 × 25 mm), one in the inferior left lung (27 × 27 mm), and the largest one at the base of the right lung (82 × 81 mm). A computer tomography of the chest (►**Fig. 1A**) revealed multiple, irregular-shaped arterial aneurysms, pulmonary embolism, and pulmonary consolidation at both lung bases, as well as, ground-glass opacities in the middle and lower lobes of the right lung. The computed tomography (CT) also showed partial inferior vena cava thrombosis and left external iliac and femoral vein thrombosis. Doppler study of the lower limbs vein confirmed DVT, demonstrating filling defects in the left common femoral vein and in the left popliteal vein. CT angiography revealed an ecstatic venous vessel with drainage to the right transverse sinus, and Fluorescein angiography detected a delayed filling of the inferior nasal vascular arch and dilation of the venous capillary plexuses in the left optical disc.

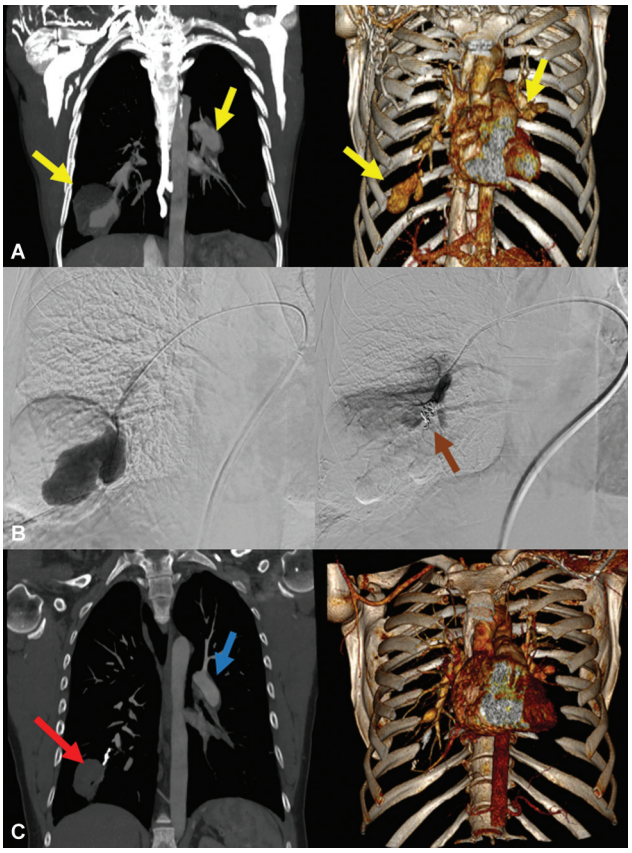

Fig. 1 (**A**) Preoperative image computer tomography (CT) maximum intensity projection (MIP) and shaded surface display (SSD) reconstruction showing pulmonary aneurysms: patent right great aneurysm and little left aneurysm (*yellow arrows*). (**B**) Intraoperative selective angiography confirming the nonruptured aneurysm and successful coil embolization (*brown arrow*) with aneurysm exclusion. (**C**) Postoperative image CT MIP and SSD reconstruction showing complete right aneurysm exclusion (*red arrow*) and unmodified left aneurysm (*blue arrows*).

During hospitalization, the patient underwent urgent embolization of the largest pulmonary arterial aneurysm (►**Fig. 1B**) and implantation of a vena cava filter.

After the aneurysmal embolization and a careful risk–benefit analysis, the patient was initiated on anticoagulation therapy with LMWH in a preventive dose, 2,000 IU 1 vial subcutaneously (fl sc) in the morning and 4,000 IU 1 fl sc in the evening.

Further laboratories exams showed a heterozygous mutation in the methylenetetrahydrofolate reductase (MTHFR) C677T gene increased Factors IX (144%) and VIII (176%) and a Factor VII less than 50% (nv: 50–200%). In addition to this, protein electrophoresis showed mildly increased beta1 and beta2 fractions complement component 3: 185 mg/dL (nr: 80–185 mg/dL) and 4: 36.8 mg/dL (15–53 mg/dL), homocysteine: 15.91 µmol/L (nv: < 15 µmol/L), anti-nuclear antibodies, anti-neutrophil cytoplasmic antibodies, anti-cardiolipin antibodies, and anti β2-glycoprotein antibodies were all normal. Moreover, genetic evaluation revealed HLA-B51:01:01 and HLA-B18:01:01 haplotype, which are

associated with BD in 50 to 70% of cases, mainly the B51:01:01 subtype.

