A missed Behçet’s case presenting with spontaneous epidural hematoma
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Abstract
Spinal vascular events related to Behçet’s disease are relatively uncommon. Deep vein thrombosis is the most frequent vascular involvement. Anticoagulant therapy is a debated issue in Behçet’s disease. In this case report, we present a patient with a delayed diagnosis of Behçet’s disease after development of cervical epidural hematoma following anticoagulant therapy due to deep venous thrombosis. Anticoagulant therapy without immunosuppressive therapy leading to uncontrolled systemic inflammation may be the cause of spinal epidural hematoma. On the other hand, epidural vascular involvement as a vasculitic manifestation of Behçet’s disease in conjunction with anticoagulant therapy may have predisposed to spontaneous bleeding via increased fragility or microaneurysms in epidural arteries. In this case report, the importance of a multidisciplinary approach to Behçet’s disease is emphasized.

Keywords: Behçet’s disease, spinal epidural hematoma, anticoagulants

Introduction
Behçet’s disease is a multisystemic disease which can present with various organ system involvements such as mucocutaneous, ophthalmic, musculoskeletal, vascular, neurogenic, gastrointestinal manifestations. The disease may evolve by the cumulation of symptoms within months/years. Holistic approach to the Behçet’s patient and immunosuppressive treatment are essential in the management (1).

Vascular manifestations have been reported in 7.7-38% of Behçet’s patients. Deep vein thrombosis is the most frequent vascular involvement (2). Spinal vascular events related to Behçet’s disease are relatively uncommon (3).

Spontaneous spinal epidural hematoma (SEH) is a rare condition with an estimated incidence of 0.1/100,000 (4). Coagulopathy, vascular malformation, and malignancy are non-traumatic causes of SEH (5). On the other hand, the use of unfractionated heparin, warfarin, aspirin, and tissue plasminogen activator may predispose to spontaneous SEH (6). Anticoagulant therapy in Behçet’s disease is a debated issue. There is no controlled study regarding the necessity of anticoagulant therapy in vascular involvement of Behçet’s disease (1). Moreover, inattentive anticoagulation therapy increases the risk of hemorrhage especially in the presence of an aneurysm or other vascular malformations.

To our knowledge there has been no report of SEH in a Behçet’s case treated with/without anticoagulant therapy. In this case report, we present a patient with a delayed diagnosis of Behçet’s disease after development of cervical SEH following anticoagulant therapy due to deep venous thrombosis.

Case Presentation
A 42-year-old man was admitted to our outpatient clinic due to weakness in extremities. Three months ago, numbness in his extremities, progressive muscle weakness causing impairment of gait and urinary retention developed within a 24-hour period following a sudden and severe neck pain. No history of antecedent trauma, iterative coughing or spinal invasive procedure was reported. He had been on warfarin prescribed by a cardiovascular surgeon for three months because of bilateral deep venous thrombosis with pulmonary embolism. Prothrombin time (PT) expressed as international normalized ratio (INR) was not monitored regularly during this period. MRI revealed a cervical epidural hematoma spanning posterolaterally on the right side between C4 and C6 vertebral levels (Figure 1, 2). INR value was found to be 3.81 and an urgent surgical decompression within 36 hours after the first symptom was carried out.

At the time of admission to our outpatient clinic following surgery, muscle strength and ambulation had improved. However he complained about diminished effort level, difficulty in walking and urinary
Spontaneous SEH may affect all size of arteries and veins. Vasculitis, arterial aneurysm and arteriovenous malformations are characterized by vessel fragility and predispose to spontaneous bleeding which may lead to neurologic deficits (10). Considering deep venous thrombosis, uveitis and oral aphthous history, our patient may have been suffering from longstanding inflammation of undiagnosed Behçet’s disease. Consequently epidural vascular involvement in conjunction with anticoagulant therapy may have predisposed to spontaneous bleeding via increased fragility or microaneurysms in epidural arteries. Since angiographic evaluation was not available before surgical decompression, we cannot prove this hypothesis.

Multidisciplinary approach and collaboration are important in the management of multisystemic diseases like Behçet’s disease promoting early recognition and treatment. Individual treatments provided by the ophthalmologists, cardiovascular surgeons or dermatologists without holistic view would lead to a delay in diagnosis. Delayed diagnosis and treatments may conclude with unexpected permanent sequelas and even death.

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Informed Consent: Written informed consent was obtained from the patient.

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