Balanitis xerotica obliterans

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Balanitis xerotica obliterans (BXO) / lichen sclerosus of the male genitalia is a common cause of acquired phimosis, and was first described by Stuhmer in 1928 [1]. It is described in medical literature as a chronic inflammatory condition of unknown aetiology that affects the foreskin, glans and the external urethral meatus and urethra in severe cases.

Most patients present with phimosis with varying degrees of difficulty or inability to retract the foreskin [2]. The phimosis is usually progressive in nature with the presence of initially a white sclerotic ring in the distal prepuce (Figure 1) that becomes more apparent on retraction of the foreskin and at later stages complete inability to retract with fissuring and haemorrhagic blistering during sexual activity [2,3]. Obstructive symptoms, dysuria, poor urinary stream and urinary retention should also raise the suspicion of possible involvement of the external urethral meatus [2,4]. In aggressive cases, the surface of the glans and inner prepuce may ulcerate and fuse, therefore making circumcision technically difficult [3].

BXO can present at any decade with studies reporting an age range between two and ninety years. However, the highest incidence is reported in the third decade [5]. It is a common finding in urological practice, with the majority of cases being referred for consideration of circumcision due to the presence of phimosis.

Histological characteristics

Microscopically, marked fibrosis, epidermal atrophy, interface dermatitis, and dermal oedema are observed [6,7]. It is characterised by the presence of hyperkeratosis with an atrophic epidermis and with thinning of the rete pegs [6,7]. Dermal oedema with an increase in the number and size of the basal layer cells

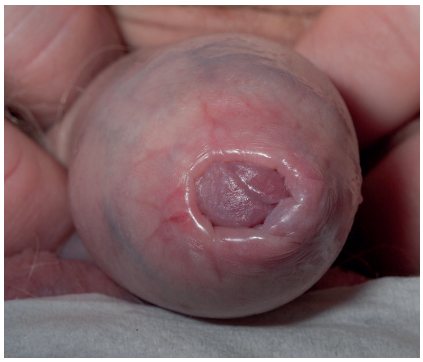

Figure 1

may also be present [6,7]. The papillary and reticular dermis present with a 'washed out' appearance with the dermal collagen forming a homogenous band at the dermal-epithelial junction in conjunction with elastin fibres, to produce an amorphous hybrid substance with infiltration of T lymphocytes, and less frequently B lymphocytes [6,7].

Aetiology

The aetiology of BXO is unknown. It demonstrates the Koebner phenomenon and arises from areas that have undergone trauma, old scars, skin graft and most importantly at sites prone to constant friction or after burns or radiation treatment [8].

Several studies have suggested that the warm, moist, urine-exposed environment that exists under the foreskin contributes to the pathogenesis of BXO. This explains why circumcision arrests the progression of the disease, whereas in cases where the circumcision was incomplete it can recur. Constituents in the urine leading to lichen sclerosus also explain why females present with vulvar lichen sclerosus as well as peristomal lichen sclerosus in patients with incontinent urinary diversion [3,9,10].

Studies on lichen sclerosus of the female genitalia have also suggested an autoimmune association but the evidence is sparse in BXO [3,11]. There is no reported association between BXO and human papillomavirus infection, however other organisms and viruses such as acid-fast bacilli, spirochates, Borrelia burgdorferi and Epstein Barr virus have not been ruled out [12].

BXO and penile squamous cell carcinoma

Squamous cell carcinoma of the penis has been reported in patients with longstanding BXO. It is thought that the intermittent ulceration and chronic instability of the glans may contribute in the progression to penile squamous cell carcinoma [3]. The risk of malignant change in patients with BXO has been quantified in a few studies. Depasquale et al. reported a 2% rate in a cohort of 522 patients [3]. Nasca et at. calculated a 5.8% risk with a mean range of 17 years from diagnosis to malignant transformation. However, 80% of their cohort did have concomitant HPV infections [13]. In a retrospective study from the UK, Edmonds et al. reported a 0% risk following a review of 329 cases with BXO. All patients were diagnosed and treated effectively either with ultra-potent topical steroid or circumcision [14].

Management

The decision on whether to proceed or not with conservative or surgical treatment of BXO is dependent on the severity and the extent of the disease as well as the impact on the patient's quality of life and patient's choice.

Medical treatment

Topical steroids seem to offer a reliable option only in the management of mild BXO limited to the prepuce with minimal scar formation. Once established scarring and phimosis are present, steroids are ineffective [7]. In the early stages of this condition however, intermittent topical steroid therapy reduces disease progression and therefore the need for surgical management [15]. Recent evidence has additionally shown that intra-urethral steroids may also assist in the management of distal urethral strictures secondary to BXO [16].

"Patients need to be educated with regards to BXO once diagnosis is confirmed and following circumcision, monitoring of the penile shaft skin and urethral meatus is required."

Surgical treatment

Surgical management is reserved for cases with progressive phimosis or where conservative management has not been successful. The underlying benefit of performing a circumcision is to remove the urine exposed warm, moist environment provided by the foreskin aiding the development of BXO.

Circumcision is recommended when BXO is restricted to the foreskin with or without the involvement of the external urethral meatus.

In the presence of a urethral stricture, surgical procedures vary depending on the location of the stricture, the length and previous treatments. Therefore, urethral dilatation, urethrotomy and urethroplasty are all recognised treatments for the management of this disease [17,18].

Follow-up

Long-term monitoring of the foreskin is required in patients managed conservatively as progressive phimosis and adhesions between the inner prepuce and the glans may lead to a less favourable cosmetic result following circumcision. Recurrence of BXO is a recognised complication especially in individuals where the circumcision was incomplete or where there is a formation of 'neo'-foreskin due to weight gain, increased pre-pubic fat and burying of the penis. Patients need to be educated with regards to BXO once diagnosis is confirmed and following circumcision, monitoring of the penile shaft skin and urethral meatus is required [19].

Good cosmetic and functional outcomes are dependent on the severity and the extent of the disease. Early diagnosis in primary care will enable timely conservative management under specialist dermatologist teams [20].

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TAKE HOME MESSAGE

- BXO is a common cause of acquired phimosis that affects the foreskin, glans and the external urethral meatus and urethra in severe and neglected cases.
- It is suspected that the warm, moist, urine-exposed environment that exists under the foreskin contributes to the pathogenesis of BXO. This also explains why females present with vulvar lichen sclerosus as well as peristomal lichen sclerosus in patients with incontinent urinary diversion
- Early diagnosis in primary care will enable timely conservative management under specialist dermatologist teams with better cosmetic and functional outcomes.

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