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Hughes-Stovin Syndrome: A Rare Cause of Concurrent Pulmonary Artery Aneurysm and Deep Vein Thrombosis

Syndrome de Hughes-Stovin : Une Cause Rare d'Anévrisme Simultané de l'Artère Pulmonaire et de Thrombose Veineuse Profonde

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Résumé

Le Syndrome de Hughes-Stovin (SHS) est un trouble vasculitique rare et potentiellement fatal, caractérisé par des anévrismes de l'artère pulmonaire (AAP) et une thromboembolie veineuse. Il partage des caractéristiques communes avec la maladie de Behçet (MB), conduisant souvent à des difficultés diagnostiques. Nous présentons un patient mâle de 23 ans atteint de la maladie de Behçet qui a développé une hémoptysie, une dyspnée et des anévrismes de l'artère pulmonaire avec des thrombus intracardiaques, suggérant le SHS comme une complication vasculaire de la MB. Un traitement immunosuppresseur précoce par glucocorticoïdes et cyclophosphamide a été initié, stabilisant l'état du patient. Ce cas souligne l'importance d'une reconnaissance rapide et d'une immunosuppression agressive dans la prise en charge du SHS pour prévenir des complications fatales telles que l'hémoptysie massive ou la thromboembolie.

Mots-clés : Maladie de Behçet's, Syndrome de Hughes-stovin, Vasculite artérielle large, Anévrysme artériel pulmonaire périphérique, thrombose
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Summary

Hughes-Stovin Syndrome (HSS) is a rare and life-threatening vasculitic disorder characterized by pulmonary artery aneurysms (PAAs) and venous thromboembolism. It shares overlapping features with Behçet's disease (BD), often leading to diagnostic challenges. We present a 23-year-old male with Behçet's disease who developed hemoptysis, dyspnea, and pulmonary artery aneurysms with intracardiac thrombi, suggesting HSS as a vascular complication of BD. Early immunosuppressive therapy with glucocorticoids and cyclophosphamide was initiated, stabilizing the condition. This case highlights the importance of prompt recognition and aggressive immunosuppression in managing HSS to prevent fatal complications such as massive hemoptysis or thromboembolism.

Keywords: Behçet's disease, Hughes-stovin syndrome, Large-artery Vasculitis, Peripheral Pulmonary Artery Aneurysm, Thrombosis
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Introduction

Hughes-Stovin Syndrome (HSS) is an exceptionally rare and fulminant systemic vasculitis, first delineated in 1959. It is defined by the pathognomonic constellation of multiple pulmonary artery aneurysms (PAAs) and recurrent deep venous thromboses, constituting a high-mortality clinical entity (1). The underlying pathophysiology is not fully elucidated; however, the prevailing model implicates a vicious cycle of transmural vascular inflammation, consequent endothelial

injury, and a secondary hypercoagulable state. This triad results in the simultaneous, paradoxical findings of vessel wall degradation leading to aneurysm and luminal thrombosis (2-3). Nosologically, HSS resides at the intersection of vasculitis and autoinflammation, sharing considerable clinical and histopathological overlap with Behçet's disease (BD). The common manifestations, including mucocutaneous ulceration, ocular inflammation, and vascular involvement have led many authorities to posit HSS as a severe variant or a distinct phenotype within the BD spectrum, sometimes referred to as "incomplete" BD (4-5). A critical distinction lies in HSS's predilection for catastrophic cardiopulmonary vascular pathology, which may present in relative isolation, suggesting potential unique pathogenic drivers (6). Diagnostic confirmation remains a significant clinical challenge, as definitive criteria are lacking. Consequently, diagnosis hinges on a high index of suspicion in the appropriate clinical context, supplemented by advanced cross-sectional imaging, most reliably computed tomography pulmonary angiography (CTPA), to visualize PAAs and thromboses (5). This diagnostic ambiguity often contributes to deleterious delays in intervention. The natural history of untreated HSS is grave, with mortality predominantly resulting from exsanguination due to PAA rupture or from massive thromboembolism. Given its rarity and poor prognosis, every documented case contributes to the evolving understanding of its management. This report details the case of a young male with an established diagnosis of Behçet's disease who progressed to develop the classic features of HSS. This progression underscores the necessity for rigorous surveillance for severe vascular sequelae in BD patients and emphasizes the imperative for timely and aggressive multimodal therapy to mitigate the syndrome's high fatality rate.

