Erectile dysfunction in Behçet’s disease without neurological involvement: two case reports

Sir, Behçet’s disease (BD) is a unique systemic vasculitis of unknown aetiology, which may affect both veins and arteries of different sizes and localization [1]. Erectile dysfunction, which may be described as the lack of penile erection for successful vaginal penetration, may be due to vascular, neurological, (rarely) endocrinological or psychological factors or various drugs. Recently, erectile dysfunction in BD patients with neurological involvement was reported to have a high prevalence [2]. However, there is no knowledge about erectile dysfunction in BD without neurological involvement. We present two cases of Behçet’s disease with erectile dysfunction but without neurological findings. These two cases are remarkable for having venous leak as the aetiology of erectile dysfunction, one patient being treated successfully by deep dorsal vein ligation.

Case 1 was a 35-yr-old man with a 17-yr history of BD. The main clinical features were recurrent oral and genital ulcerations, erythema nodosum, deep vein thrombosis, subcutaneous thrombophlebitis and intermittent oligoarthritis. There was no ocular or neurological involvement. The pathergy test was positive. He had been receiving colchicine treatment irregularly for the last 12 yr. He described loss of penile erection for the past year. Libido was normal. He was a non-smoker.

Case 2 was a 55-yr-old man with a 23-yr history of BD. The main clinical features were recurrent oral and genital ulcerations, erythema nodosum, deep vein thrombosis and intermittent oligoarthritis. There was no
ocular or neurological involvement. The pathergy test was positive. He had been receiving colchicine treatment regularly (1 mg/day) for the last 4 yr. He described loss of penile erection for the last 2 yr. Libido was normal. He was a non-smoker.

These two patients, both being followed by Ege University Rheumatology Department, were investigated because of their complaints about erectile dysfunction during routine tests. The patients did not have a history of any chronic infection, diabetes mellitus or any other co-existing disease. Both of the patients were normotensive with normal cardiac and pulmonary examinations. There was no organomegaly or lymphadenopathy. Urological physical examinations were normal, except for scrotal scars. The psychiatric and neurological consultations were also normal. Biochemical examinations, including liver and kidney function tests, serum glucose levels, full blood count, urine analysis, serum protein levels and lipid profile, were within normal limits in both patients. Endocrinological tests, including free and total testosterone, oestadiol, gonadotrophins, prolactin, adrenocorticotrophic hormone and cortisol, were also normal. Electromyography was performed in both patients, but polynuropathy was not detected.

As the next step, penile electrophysiological tests, penile colour Doppler ultrasound and cavernosography were performed. Electrophysiological tests included analysis of the bulbocavernous reflex. Colour Doppler ultrasonography was performed after intracavernosal injection of 60 mg papaverine as a vasoactive agent. In accordance with the literature, arteriogenic impotence is diagnosed when maximum systolic velocities measured in both of the cavernosal arteries are less than 35 cm/s [3]. End-diastolic velocities in cavernosal arteries greater than 7 cm/s with normal maximum systolic velocities are accepted as venous leak [4]. Patients with Doppler findings suggestive of venous leak undergo confirmative cavernosography. During cavernosography, 40 mg papaverine is injected intracavernously. Penile tume-scence and rigidity are observed. A 21-gauge intravenous canula is then inserted and 50% diluted contrast medium with 76% iodine is injected into the cavernous body. Serial fluoroscopic images are then taken. These last two tests (penile colour Doppler ultrasonography and cavernosography) revealed venous leak in both patients (Table 1).

Table 1. Results of investigations

<table>
<thead>
<tr>
<th></th>
<th>Case 1</th>
<th>Case 2</th>
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<tbody>
<tr>
<td>Erythrocyte sedimentation rate (mm/h)</td>
<td>42</td>
<td>21</td>
</tr>
<tr>
<td>C-reactive protein (mg/l)</td>
<td>36</td>
<td>11</td>
</tr>
<tr>
<td>BUN</td>
<td>12</td>
<td>15</td>
</tr>
<tr>
<td>Hb (g/dl)</td>
<td>13.7</td>
<td>14.2</td>
</tr>
<tr>
<td>HLA-B51</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>EMG</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Bulbocavernous reflex</td>
<td>Venous leak</td>
<td>Venous leak</td>
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<tr>
<td>Penile colour Doppler US</td>
<td>Venous leak</td>
<td>Venous leak</td>
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<tr>
<td>Cavernosography</td>
<td>Venous leak</td>
<td>Venous leak</td>
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</table>

For treatment of erectile dysfunction, patient 1 underwent deep dorsal vein ligation. During the operation, a biopsy of the cavernous body was also performed; it revealed hypocellular, collagen-rich tissue but no signs of vasculitis. Complete recovery of penile erection was attained in the post-operative period. Patient 2 refused the penile operation.

Since BD is a chronic systemic vasculitis with many different clinical findings, erectile dysfunction may be expected to occur during its course. Besides secondary psychiatric problems, various drugs used in BD may contribute to erectile dysfunction. Neurological involvement (neuro-Behçet disease) is known to be able to cause erectile dysfunction. However, erectile dysfunction in BD, and its prevalence, have not been studied extensively. In the literature we could find only a single study, by Erdoğru et al. [2], investigating the prevalence of erectile dysfunction in BD with neurological involvement. They found erectile dysfunction in 14 out of 24 (63%) neuro-Behçet patients. With respect to the aetio-logy of erectile dysfunction, they reported a mixed type of vasculogenic impotence in seven patients, arterial insufficiency in two patients, veno-occlusive dysfunction in two patients and neurogenic impotence in one patient.

The cause of erectile dysfunction in our patients was severe venous leak, detected by penile Doppler ultrasound and cavernosography. Since venous thrombosis is a well-known clinical finding in BD, penile veins may also be affected. The occurrence of venous leak may be related to recanalized microthrombosis. During the recanalization process, the sphincter structure of the veins may be disturbed, causing venous leak.

Although colour Doppler ultrasonography is reported to have high sensitivity in diagnosing arteriogenic impotence, its performance in the diagnosis of venous leak is still debatable [3, 4]. For this reason, we prefer confirmative cavernosography in addition to Doppler results indicating venous leak.

The histological findings of the cavernous body biopsy taken during the operation on patient 1 merit further discussion. The biopsy revealed hypocellular, collagen-rich tissue with no signs of vasculitis. As colchicine has an antifibrotic effect, [5], fibrosis of the cavernous body may be regarded as interesting. On the other hand, since BD is a chronic vasculitis, one might have expected to find vasculitic signs in this biopsy.

While evaluating the aetiopathogenic factors of erectile dysfunction in BD, one should keep in mind the possible effects of the drugs used in BD. Colchicine, which was used in our patients, is the most widely used drug in BD, and myoneuropathic effect is well known [6]. On the other hand, neither of our patients had used other drugs, such as cyclophosphamide, sulphasalazine, indomethacin and naproxen, which are well known to cause erectile dysfunction [7].

We conclude that erectile dysfunction may be expected even in the absence of neurological involvement.
Venous leak, as in our patients, should be kept in mind as a treatable cause of erectile dysfunction in BD.


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