CASE REPORT

Acquired urethral meatal stenosis: a rare sequel of an aggressive form of Behçet's disease

Soumik Ghosh, Manish Kumar, Pushpa Kumari, Adesh Kumar Gadpayle

Department of Medicine,
PGIMER & Dr Ram Manohar
Lohia Hospital, New Delhi,
India

Correspondence to
Dr Soumik Ghosh,
soumik13joy@gmail.com

SUMMARY

Behçet's disease (BD) is characterised by the triad of recurrent oral and genital aphthous ulceration, skin lesions and ocular inflammation. This disease frequently affects other systems including neurological, locomotor, cardiac or vascular systems. Recurrent oral and genital aphthous ulcerations are the hallmarks of BD. We hereby report a case of rapidly evolving and aggressive form of BD in a young man presenting with extensive ulcerative lesions in orogenital region leading to obstructive uropathy resulting in acquired urethral meatal stenosis due to inflammatory activity.

BACKGROUND

- Behçet's disease (BD) has a long natural history and not all symptoms may surface simultaneously. The present case, with a short evolutionary period presented with frank and typical manifestation, reiterating the aggressiveness of the disease.
- Genital ulcers, although an important component of BD, but balanitis and urethritis is quite uncommon.
- Acute retention of urine as a result of cicatricial healing of glanular and urethral ulcers in a case of BD is completely an unknown entity till date.

CASE PRESENTATION

A 16-year-old man presented with complaints of redness of both eyes along with mucopurulent discharge for 5 days. It was associated with multiple painful oral ulcers over the lips, buccal mucosa and lateral surface of tongue which restricted him from opening the mouth and food intake. Ulcers were also present on the genitalia involving the prepuce, glans penis and the penoscrotal region. They were painful with burning micturition, progressing for the same duration. The patient complained of having recurrent oral ulcerations of 3–4 episodes during the last 1 year with ocular inflammation but no genital involvement as yet. There was also a history of single episode of diffuse arthralgia with swelling of large joints 3 months back for a week's duration which subsided on its own. He denied any history of fever, headache, diarrhoea, drug intake, alopecia, immunisation or recent travel. No history of tuberculosis, diabetes or similar complaints in the family was found. No history of penile circumcision in the recent past was given.

After examination the patient was found to be afebrile and conscious with stable vitals. He had intense conjunctival injection with mucopurulent discharge from both eyes. Further ocular examination

Figure 1 Extensive coalescent aphthous oral ulcers with occasional haemorrhagic foci at ulcer base (topical ointment applied).

revealed bilateral bulbar conjunctivitis, scleritis (figures 1 and 2), anterior uveitis and a visual acuity of 6/12. There was extensive oral ulceration, few aphthous in buccal mucosa and coalescent raw beefy ulcers along mucocutaneous junction of lips and angle of mouth which restricted its opening. Oropharynx and palate were however spared. There were discrete, erythematous, pustular targetoid skin lesions over the extremities. Genital examination showed tender ulcers involving the glans and inner prepuce surface with pustule formation, external urethral meatus and penoscrotal area with crusting (figures 3 and 4). There was no lymphadenopathy and rest of the physical examination was within normal limits.

Figure 2 Bulbar conjunctivitis and scleritis.

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Figure 3 Penile ulcers in the glans with haemorrhagic necrosis and encrustation in external urethral meatus.

INVESTIGATIONS

Investigations revealed erythrocyte sedimentation rate (ESR)–42 mm during the first hour, haemoglobin 13.9, total leucocyte count (TLC) 9900, platelets 2.4 l/ μ l. Kidney function tests and liver function tests were within normal limits. Urinalysis showed 2–4 pus cells/hpf with no trace of albumin. Blood and urine cultures were sterile. Chest x-ray was normal. Rheumatoid factor and anti-nuclear antibody were negative but C-reactive protein 32 mg/l. HIV 1 and 2 were non-reactive. Serum HLA B 51 disease association was positive. Two-dimensional echo revealed mild tricuspid regurgitation with mild pulmonary artery hypertension. Ultrasound abdomen showed normal prostate size and echogenicity. Dermal pathergy test with a 26-gauge hypodermic needle was performed which gave a positive 2 mm erythematous reaction after 48 h (figure 5). Skin biopsy reported features of

Figure 5 Pathergy test done with a 26-gauge needle prick evaluated after 48 h duration.

hyperkeratosis and parakeratosis of epidermis with upper dermis showing dense perivascular chronic inflammatory infiltrates invading the vascular wall with endothelial cell swelling suggestive of lymphocytic vasculitis (figures 6 and 7).