Case Presentation

A twenty-three-year-old male patient, employed as a private security officer, was admitted for evaluation of a progressive, one-month history of constitutional and respiratory symptoms. The patient carried a pre- existing diagnosis of Behçet's disease, established three years prior based on the clinical triad of recurrent bipolar aphthosis and recurrent episodes of uveitis. His outpatient therapeutic regimen consisted of a low- dose glucocorticoid taper, maintained on prednisone at five milligrams daily. The presenting symptomatology was characterized by an insidious onset of a non-productive chronic cough, followed by the development of exertional dyspnea consistent with New York Heart Association (NYHA) Functional Class II. This clinical course culminated in a single, low-volume episode of hemoptysis. These pulmonary manifestations were accompanied by nonspecific systemic symptoms, most notably a significant but unquantified reduction in body weight, suggesting an underlying inflammatory or vasculitic exacerbation.

Physical examination upon admission revealed a hemodynamically stable individual with normal vital parameters, including unremarkable respiratory rate and oxygen saturation. Cutaneous and mucosal pallor was evident; however, a comprehensive cardiopulmonary auscultation revealed no adventitious heart sounds, murmurs, or pulmonary crackles. The patient was afebrile, and the absence of stigmata of active infection shifted the initial diagnostic consideration towards a non-infectious complication of his underlying systemic inflammatory disorder.

Initial laboratory investigation, as summarized in table 1, revealed evidence of chronic inflammation and mild anemia. Notably, the C-reactive protein level was markedly elevated, indicative of significant systemic inflammatory activity. In contrast, procalcitonin levels remained within the normal range, effectively arguing against a concurrent bacterial infectious process. A comprehensive serological evaluation, including assays for antinuclear antibodies, antineutrophil cytoplasmic antibodies, and antiphospholipid antibodies, yielded negative results, thereby excluding other common autoimmune and pro-thrombotic syndromes, Standard coagulation studies and an extended thrombophilia screen were similarly within normal limits.

Table 1. Pertinent Admission Laboratory Findings

Parameters	Result	Reference Range	Interpretation
Hemoglobin	11 g/dL	13.5–17.0 g/Dl	Decreased
C-reactive Protein	115 mg/L	< 5 mg/L	Markedly Increased
Procalcitonin	0.1 ng/mL	< 0.5 ng/mL	Normal
Antinuclear Antibody	Negative	Negative	Normal
Antineutrophil Cytoplasmic Antibody	Negative	Negative	Normal
Antiphospholipid Antibodies	Negative	Negative	Normal

Diagnostic imaging provided the definitive etiological clarity. Initial chest radiography demonstrated nonspecific bilateral parahilar opacities. Subsequent contrast-enhanced computed tomographic angiography (CTA) of the thorax and abdomen revealed the pathognomonic findings of Hughes-Stovin Syndrome (figures 1-3). These included a saccular aneurysm arising from the posterior segmental branch of the right pulmonary artery, with associated thrombotic occlusion of an adjacent lateral segmental arterial branch.

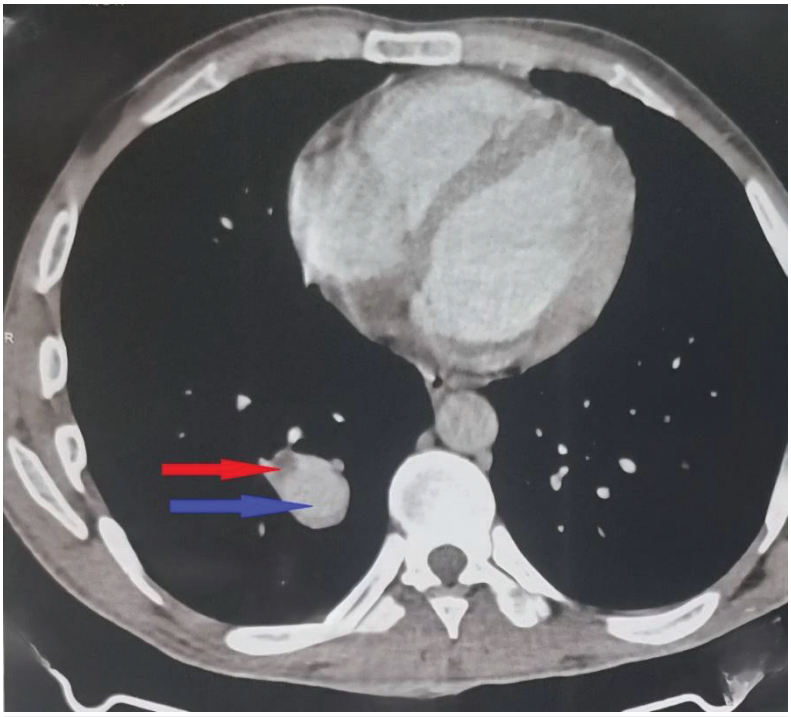

Figure 1. Contrast-enhanced CT with maximum intensity projection (MIP) clearly delineating the pulmonary artery aneurysm (blue arrow) and an adjacent mural thrombus (red arrow).

