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Recurrent Haemoptysis Revealing Hughes-Stovin Syndrome

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Abstract **Case Report**

Hughes-Stovin Syndrome (HSS) is a rare and severe disorder characterized by the combination of pulmonary artery aneurysms and deep vein thrombosis. We report the case of a 42-year-old male presenting with recurrent hemoptysis revealing HSS. Management consisted of high-dose corticosteroids and cyclophosphamide. The patient showed marked clinical and radiological improvement. HSS, as a rare form of Behçet's disease, is associated with high morbidity and mortality due to thrombosis and potential rupture of aneurysms. Immunosuppression remains the cornerstone of management. Anticoagulation is controversial and requires careful consideration. Early diagnosis and prompt treatment are critical in managing HSS.

Keywords: Hughes-Stovin Syndrome, Behçet's disease, Pulmonary artery aneurysm, Deep vein thrombosis, Immunosuppression.

INTRODUCTION/BACKGROUND

Hughes-Stovin Syndrome (HSS) is a rare and life-threatening disorder considered a cardiovascular variant of Behçet's disease. It was first described in 1959, characterized by the coexistence of pulmonary artery aneurysms and deep vein thrombosis. With fewer than 60 cases reported globally, its exact pathogenesis remains poorly understood, emphasizing the need for heightened clinical awareness [1]. This case highlights the importance of early diagnosis and multidisciplinary management in improving patient outcomes.

CASE DESCRIPTION

A 42-year-old male with no history of smoking or tuberculosis presented with hemoptysis occurring 2–3 times per week for 14 months, associated with a dyspnea grade 2 using the modified Medical Research Council (mMRC) scale. Physical examination revealed generalized pallor, acne, multiple oral and genital ulcers recurring three times annually over five years and abdominal examination revealed peri-umbilical collateral venous circulation (Figure1).

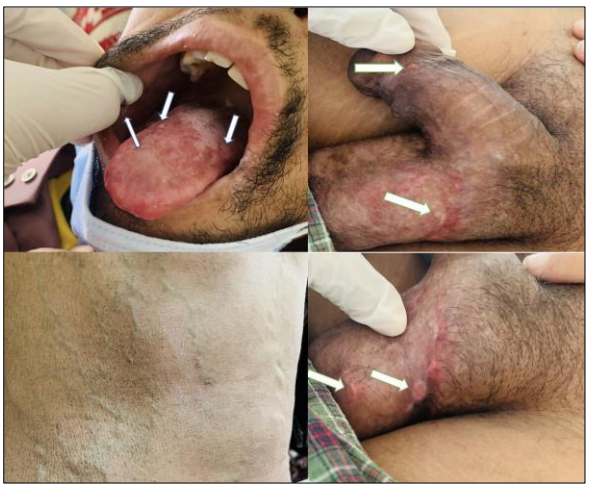

Figure 1: Clinical findings: Oral and genital aphthous ulcers and peri-umbilical collateral venous circulation

The chest x-ray revealed a left hilar opacity suggestive of a vascular origin (Figure2).