TREATMENT

Based on the clinical examination and ancillary investigation reports, a diagnosis of BD was made, as the case fulfilled the diagnostic criteria proposed by International Study Group for BD. He was started with intravenous methylprednisolone and oral colchicine. Oral ulcers regressed significantly but the patient developed acute urinary obstruction, which was

Figure 4 Discrete targetoid skin lesion with secondary pustule formation.

Figure 6 Low-power histopathology section of skin from lesional area showing prominent perivascular inflammatory infiltrates.

Figure 7 High-power microscopy of a single vessel showing lymphocytic vasculitis and intravascular thrombosis.

attributed to cicatricial healing of urethral ulcerative inflammation. Orificial adhesiolysis was performed and obstruction could only be relieved by Foley's catheterisation. Indwelling catheter was kept for 7 days and then removed thereafter. The patient consequently had no urinary symptoms after Foley's removal and his uroflowmetry was normal. One week later the patient underwent a synchronous retrograde and micturating urethrography which reported a residual partial stricture extending approximately 1 cm proximal to external urethral meatus. However for his persistent uveitis, cyclophosphamide was added and initiation of infliximab is being contemplated.

DISCUSSION

BD is a rare, chronic, multisystemic vasculitis¹ first described in 1937 by Hulusi Behçet, a Turkish dermatologist, that affects mainly young adults in Mediterranean, Middle Eastern and Far Eastern countries² a link to the ancient Silk route. Turkey has the highest prevalence of BD, with 420 cases per 100 000 population. The prevalence in Japan, Korea, China, Iran and Saudi Arabia ranges from 13.5 to 22 cases per 100 000 population. The prevalence in North America and Europe is much less, with one case per 15 000–500 000 population.³

Genital ulcers are a common symptom of BD, occurring in an estimated 70–95% of cases. In men, the ulcers usually appear on the scrotum and in women they usually appear on the cervix, vulva or vagina.⁴ However, genital ulcers can appear anywhere in the groin area, but ulcers affecting the glans penis and the external urethral meatus is a rare entity. The genital ulcers are usually painful and leave scarring in around half of all cases. Men may also experience inflammation (swelling) of the testicles and women may find that the ulcers make intercourse painful. Genital ulcers that are caused by BD are not contagious and cannot be spread through sexual intercourse.

Both genetic and environmental factors play a role in the pathogenesis of BD. The age of onset of uveitis is usually in the third to fourth decades of life, with men being more commonly affected than women. However, the uveitis of BD is believed to be most severe in young men between 15 and 25 years of age.⁵ A recent study analysing polymorphic microsatellite markers near the HLA-B gene in Japanese, Greek and Italian patients with BD strongly suggests that it is the HLA-B51 gene itself that is related to disease pathogenesis and not other genes located in the vicinity of HLA-B.⁶ The manner in which HLA-B51 relates to disease susceptibility is unknown.

The diagnostic criterion was proposed in 1990 by the International Study Group for BD.⁷ Using this, the diagnosis of BD requires the presence of recurrent oral ulceration, which have recurred at least three times over a 12-month period plus at least two other findings among the following: recurrent genital ulceration, uveitis, skin lesions and positive pathergy test result.

This patient suffered from an episode of acute obstructive uropathy due to inflammatory meatal adhesion as a result of cicatricial healing of urethral and glanular ulcers, a complication totally unprecedented in medical literature. In paediatric and adolescent age group, urethral meatal stenosis is generally encountered after urological procedures or post-circumcision.⁸ Synchronous retrograde and micturating urethrography is a novel, affordable and easily reproducible investigation to clearly visualise both anterior and posterior urethra and any length of stricture, which will enable one to make a judicious treatment plan.⁹

To conclude, although genital ulcers are quite frequent in BD, meatal stenosis as a complication of healing urethral ulcers has not been reported earlier in medical literature.

Learning points

- ▶ Behçet's disease (BD) is a rare chronic inflammatory disease with recurrent ulcerations due to underlying vasculitis which is diagnosed solely on clinical grounds. Thus, a high index of suspicion should be kept while dealing with oral or genital ulcers for an internist.
- ▶ Acute flares of BD has high inflammatory activity and propensity for cicatrization during healing stage. As it may involve delicate mucosal surfaces, care should be taken to prevent adhesion formation.
- ▶ Appropriate anti-inflammatory drugs should be prescribed in adequate dosages to prevent further flares or complications of vasculitis in the form of disabling ulcers and vascular aneurysms, thus to provide a better quality of life.

Contributors SG and PK were involved in writeup of the case. MK was involved in diagnosed the case. AKG was involved in management and follow-up of the patient.

Competing interests None.

Patient consent Obtained.

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