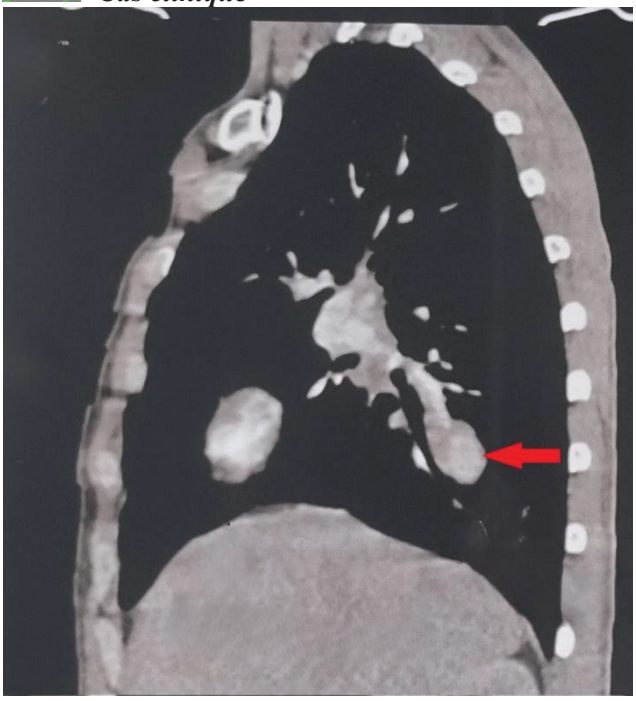

Figure 2. Parasagittal image localizing the saccular aneurysm (red arrow) within the central pulmonary arterial system

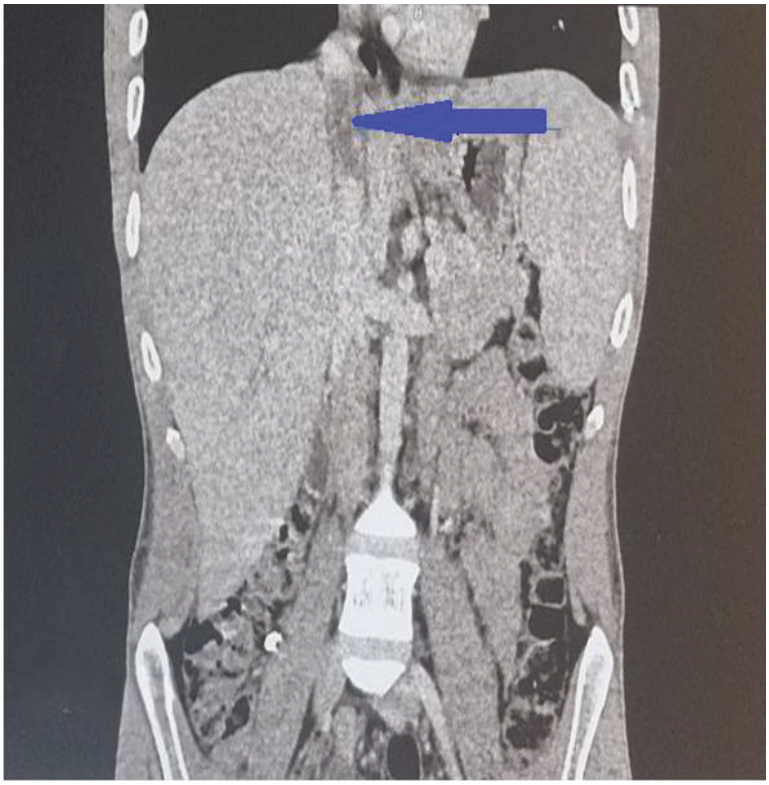

Figure 3. Coronal image localizing the thrombus (blue arrow) within the inferior vena cava

Further, an extensive, partially occlusive thrombus was visualized within the infrarenal inferior vena cava, propagating cephalad to the cavoatrial junction. Complementary transthoracic echocardiography confirmed the intracaval extension, identifying two distinct thrombi within the right atrium (figure 4): a mobile, pedunculated thrombus adherent to the atrial aspect of the tricuspid valve apparatus

and a second, fixed thrombus originating from the atrial roof.