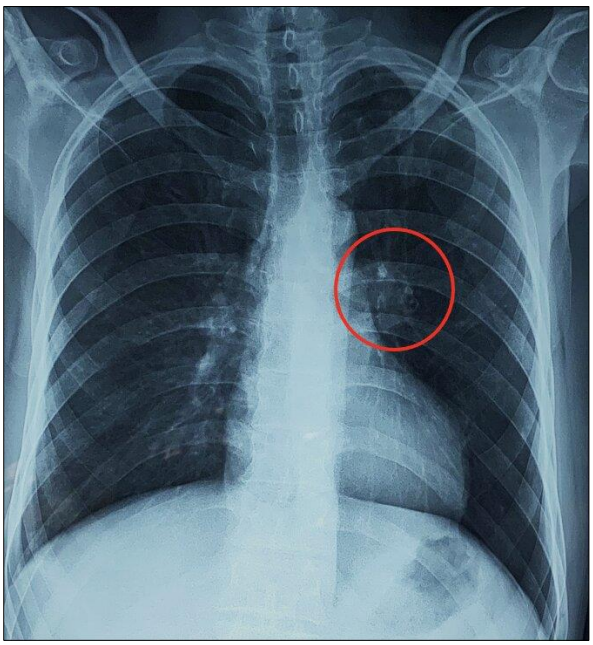

Figure 2: Chest X-ray showing a left hilar opacity

The thoraco-abdominal computed tomography angiography (CTA) identified a thrombosed aneurysm of the left pulmonary artery branch measuring (21×13 mm, length 30 mm) with surrounding ground-glass opacity indicative of intra-alveolar hemorrhage and chronic pulmonary embolism of the left pulmonary artery, associated with thrombosis of the inferior vena cava and the inferior mesenteric vein (Figure3).

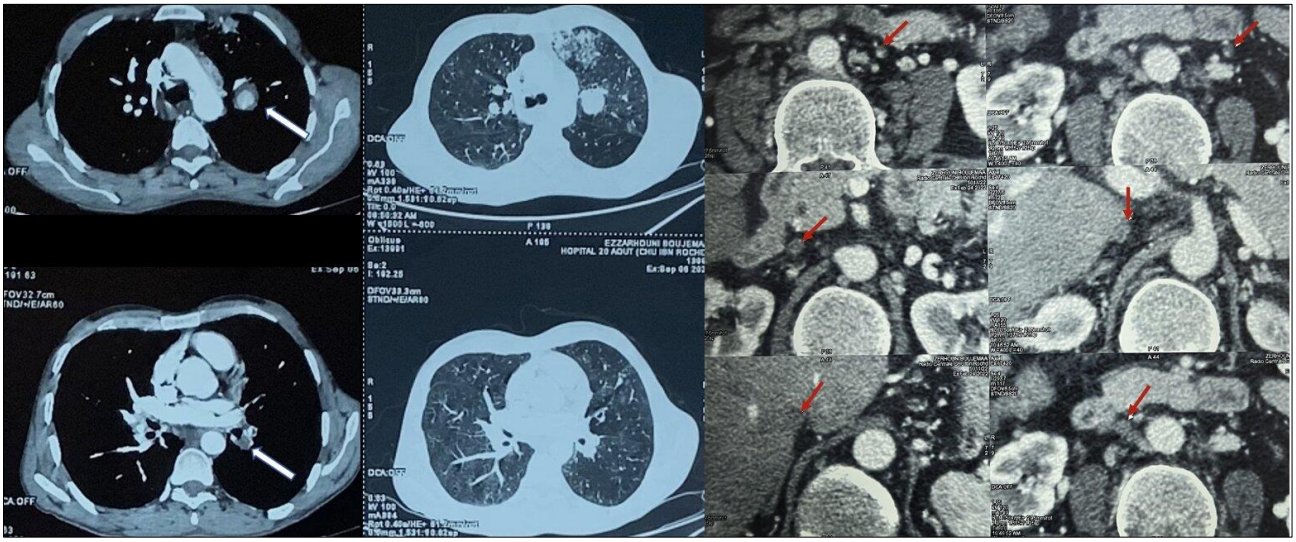

Figure 3: Thoraco-abdominal CTA showing a thrombosed aneurysm of the left pulmonary artery branch

Blood count test showed severe anemia (hemoglobin 6 g/dL) necessitating two packed red cell units' transfusion, raising hemoglobin to 9.1 g/dL. Coagulation tests were normal. Additional investigations included a negative pathergy test, no active uveitis on ophthalmological evaluation. Echocardiography revealed mildly dilated right heart chambers, tricuspid regurgitation, and normal left ventricular function (ejection fraction 63%) and lower limb Doppler ultrasound showed no abnormalities.

The diagnosis of Hughes-Stovin Syndrome, a particular form of Behçet's disease, was established based on the International Criteria for Behçet's Disease (2013), considering bipolar aphthosis, cutaneous signs, and vascular involvement.