After rheumatological consultation, the patient was treated with initial regimen consisted of prednisone (1 mg/kg/d) in combination with oral cyclophosphamide (2 mg/kg/d–100 mg/d). The patient remained in a stable condition with tapering doses of prednisone in a month and daily oral cyclophosphamide. At the follow-up (1 and 3 months later), no genital and oral ulcers nor vascular thrombosis occurred, and acute phase reactants were in normal range

Follow-up

The patient remains in good general condition at 30-month follow-up, with the vascular filter in place. Follow-up CT scan (►**Fig. 1C**) shows persistent exclusion of right pulmonary artery aneurysm and stable diameter of the contralateral pulmonary artery aneurysm.

Discussion

BD is a relapsing multisystemic inflammatory disease characterized by oral and genital aphthae, uveitis, thrombophlebitis, and which frequently involves the joints, skin, central nervous system, cardiovascular system gastrointestinal tract. Its etiology is unknown, although some scientific studies have hypothesized a genetic association with the HLA-B51 antigen.

HSS can be evoked before any deep vein thrombosis arising in a young man with BD and a fortiori when repeated hemoptysis occurs. In these conditions, chest CT scans must be performed systematically looking for aneurysmal imaginings.

Behçet Disease and Genetic Pathogenesis

The epidemiology of BD is uniquely distributed along the ancient Silk Road from Mediterranean countries to Middle Eastern and East Asian countries. BD is rarely seen in Northern Europe (0.64 cases per 100,000 population), North America (0.12–0.33 cases per 100,000 population), Australia, and Africa.

The pathogenic mechanism of BD is not completely clear. It is known that genetic susceptibility together with trigger factors and immunological abnormalities are the influence manifestation of the disease.

An important role is played by the presence of the HLA-B_51 allele of the major histocompatibility complex, through the combination of different HLA class I-associated functions and/or structural characteristics of the HLA-B_51 heavy chain. These data have been confirmed by several independent studies on 4,800 BD patients and 16,289 controls from around the world. A carrier of HLA-B5/B_51 alleles from Italy, Germany, Middle Eastern, and Far Eastern countries will have an odds ratio of 5.78 for developing the disease. Nevertheless, HLA-B_51 allele accounts for less than 20% of the genetic risk. HLA-B52 is also associated with the disease in Israel (21 vs. 9%) and HLA-B57 in the United Kingdom. HLA-B5101 and, to a lesser extent, HLA-5108 alleles have been most closely linked to patients along the Silk Road.

Moreover, a genetic contribution to disease severity has also been reported, always involving the HLA-B51 allele, which is associated with a worse clinical phenotype. There are several potential mechanisms explaining the association with HLA-B51, among which are an alteration in the B pocket of the antigen-binding groove by HLA-B51, cross-reactivity between HLA-B51, and organ-specific antigens and linkage disequilibrium with other disease associated genes. A familial pattern of occurrence has been described, indicating an increase of disease risk among first-degree individuals. Sporadic cases have been reported, even though HLA-B51 presence is higher in familial cases.

Also, common variants in interleukin 10 (IL-10) and at the IL-23R–IL-12RB2 locus might predispose individuals to BD. These variants cause a reduced expression of this anti-inflammatory cytokine, which may lead to a susceptible inflammatory state and thus to an increased susceptibility to BD.

Genome-wide studies have also evidenced non-HLA regions involved in the disease as well polymorphisms in some genes, such as the intercellular adhesion molecule-1, endothelial nitric oxide synthase, tumor necrosis factor genes, vascular endothelial growth factor gene, Manganese superoxide dismutase gene, cytochrome P450 gene, endoplasmic reticulum aminopeptidase 1, the IL-10, and IL-23 receptor gene.

In our case the observed mutation of C677T gene may be correlated with appearance of DVT. Some authors suggested the involvement of this mutation in BD patient's venous diseases. Canataroglu et al[1] suggested that elevated plasma homocysteine level and mutation of C677T may play a role in the pathogenesis of venous thrombosis in BD. On the contrary, Messedi et al[2] did not observe any difference when comparing BD patients versus healthy subjects regarding the MTHFR polymorphisms, observing only an influence on homocysteinemia values

The environmental trigger hypothesis has also been proposed in BD patients with genetic susceptibility. Trigger factors such as bacteria or viruses may have a high affinity for HLA-B51 molecules.