The syndromic convergence of multiple pulmonary artery aneurysms and extensive deep venous thrombosis in a patient with established Behçet's disease confirmed the diagnosis of Hughes-Stovin Syndrome as a severe vascular phenotype. Given the preeminent mortality risk from aneurysmal rupture and hemorrhage, therapeutic anticoagulation was considered contraindicated. The therapeutic strategy was therefore exclusively centered on aggressive immunosuppression to halt the underlying vasculitic process. Induction therapy was initiated with intravenous pulse methylprednisolone administered at a dose of one gram daily for three consecutive days. This was followed by consolidation with high-dose oral prednisone dosed at one milligram per kilogram per day. For long-term disease modification and maintenance of remission, monthly intravenous cyclophosphamide induction was commenced at a dose of 750 milligrams per square meter of body surface area.

The clinical and radiographic response to this immunosuppressive regimen was favorable. The patient experienced complete resolution of hemoptysis following initiation of therapy. A follow-up imaging assessment conducted at six weeks demonstrated stabilization of the thrombotic burden within the venous system and no interval progression in the size or morphology of the pulmonary artery aneurysm, confirming an initial positive therapeutic response and adequate disease control.

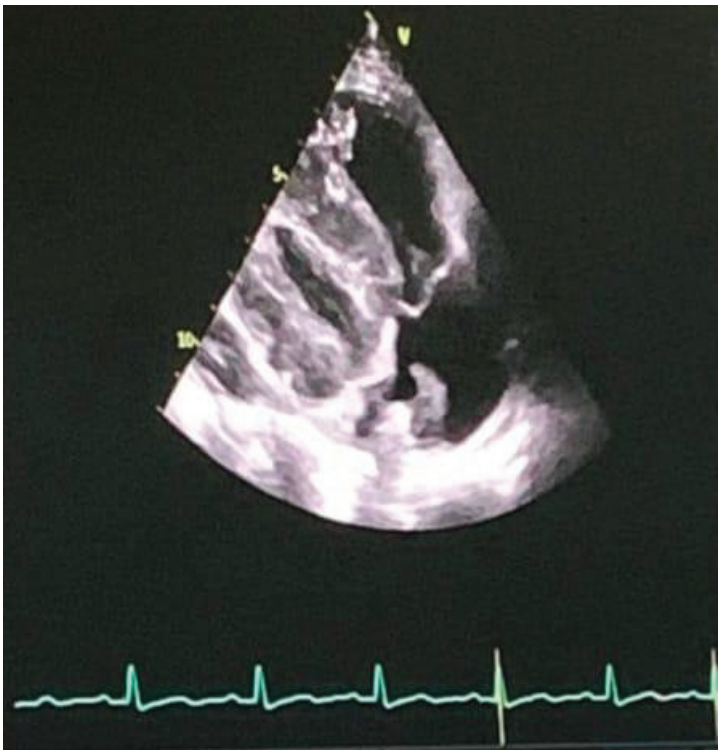

Figure 4. Transthoracic echocardiography (apical four-chamber view) demonstrating a hyperechoic mass within the right atrium, consistent with right atrial thrombus

Discussion

Hughes-Stovin Syndrome (HSS) remains a rare and enigmatic vasculitic disorder, often regarded as part of the clinical spectrum of Behçet's disease (BD) due to overlapping features such as recurrent thrombosis, pulmonary artery aneurysms (PAAs), and systemic inflammation (4,6). The case presented here illustrates this association, as the patient had a confirmed diagnosis of BD prior to developing vascular complications typical of HSS (1,6), including hemoptysis, dyspnea, and imaging-confirmed PAAs with intracardiac thrombi. This reinforces the hypothesis that HSS may represent an "incomplete" or vascular-predominant variant of BD, particularly when classic mucocutaneous or ocular manifestations are absent. The pathophysiology of HSS is still not fully understood, but it is believed to involve