Following negative infectious screening, the patient received high-dose methylprednisolone (15 mg/kg/day) for three days, followed by oral prednisone (1 mg/kg/day), tapered gradually after four weeks. Cyclophosphamide induction therapy (600 mg/m²) was

received on day four with subsequent doses every two weeks for the first month and every three weeks thereafter, totaling six doses. The adjuvant therapy included colchicine, potassium, calcium, vitamin D supplementation. Anticoagulation was not initiated because of the bleeding risk.

The patient improved significantly with no further episodes of haemoptysis or dyspnea reported and disappearance of bipolar aphtosis (Figure4).

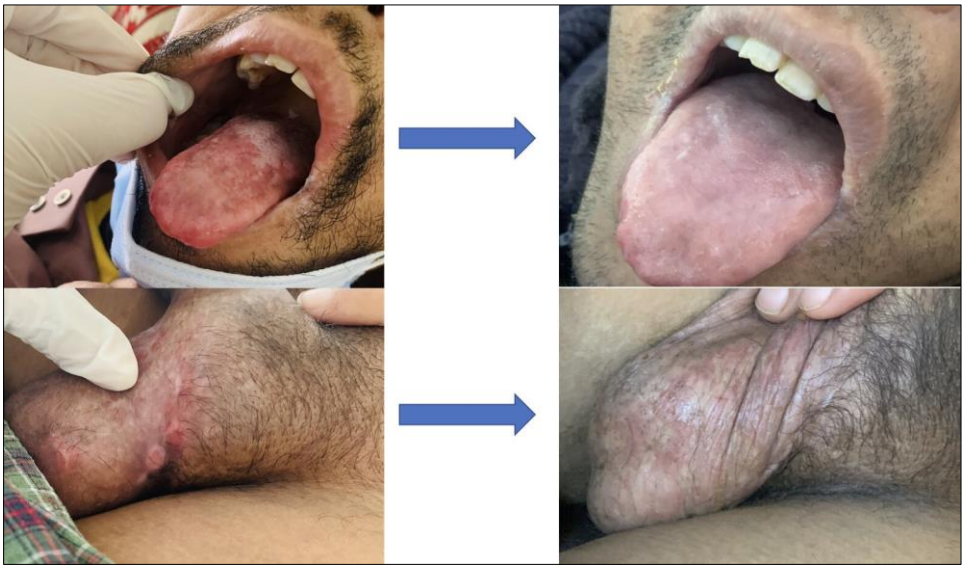

Figure 4: Clinical improvement: Disappearance of bipolar aphthosis

Follow-up CT angiography revealed complete resolution of vascular abnormalities (Figure5).

