Review

Bladder Involvement in Behcet's Disease

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Purpose: To review different aspects of the bladder involvement in Behcet's disease as a rare complication.

Materials and Methods: We searched PubMed, Ovid, and Google Scholar for Behcet's and neuro-Behcet's disease and neurogenic and neuropathic bladder, bladder involvement, voiding dysfunction, and urologic manifestations. Fourteen full-texts and one abstract were retrieved.

Results: Most involved patients are young to middle-aged men. Both bladder filling and emptying problems can be seen, with the storage symptoms being the most common finding. Sphincter function could be normal, dyssynergic, or deficient. The most common urodynamic finding is detrusor overactivity. In cystoscopic examination, ulcers or nodules due to vasculitis can be seen, which along with neurologic causes give rise to the voiding symptoms. The rate of cancers does not increase in Behcet's disease. Surgery and chemotherapy are tolerated well. However, radiotherapy may be associated with increased complication rates.

Conclusion: Treatment plan should be tailored according to the specific type of the bladder involvement. Periodic re-evaluation is required because of the changing nature of the bladder behavior.

Keywords: Behcet syndrome, urinary bladder, complications, neurogenic, etiology

INTRODUCTION

Behcet's disease (BD) is a systemic vasculitis with an unknown etiology, which affects the small and large vessels in both arterial and venous systems.[1] The hallmark of this disease is recurrent oral aphtae[2] that when accompanies with at least two of the followings, BD is suggested: recurrent genital ulcer, skin lesions, eye lesions, and a positive pathergy test.[3] In the International Criteria for Behcet's Disease (ICBD), vascular manifestations have also been added to increase the sensitivity of the diagnosis.[4] This disease may involve cardiovascular, central and peripheral nervous, musculoskeletal, respiratory, and gastrointestinal systems as well.[5]

Turkey and other countries along the ancient Silk Road have the highest prevalence of BD.[6] In these countries, males and females are affected almost equally, usually in their 3rd to 5th decades of life.[7]

Aside from genital ulcers, other urological problems are encountered infrequently, usually in the form of epididymitis and sterile urethritis.[8] Even rarer is the bladder involvement that has been addressed in a few case reports and case series. In this article, we aim to review different aspects of the bladder involvement in BD.

MATERIALS AND METHODS

In September 2011, we performed a detailed internet search on PubMed, Ovid, and Google Scholar, looking for articles relating Behcet's and neuro-Behcet's disease and neurogenic or neuropathic bladder, bladder involvement, voiding dysfunction, and urologic manifestations. Checking the search results and their references, we retrieved 14 full-text articles and one abstract. One of the articles was in Japanese, which was translated. Most of the articles were case reports or small case series that underscores the rarity of the bladder involvement in BD.

RESULTS

Most involved patients are young to middle-aged men.[2,9-17] The most common symptoms are storage symptoms,[2,9-11,13,14,16] and sometimes urge incontinence.[9,10,12-14] However, emptying symptoms[2,11,12,14,16] and urinary retention[11,16] may also occur.

Sphincter function can be normal. However, detrusor sphincter dyssynergia[9,12,16] or sphincter deficiency[2] may also be seen. Meatal ulceration,[14] sterile urethritis,[8] or cystitis[18] can give rise to dysuria. Gross hematuria is very rare.[2,15]

Bladder cancer has been reported either sporadically[19] or after cyclophosphamide therapy.[20] Other rare sequelae of BD are urethrovaginal and vesicovaginal fistulas.[14]

As in other causes of neuropathic bladder dysfunction, hydroureteronephrosis could ensue in BD as a consequence of increased intravesical pressure or severe bladder wall trabeculation.[2]

When urodynamic study is performed, following findings may be observed: detrusor overactivity either alone[2,9,12-14,16] or with impaired contractility,[2,11,16] decreased bladder compliance[2,14] or capacity,[9,10] bladder hypersensitivity,[9,10,12] hypo or acontractility alone,[11,14,16] and increased post-void residual urine,[9,12] with the detrusor overactivity being the most common.

In cystoscopic examination of the bladder, self-healing ulcers[2,14] or mass lesions that may resemble the bladder tumor have been reported. The pathologic examination of the mass shows vascular thrombosis, perivascular inflammatory exudates, and severe mucosal ulceration in one report[15] while in the other, it was reported as an indurated hypervascular lesion.[2]

DISCUSSION

Neurologic involvement (neuro-Behcet's disease) has been reported in 5% to 10% of patients with BD,[7] of whom 5% manifest voiding symptoms.[17] Cetinel and colleagues estimated the incidence

of the bladder involvement to be 0.07%.[2] Central nervous system involvement in BD is progressive in 85% of the patients and has an intermittent nature in the remaining 15%. It usually affects the brain stem and may resemble central nervous system infection, stroke, or multiple sclerosis.[9-11] Therefore, one reason for high prevalence of frequency and urgency in BD patients with lower urinary tract symptoms could be involvement of the pontine micturition center by a vasculitis process.[10]

Direct involvement of the bladder, however, is possible in the form of ulceration and nodules[2,14,15] or recurrent cystitis.[18] Storage symptoms in these patients, as a result, could be due to neuro-Behcet's disease, direct bladder wall involvement, or their combination.[14]

In a study, Cetinel and associates compared a group of male patients with BD with a control group and found that irritative voiding symptoms, but not obstructive ones, were more prevalent in BD group.[14]

One should keep in mind that the pattern of the bladder involvement can change during time. Porru and coworkers reported a case whose bladder's behavior changed from areflexia to instability with impaired contractility.[11]

Neurologic symptoms usually become manifested five to six years after the beginning of non-neurologic manifestations. Nevertheless, they can occur concurrently or even prior to other symptoms.[21] In the Cetinel's study, the bladder involvement became evident 1 to 10 years after the onset of neuro-Behcet's disease. However, in one patient, it preceded neurological involvement. Urodynamic abnormalities can be present even in patients without neurologic involvement.[2]

Treatment of the bladder dysfunction is individualized for each patient according to their urodynamic and imaging findings. Emptying failure is usually managed by clean intermittent catheterization while storage problems have been treated by anticholinergics; however, sometimes augmentation cystoplasty becomes mandatory.

Patients with BD have hyperreactivity of the skin and other tissues to minor trauma (pathergy). Despite this hyperreactivity, cystoscopy, bladder biopsy, and even ileocystoplasty or radical cystoprostatectomy have been done in them without complication.[2,19]

Association of BD and malignancies is rare.[22] Some believe that the incidence of malignancies does not increase in BD.[19] Increased toxicity of chemotherapy has not been reported in BD. However, radiotherapy can have such late adverse effects as ureteral stricture or skin break-down.[22,23]

CONCLUSION

Bladder involvement in BD can be neuropathic and/or due to vasculitis in the bladder wall. Different types of voiding dysfunction due to sphincter and bladder dysfunction, either in filling or emptying phases can occur.

CONFLICT OF INTEREST

None declared.

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