Geographic and Genetic Perspective of Hughes–Stovin Syndrome

HSS was first described in 1959 by John Patterson Hughes and Peter George Ingle Stovin.[3] Case reports and case series have been described in 42 papers published in the medical literature. We conducted an analysis concerning the geographic distribution of PubMed published papers. The number of cases and the publication location of the cases are demonstrated in ►**Fig 2**. The analysis documented a prevalent diffusion in the Mediterranean and Middle Eastern regions. This evidence could be correlated with genetic analyzes that hypothesize the presence of the alteration of the HLA-B51 antigen in BD. In fact, familiarity and the contemporary geographical area of belonging could correspond to a single belonging phenotype. Although there is no consensus on the etiology, HSS should be included among the pathologies in which genetics play a potential role and may provide opportunities for prevention. In our analysis, despite the rarity of occurrence, the geographic distribution

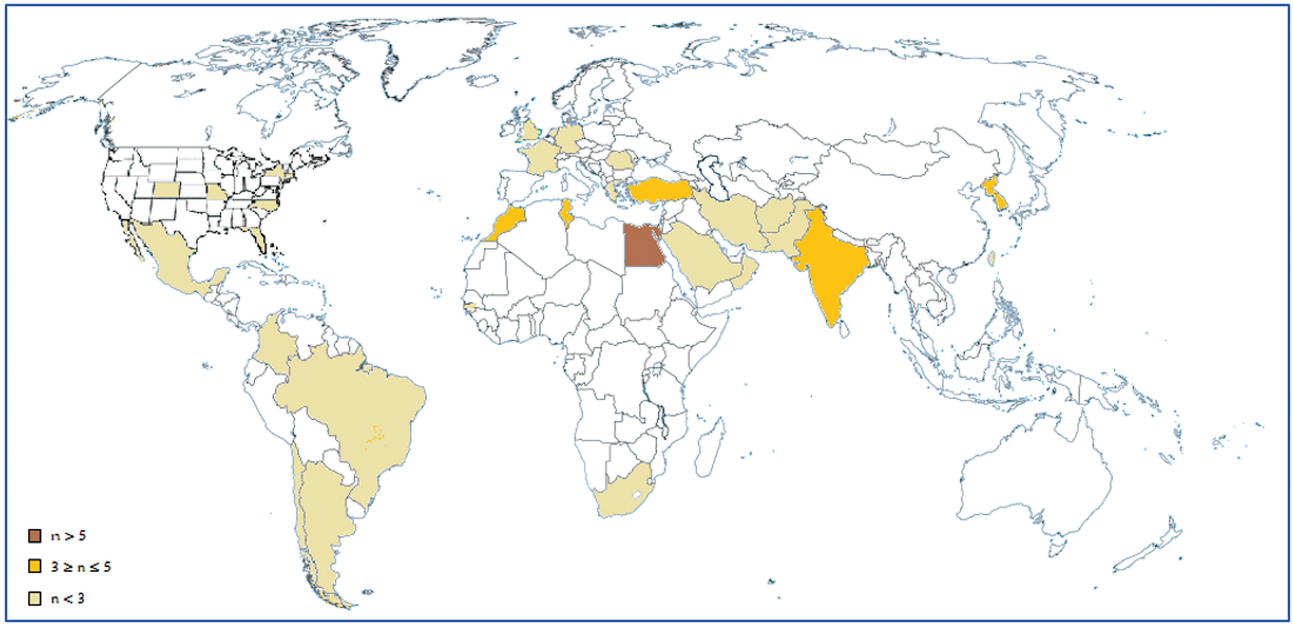

Fig. 2 Number of confirmed cases of Hughes–Stovin syndrome reported by geographic area.

of the disease suggests the need for genetic evaluations for better definition and prevention, to develop future genetic treatment models. However, it is important to identify the disease early and allow prompt treatment of any complications with accurate follow-up.

