

Is Warfarin Indicated in Behcet's Syndrome Associated with Thrombosis? Reflections on Treatment and Death of a Patient

Yong Chen, Jian-Long Guan

Department of Rheumatology and Immunology, Huadong Hospital Affiliated to Fudan University, Shanghai 200040, China

A 37-year-old male patient was admitted to Huadong Hospital with diagnosis of Behcet's syndrome (BS) for 5 years and deep vein thrombosis (DVT) for 2 months. The patient complained of recurrent oral ulceration, approximately five to seven times per year starting 5 years before presentation. He had a history of genital ulceration one to two times every year. The patient did not follow a strict treatment regimen. Seven months before admission, he developed an inflammatory reaction in his conjunctiva bilaterally, with diminution of vision in both eyes. Two months before admission, he presented with swelling and pain in his left lower limbs. An imaging examination in another hospital suggested DVTs in both lower limbs. He was treated with glucocorticoid and warfarin. The symptoms related to thrombosis rapidly progressed with swelling and pain, leading to his losing the ability to walk [Figure 1a]. B-ultrasound in our hospital showed varicosity and thrombosis (complete occlusion) of the soleus venous plexus [Figure 1b]. Erythrocyte sedimentation rate was 45 mm/h, fibrinogen was 3.5 g/L (normal range 2–4 g/L), and D-dimer was 2.59 mg/L (normal range 0–0.55 mg/L).

Considering his history of irregular treatment, we prescribed glucocorticoid, thalidomide, and cyclosporine A. To prepare him for gastrointestinal endoscopy, we discontinued his warfarin, which he had been taking for 2 months before admission. His general condition improved quickly; his pain decreased rapidly, and he was able to rise from bed and walk. On his 7th day in hospital, the patient complained of upper abdominal pain with sweating at night. The pain lasted for around 2 h, accompanied by dyspnea. The resident doctor on call arranged for urgent abdominal computed tomography (CT), which revealed multiple microstones in the left kidney calices and upper ureter and multiple cystic lesions of the liver. No other abnormality was detected. No specific treatment was applied apart from indomethacin administered per rectum.

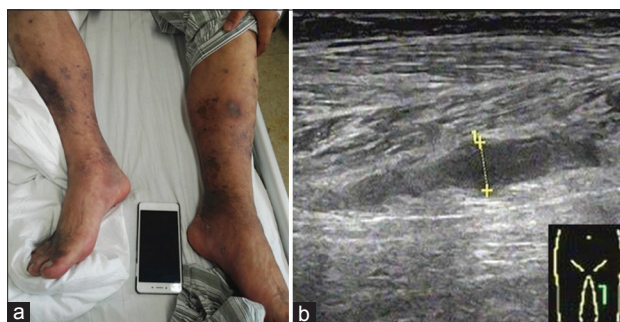


Figure 1: Bilateral lower limbs were swollen and with ecchymosis skin (a). B-ultrasound showed varicosity and thrombosis (complete occlusion) of soleus venous plexus (b).

His symptoms recurred the following morning. He complained of chest pain accompanied by dyspnea and sweating. Electrocardiogram and blood tests excluded coronary heart disease. Fibrinogen was 38.7 g/L and D-dimer was 10.36 mg/L. Shortly thereafter, the patient went into cardiac arrest. The patient was declared dead after 1 h of resuscitation.

BS is a systemic vasculitis with vessel involvement of all types and sizes. Vessel disease is one of the leading causes of mortality in BS. Systemic inflammation appears to play a major role in the pathophysiology of thrombosis in BS, whereas other thrombophilic factors are less important. BS may thus be considered a model of inflammation-related thrombosis.^[1] The objective of treatment in vascular BS is suppression of inflammation or autoimmunity, with

Address for correspondence: Prof. Jian-Long Guan,
Department of Rheumatology and Immunology,
Huadong Hospital Affiliated to Fudan University,
Shanghai 200040, China
E-Mail: guanjianlong@medmail.com.cn

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prevention of complications.^[2] The role of anticoagulants is controversial in the management of BS associated with thrombophlebitis. Immunosuppressive therapy (with cyclophosphamide and corticosteroids) was reported to be preferred for treatment of large vessel involvement. Anticoagulant therapy may be beneficial in BS patients with thrombosis if DVT persists after initiation of immunosuppressive therapy, provided that aneurysm is ruled out.^[3]

Warfarin is indicated for the tendency for thrombosis or as secondary prophylaxis in individuals who have already developed a thrombus. Warfarin treatment may help reduce the risk of embolism.^[4] Although an autopsy was not conducted, considering the presence of DVTs and the clinical manifestations after warfarin discontinuation, we highly suspect the patient died from pulmonary embolism. Although the use of anticoagulants in BS with thrombosis is still debatable, we believe warfarin should prevent the death in this patient, the treatment guidelines should be revisited, and further clinical studies should be performed.

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 names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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