Case Report

Hughes–Stovin Syndrome

Abstract

We present an extremely rare case of Hughes–Stovin syndrome, of which we believe <60 cases have been reported in English medical literature. We wish to draw the attention of our fellow cardiologists to consider this when coming across patients with pulmonary artery aneurysm in their clinical practice. Appropriate treatment, if instituted promptly and early in the course of the disease, has the potential to induce remission.

Keywords: *Hughes–Stovin syndrome, pulmonary artery aneurysm, venous thrombosis*

Idris Ahmed Khan, Vivek Sullere

Department of Cardiology, Bombay Hospital Indore, Madhya Pradesh, India

Introduction

Hughes–Stovin syndrome (HSS) is named after two British physicians, Drs. John Patterson Hughes and Peter George Ingle Stovin. They in 1959, first described the syndrome (deep venous thrombosis and segmental pulmonary artery [PA] aneurysms) in four male patients having PA aneurysms (PAAs).[1] HSS is a rare disorder of unknown etiology. It is characterized by the findings of thrombophlebitis and multiple pulmonary and/or bronchial aneurysms.[2]

Another syndrome, Behcet's disease (BD), is also associated with aneurysm–thrombosis combination. BD constitutes the syndrome of hypopyon, iritis, and orogenital ulcers as described in 1937 by H. Behcet.[3]

In a patient having the findings of aneurysms and thrombosis, after ruling out other causes, the patient either has HSS or BD. However, if its other distinctive features are absent in the patient, BD can be ruled out.

HSS is an extremely rare disorder with approximately 57 published cases in English medical literature up to May 2021.[4] In this report by the HSS International Study Group, published in the *International Journal of Cardiology*, authors have critically analyzed and presented the syndrome's clinically important issues summarized as follows.

At initial presentation, deep-vein thrombosis (DVT) was observed in 29 (33.3%), thrombophlebitis in 3 (5.3%), hemoptysis in 24 (42.1%), and diplopia and seizures in one patient each. During the course of disease, DVT occurred in 48 (84.2%) patients, and superficial thrombophlebitis was observed in 29 (50.9%). Hemoptysis occurred in 53 (93.0%) patients and was fatal in 12 (21.1%). PAAs were bilateral in 53 (93%) patients. PAA was located within the main PA in 11 (19.3%), lobar in 50 (87.7%), interlobar in 13 (22.8%), and segmental in 42 (73.7%). Fatal outcomes were more common in patients with inferior vena cava thrombosis ($P = 0.039$) and ruptured PAAs ($P < 0.001$). Death was less common in patients treated with corticosteroids ($P < 0.001$), cyclophosphamide ($P < 0.008$), azathioprine ($P < 0.008$), and combined immune modulators ($P < 0.001$). No patients had uveitis; 6 (10.5%) had genital ulcers and 11 (19.3%) had oral ulcers.

Case Report

A previously healthy 24-year-old Indian male presented with symptoms of, on and off fever, for 9 months and cough with hemoptysis for 6 months.

His vital parameters were within normal limits. His routine complete blood count, erythrocyte sedimentation rate, and coagulation tests were within normal limits. Antinuclear antibodies, antineutrophil

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Address for correspondence:
Dr. Vivek Sullere, Room 28, 1st Floor, Bombay Hospital, Indore, Madhya Pradesh, India.
E-mail: drsullere@gmail.com

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cytoplasmic antibodies, and venereal disease research laboratory tests were negative. Chest X-ray showed round opacity in the left upper lobe [Figure 1].

A plain and contrast computed tomography chest showed saccular aneurysmal dilatation with mural thrombosis of descending/lower lobe segmental branches of PA on either side and upper lobe segmental artery on the left side. The average size of aneurysmal dilatation was about 2.5–3.2 cm. Filling defect/thrombosis was seen in one of the segmental branches in the left lower lobe [Figure 2].

Diagnosis of HSS was made on the basis of clinical features and PA aneurysm. Patient's symptoms resolved with intravenous (IV) steroids and IV cyclophosphamide.

The patient reviewed after 1 month showed clinical improvement; no episode of hemoptysis or fever. The size of PA aneurysm had reduced [Figure 3].

