Hughes–Stovin syndrome: the diagnostic and therapeutic challenges of peripheral pulmonary artery aneurysms

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Case report

CASE REPORT

Hughes–Stovin syndrome: the diagnostic and therapeutic challenges of peripheral pulmonary artery aneurysms

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Learning points for clinicians

Pulmonary artery aneurysms (PAA) have a diverse differential diagnosis. Vasculitic features, without evidence of infection, suggests Hughes–Stovin syndrome (HSS) or Behçet's disease (BD). In solitary peripheral PAA, we recommend operation or embolization for rapidly growing or high-pressure aneurysms. Multiple, small PAA warrant early immunosuppression. Anticoagulation is typically contraindicated by bleeding risk.

Introduction

Pulmonary artery aneurysms (PAA) are uncommon, with a diverse differential diagnosis. Although most are asymptomatic at diagnosis, fatal aneurysm rupture occurs in one-third and dissection in one-fifth.[1] Hughes–Stovin syndrome (HSS) is a rare condition, characterized by pulmonary/bronchial artery aneurysms and thrombophlebitis, without diagnostic features of Behçet's disease (BD).[2] The diagnostic work-up, role of immunosuppression, anticoagulation and operative indications remain poorly defined.

Case presentation

A 21-year-old male from Missouri, USA was admitted with haemoptysis following 1 month of intermittent fevers, non-productive cough and weight loss. Past medical history was significant for recurrent oral ulcers. Physical examination found fever, tachycardia and reduced air entry to his right lung base. Ophthalmological examination excluded uveitis, vitritis or retinal vasculitis and he did not have genital ulceration or skin manifestations of BD. He developed superficial thrombophlebitis at venous access sites but no pathergy. There was no previous trauma or features of connective tissue disorders.

Laboratory testing found microcytic anaemia (Hb 104 g/l, MCV 78 fl), normal white blood cells, raised CRP (73 mg/l), ESR (84 mm/h), ferritin (870 μg/l) and D-dimer (383 ng/ml). Urinalysis and protein: creatinine ratio were normal. Chest x-ray demonstrated a right lower zone circumscribed lesion. He was treated with antibiotics for community-acquired pneumonia. Computed tomography (CT) demonstrated a 35 mm right lateral segmental PAA (Figure 1a), with multiple pulmonary artery thromboses. CT angiogram found normal mesenteric, hepatic and renal vasculature. Echocardiogram excluded pulmonary artery hypertension (PAH) and structural cardiac disease. Our differential was vasculitis (including BD/HSS, Takayasu's arteritis, granulomatosis with polyangiitis and eosinophilic granulomatosis with polyangiitis) vs. infection (including syphilitic, mycotic or Rasmussen aneurysms).

Extensive microbiological investigation found no evidence of infection. Immunological testing found a weak-positive ANA (1:80 titre, speckled) and raised C3/C4. PAA resection by thoracotomy and lower lobe basal segmentectomy was performed. He returned to America post-operatively for immunosuppressive treatment. Tissue pathology demonstrated necrotising lymphocytic vasculitis with pulmonary infarction. Based on clinical and histopathological findings, without meeting BD criteria, he was diagnosed with HSS.

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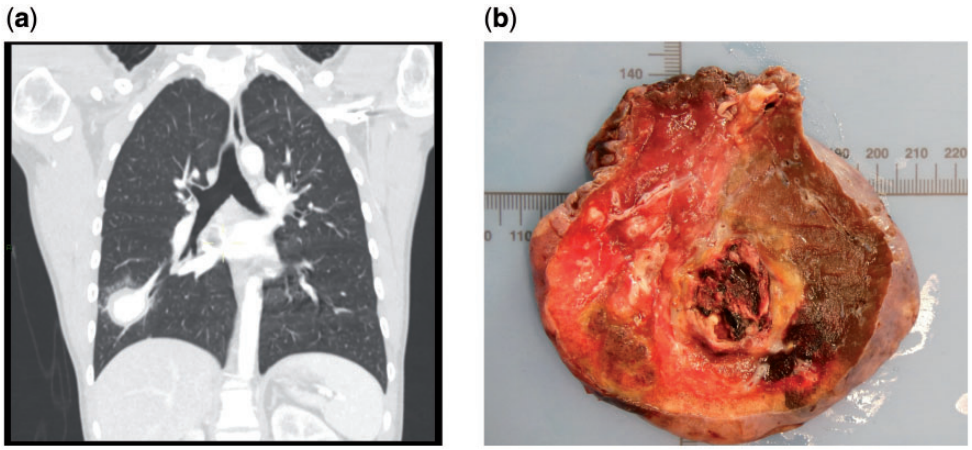

Figure 1. (**a**) CT chest demonstrating 35 mm right lateral segmental PAA. (**b**) Resected pulmonary artery aneurysm and adjacent pulmonary infarction.

Discussion

Without PAH (mean pulmonary artery pressure ≥ 25 mmHg) or features of trauma/iatrogenic injury, congenital abnormalities or connective tissue disorder, the differential diagnosis of PAA is infection vs. vasculitis. Aneurysm-thrombosis complexes, with negative microbiology/mycology testing, suggest HSS or BD-PAI.

Both conditions predominantly affect young males, present with fever and haemoptysis, and involve the right lower lobar artery.[2,3] HLA-B51 is associated with BD and reported in the only case of HSS tested.[2] Due to significant correlation, authors suggest that HSS is a variant form of BD.[2,3] Recently, the concept of 'vasculo-Behçet's,' with higher frequencies of vascular and cardiac lesions and less ocular, genital or joint involvement has emerged.[4]

Operative thresholds for HSS/BD-PAI are controversial and based on case reports. For solitary lesions, we recommend that large (≥30 mm) or expanding (≥3 mm in 6 months) aneurysms, PAH and evidence of rupture or dissection should be considered as operative indications based on risk of aneurysm rupture.[1] Multiple, bilateral PAA and small, stable lesions should be treated with immunosuppression. Immunosuppression may stabilize or promote regression of PAA.[2,5] Transcatheter arterial embolization offers an alternative for inoperable cases. However, long-term outcome data does not exist and the procedures risk aneurysm rupture, distal infarction, and procedural failure, particularly for larger aneurysms.[3] Despite the prothrombotic nature of the condition, anticoagulation carries a high risk of fatal bleeding and has not provided survival benefit in retrospective studies.[3,6] It should therefore be reserved for cases where the risk of thrombosis clearly outweighs the risk of haemorrhage.

Future research should focus on the immunogenetic relationship between HSS and BD, optimization of immunosuppressive strategies through prospective controlled trials and evaluation of the long-term outcomes of embolization procedures.

Conflict of interest: None declared.

Informed consent

Written informed consent was obtained from the individual included in the report.

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