immune-mediated vascular injury leading to aneurysm formation and thrombotic events (2,4). The presence of elevated inflammatory markers (CRP: 115 mg/L) in our patient further supports the role of systemic inflammation in driving vascular pathology. Diagnosing HSS remains a clinical challenge, as no validated diagnostic criteria exist (2). Instead, diagnosis relies on a combination of clinical suspicion, exclusion of alternative causes (e.g., infections, malignancies, or other vasculitis), and characteristic imaging findings (7). In this case, contrast-enhanced CT angiography played a pivotal role by revealing PAAs and thrombi in the pulmonary vasculature and inferior vena cava, while echocardiography identified high-risk intracardiac thrombi, a finding that significantly influences management decisions. The absence of thrombophilia or coagulopathy in laboratory workup further strengthened the diagnosis of HSS, as the condition typically occurs in the setting of normal coagulation profiles (4). The management of HSS is primarily immunosuppressive, mirroring the treatment approach for severe BD (1,4). High-dose glucocorticoids, often initiated as a pulse of methylprednisolone followed by oral prednisone, serve as first-line therapy to suppress acute inflammation. The addition of cyclophosphamide, as used in our patient, is critical for inducing remission and preventing aneurysm progression (1,5).

However, the role of anticoagulation remains controversial due to the heightened risk of catastrophic hemorrhage from fragile PAAs (1,3). In this case, anticoagulation was deferred in favor of immunosuppression, given the absence of acute thromboembolic events and the predominance of aneurysm-related complications. Long-term maintenance therapy with steroid-sparing agents such as azathioprine or biologic therapies (e.g., TNF-α inhibitors) may be considered to sustain remission and reduce glucocorticoid dependence.

Historically, untreated HSS carried a dismal prognosis, with mortality exceeding 50% due to fatal hemoptysis from ruptured PAAs or thromboembolic events (4). However, early recognition and aggressive immunosuppressive therapy have markedly improved outcomes, as demonstrated in our patient, who stabilized without further hemoptysis or thrombotic progression (1,5-6). This underscores the importance of prompt diagnosis and multidisciplinary collaboration among rheumatologists, pulmonologists, and cardiologists in managing this high-risk condition. Future research should focus on elucidating the underlying mechanisms of HSS, refining diagnostic criteria, and optimizing therapeutic strategies to further reduce morbidity and mortality.

Here is a comparative table summarizing the key characteristics of patients with Hughes-Stovin Syndrome (HSS) from the provided case reports (tableau 2).

Table 2. Summary of reported cases of pulmonary artery aneurysm with associated thromboembolic disease, including clinical features, management, and outcomes

Study Year	Age	Sex	Clinical Features	Treatment	Outcome
Mahjoub *et al.,* 2023 (6)	37	Male	Chronic cough, hemoptysis, dyspnea, pulmonary artery aneurysms (PAA), recurrent VTE	- Pulse methylprednisolone → oral prednisone - Cyclophosphamide (6 cycles)	Improvement (no new aneurysms at 6 months)
Mahjoub *et al*., 2023 (6)	55	Male	Chronic cough, DVT in left leg, aortic aneurysm (48mm), no lab abnormalities suggesting thrombophilia/immunologic disease. Clinical findings consistent with Behçet's Disease (HSS diagnosis).	- Pulse methylprednisolone - Cyclophosphamide (6 cycles) – Warfarin	Stable (no further aneurysm growth at 6 months)
Khursheed *et al*., 2023 (1)	47	Male	Hemoptysis, dyspnea, PAA (left lower lobe), VTE	- Corticosteroids , Cyclophosphamide	Complete remission (12-month follow-up)
Emad *et al*., 2021 (4)	26	Male	Initial DVT in right lower limb, followed by hemoptysis. Bilateral PAA on CT angiography secondary to pulmonary vasculitis.	- Corticosteroids - Cyclophosphamide	Complete remission
Khursheed *et al*., 2023 (1)	32	Male	Hemoptysis, dyspnea, PAA (bilateral), intracardiac thrombus, VTE	- Colchicine - Pulse methylprednisolone → oral steroids - Cyclophosphamide (6 cycles)	Complete remission (thrombus resolution, aneurysm stabilization)

Abbreviations: CRP: C-reactive protein; PA: Pulmonary artery; PAA: Pulmonary artery aneurysm; TNF: Tumor necrosis factor; VTE: Venous thromboembolism

The comparative table reveals that Hughes-Stovin Syndrome (HSS) primarily affects young to middle-aged males, presenting with a consistent triad of pulmonary artery aneurysms (PAAs), hemoptysis, and venous thromboembolism (VTE) (7,8,9). Treatment universally involves high-dose glucocorticoids and cyclophosphamide, with favorable outcomes (remission/stabilization) (5,6), while delayed intervention or aneurysm rupture led to mortality. Anti-TNF agents (e.g., infliximab) show promise in refractory cases (4), but anticoagulation remains controversial due to bleeding risks (3). The data underscore that early immunosuppression is critical for survival (1,6), while surgery is reserved for life- threatening complications, highlighting the need for prompt diagnosis and aggressive therapy to mitigate this rare vasculitis's high mortality.