New Clinical Findings

There is no unanimous consensus on the therapy of patients diagnosed with HSS. The review of therapies and the duration of follow-up did not show a superiority of medical therapy compared with medical and surgical therapy. Our data were collected from literature review between 1911 (this year is report in the first publication in 1959, from Hughes and Stovin) and 2020, combined and 57 papers since 1959 were selected with one or more cases of HSS. Two cases were added from the Hughes–Stovin study published in 1959, which cited the earlier cases of Beattie and Pirani.[3] Seven papers were excluded because no data are available on follow-up. Repeated papers are excluded. A total of 40 patients were analyzed.[3–59] The comparison of surgical/medical or medical alone therapy on length of follow-up is reported in ►**Fig 3**. No differences in survival rate were observed between the two groups.

Therapeutically Options

The advent of endovascular procedures has expanded the possibilities for treatment. At the beginning, therapeutically options

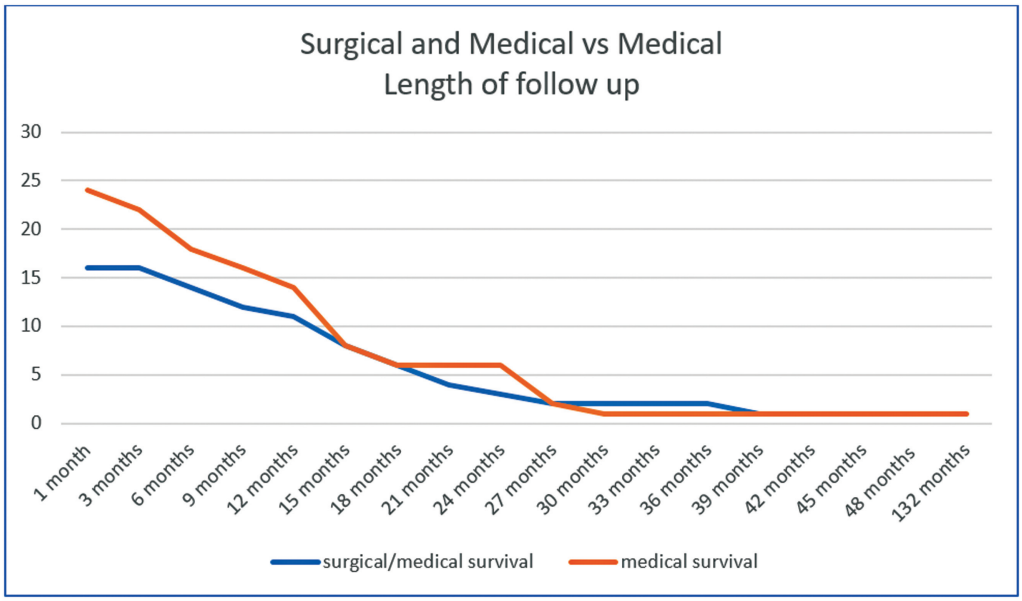

Fig. 3 Literature review of follow-up length of patients after Hughes–Stovin syndrome diagnosis: surgical versus medical/surgical therapy.

were represented by pneumectomy,[16] lobectomy,[3–5,9,25,37,50,52] and segmentectomy.[40] Aneurysm resection with saphenous vein replacement[9] or lung transplantation[10] were also proposed. More recently, embolization, even with repeated procedures, have been performed, with good immediate and mid-term results (See ►**Supplementary Materials 1–3).**[13–15,22,28,33,37,42]

The simultaneous presence of phlebitis and aneurysms makes the subsequent therapeutic path difficult because of the hemorrhagic risk associated with oral anticoagulation. In our case, the association between deep vein thrombosis and the simultaneous presence of a contralateral pulmonary aneurysm was addressed using a cava filter, which thus reduces the risk of DVT embolism and at the same time protects the patient from complications of anticoagulant therapy and aneurysm bleeding.

HSS syndrome is rare, but its consequences can be fatal. Since its mortality can reach 25% of cases, the early recognition with an appropriate therapy represents the major challenges. Verifying a genetic correlation, also linked to BD, may be represent the challenge of the future. New therapeutic options make treatment less invasive. The association with immunosuppressant has immediate and mid-term good results.

Note
This study was presented at the 70th International Congress of the European Society for Cardiovascular and Endovascular Surgery and 7th International Meeting on Aortic Diseases, Liege, Belgium, between June 20–23, 2022.

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None.

Conflict of Interest
The authors declare no conflict of interest related to this article.

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