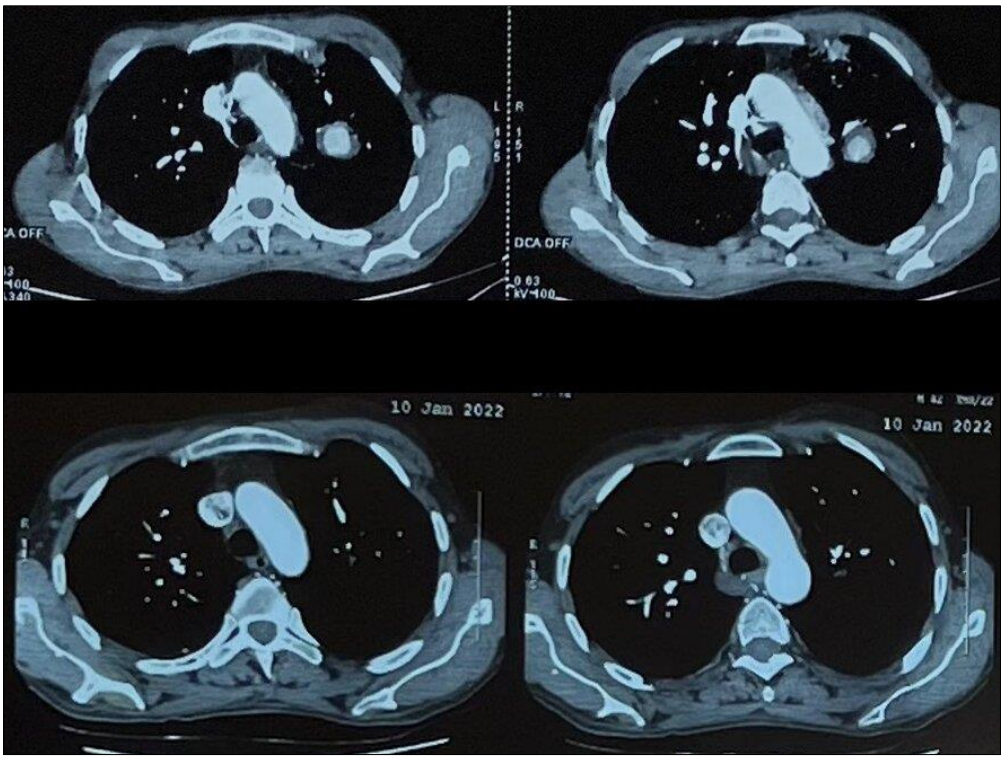

Figure 5: Follow-up CTA revealing complete resolution of vascular abnormalities

DISCUSSION

Hughes-Stovin Syndrome is a rare disorder that typically presents with a combination of pulmonary artery aneurysms and venous thrombosis. First described by Hughes and Stovin in 1959, the syndrome has been linked with conditions like Behçet's disease, but can also occur in angiodysplasia or post infectious diseases. The

HSS is associated with high mortality rate because of the combined risk of thrombosis and complications such as ruptured pulmonary aneurysms and severe hemoptysis [1].

The pathogenesis of HSS is thought to be linked to autoimmune or inflammatory processes, although specific mechanisms are still under investigation. Most commonly, cardinal features like uveitis and bipolar aphthosis are often incomplete, patients present with nonspecific symptoms such as hemoptysis, chest pain, and dyspnea, leading to potential misdiagnosis. Imaging techniques like CT scan angiography (gold standard) and Doppler ultrasound play a critical role in diagnosis, allowing clinicians to identify the characteristic pulmonary artery aneurysms and thrombotic events that can be atypical [2]. As proposed by HSS International Study Group (HSSISG), aneurysms can be unstable or stable. For instance, our patient has developed venous thrombosis in a less common location with a stable aneurysm. This case is significant because it highlights the clinical challenges of diagnosing and treating Hughes-Stovin Syndrome, especially given its rare and nonspecific presentation. It reinforces the importance of considering HSS in the differential diagnosis for patients with unexplained venous thrombosis and pulmonary symptoms, even in the absence of classic associations like Behçet's disease.

The treatment should be multimodal combining corticosteroids and immunosuppressive therapy according to the new updated EULAR (European League against Rheumatism) recommendations for the management of Angio-Behçet disease [3]. The anticoagulation should be considered with extreme caution and only in clinical situations where the benefits clearly outweigh the risks [4]. Embolization is a less invasive, enabling selective treatment of multiple bilateral aneurysms and a high risk of major bleeding. Severe cases, especially with aneurysm rupture or aneurysm more than 6cm, may require surgical or endovascular interventions [5].

CONCLUSION

This case report emphasizes the need for early recognition of HSS, prompt and tailored interventions, alongside a high index of suspicion, are essential in managing this complex and challenging syndrome

Author Contributions:

All authors contributed to the conception, drafting, and revision of this manuscript. All authors approved the final version and are accountable for its content.

Disclosure:

Conflict of Interest: The authors declare no conflict of interest.

Patient Consent: The authors confirm acquisition of written informed consent from the patient for the publication of this case report and accompanying images.

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Learning Points:

1. Hughes-Stovin Syndrome (HSS) is a rare cardiovascular variant of Behçet's disease.
2. Early recognition of HSS can prevent life-threatening complications such as aneurysm rupture and thrombosis.
3. Multidisciplinary management is crucial for improving outcomes and reducing mortality in HSS.

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