Conclusion

Hughes-Stovin Syndrome (HSS) is a rare, life-threatening vasculitis at the intersection of vascular inflammation, aneurysm formation, and thrombosis, best understood as a severe, vascular-predominant phenotype of Behçet's disease (BD). This case, like others in the literature, confirms that the syndrome's hallmark triad of pulmonary artery aneurysms (PAAs), deep venous thrombosis, and systemic inflammation can emerge in patients with known BD, necessitating rigorous surveillance for such catastrophic complications. The principal challenge in HSS remains its timely recognition, as definitive diagnostic criteria are lacking. Diagnosis therefore relies on clinical vigilance. The therapeutic imperative is unambiguous: immediate and aggressive immunosuppression is the cornerstone of management. High-dose corticosteroids combined with cyclophosphamide form the standard induction regimen to suppress the destructive vascular inflammation, stabilize aneurysms, and prevent rupture. In contrast, anticoagulation carries a prohibitive risk of fatal hemorrhage from fragile PAAs and should generally be avoided unless a clear, compelling thromboembolic indication arises.

Historically associated with mortality exceeding 50%, the prognosis of HSS has improved markedly with contemporary management centered on early diagnosis and a multidisciplinary approach. This paradigm shifts underscores that mortality is now largely a consequence of diagnostic delay or therapeutic misadventure, rather than an inevitable outcome. Future efforts must focus on establishing formal diagnostic criteria to aid recognition, investigating targeted biologic therapies, and clarifying the precise pathogenic relationship between HSS and BD.

Conflict of Interest

The authors declare no conflicts of interest in relation to this work.

Contributions for Authors

All authors contributed to the conception and design of the case report, data collection, analysis, and interpretation. All authors participated in drafting or critically revising the manuscript for important intellectual content and approved the final version to be published.

References

1. Khursheed T, Masood A, Khan MS, Sharif M, Shah S, Miraj MA. Hughes-Stovin Syndrome: A Case Report on a Rare, Life-Threatening Vasculitis. *Mediterr J Rheumatol* 2023; **29**: 565-569.
2. Ukemenam C, Muppaneni SP, De D and Lacasse A. Hugh-Stovin syndrome: the 'incomplete Behçet's disease'. A case study of a young adult with recurrent pulmonary embolism and pulmonary arterial aneurysms. *J Community Hosp Intern Med Perspect* 2021; **21**: 566-567.
3. Ascoli Marchetti A and Belvivere L. A Rare but Fatal Behçet Variant: The Hughes-Stovin Syndrome – Successful Case Report and New Evidence from Literature Review. *Aorta (Stamford)* 2023; **11**: 156-161.

4. Emad Y, Ragab Y, El-Shaarawy N and Saab AN. Hughes-Stovin Syndrome: A Case-Based Systematic Review. *Clin Rheumatol* 2021; **40**: 4427-4438.
5. Bin Pervez M, Iqbal B, Nasir N and Fatimi SH. Hughes Stovin Syndrome: A Case of Recurrent Pulmonary Artery Aneurysms. *Ann Thorac Surg* 2020; **110**: 355-356.
6. Mahjoub L and Kallala MY. Hughes-Stovin Syndrome: A Rare Entity and Challenging Therapeutic Decision. Case Series and Review of the Literature. *F1000Research* 2023; **12**: 1001.
7. Gheit A, Hegab S and Al-Nakshabandi N. Radiological Findings in Hughes-Stovin Syndrome: A 10-Year Experience. *Ann Thorac Med* 2018; **13**: 55-60.
8. Khalid U and Saleem M. Hughes-Stovin Syndrome. *Orphanet J Rare Dis* 2011; **6**: 15.
9. Hughes JP and Stovin PG. Segmental Pulmonary Artery Aneurysms with Peripheral Venous Thrombosis. *Br J Dis Chest* 1959; **52**: 116-123.

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