Discussion

Clinical features

Approximately 25% of patients having HSS develop thromboembolism, arterial aneurysms, and vascular occlusions. The typical clinical features of HSS are related to the presence of pulmonary aneurysms and peripheral venous thrombosis. Patients can have seizures, diplopia, and cephalalgia due to raised intracranial pressure because of cerebral venous sinus thrombosis.[1,5] Aneurysms seen in HSS may be single, multiple, unilateral, or bilateral. The aneurysms in HSS generally involve the pulmonary and bronchial arteries but can also occur anywhere in systemic circulation. Recurrent phlebitis often involves the large vessels resulting in thromboembolism, with even reports of thrombosis of the vena cava, cardiac chambers, jugular vein, iliac vein, femoral vein, and dural sinuses.[5-7]

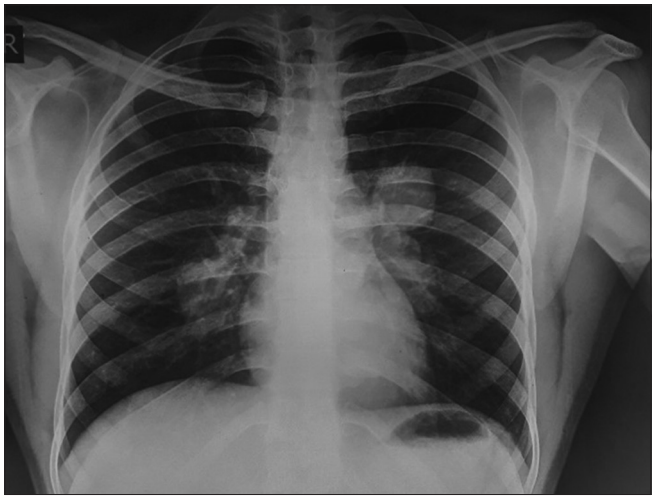

Figure 1: Chest X-ray PA view: rounded opacity in the left upper lobe. PA: Pulmonary artery

Etiopathogenesis

The exact etiology and pathogenesis of HSS are unknown. Several theories have attempted to explain the manifestations of this rare entity.[7,8] Consensus is that vasculitis is the primary pathologic process underlying HSS.[6]

1. Infections–Septic embolisms and abscesses have been proposed[2,9]
2. Angiodysplasia of bronchial arteries is another hypothesis[7,8,10]
3. Possible manifestation of Behcet's Syndrome. Some authors suggest that HSS may be a partially manifested BD due to their similar findings of vasculitis associated with PAAs[5,7,11,12]
4. Immunogenetics–BD has been associated with the human leukocyte antigen B51.

Differential diagnosis

Among the causes of PA aneurysm, there are two idiopathic, similar, syndromes that are associated with thrombosis: BD and HSS. Clinical findings unique to BD are recurrent genital ulceration, eye lesions, skin lesions, iritis, arthralgia, and a positive pathergy test.[13-15]

Management

There are no standard treatment guidelines for the management of HSS.

Figure 2: Contrast CT: saccular aneurysm, dilatation with mural thrombosis of lower lobe segmental branches of pulmonary artery on either side, upper lobe segmental artery on the left side. The average size of aneurysmal dilatation is 2.5–3.2 cm. CT: Computed tomography

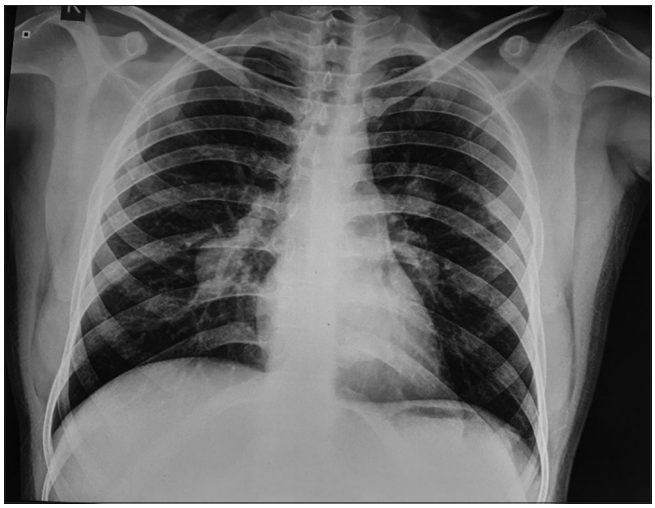

Figure 3: Chest X-ray PA view: regression of aneurysmal dilatation after cyclophosphamide and steroid therapy. PA: Pulmonary artery

a. Immunosuppressive therapy: A combination regimen of glucocorticoids and cyclophosphamide has been employed as first-line medical management in the treatment of HSS.[6,13] Monthly pulses of cyclophosphamide (1 g) plus prednisolone (1 mg/kg/day). The latter is then tapered for several months to a dose <30 mg/day[16]
b. Antibiotics: Antibiotics have no proven role in the management ofHSS[1,9,17,18]
c. Anticoagulants and thrombolytic agents: Anticoagulants and thrombolytic agents are generally considered contraindicated due to an increased risk of fatal hemorrhage, even though they have a beneficial role in an embolic state[19]
d. Surgical consultation
e. Transcatheter arterial embolization.

Prognosis

Aneurysms of arterial origin have a poorer prognosis than venular aneurysms.[6] In particular, pulmonary artery aneurysms can cause hemoptysis and have a poor prognosis.

Conclusion

Early diagnosis and timely intervention are 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.

Consent

Written informed consent was obtained from the patient for publication of this case report.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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