CASE REPORT

Wegener's granulomatosis of the penis: diagnosis and management

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A 24 year old uncircumcised man presented with a 1 month history of a painful, fungating lesion on his glans penis. Following biopsy, and further clinical developments, a diagnosis of Wegener's Granulomatosis (WG) was made. The penile lesion was treated with surgical debridement, and a penile stump was salvageable. This is only the fourth case of WG initially presenting with a penile lesion to be reported in the literature.

Key Words: Wegener's granulomatosis, vasculitis, genitourinary, penile

Introduction

Wegener's granulomatosis (WG) is a rare disease of unknown etiology. It presents with the classical triad of granulomatous vasculitis of the upper and lower respiratory tract mucosa and glomerulonephritis. In addition to this presentation, almost any organ can be affected by the vasculitic and/or granulomatous process.[1] However, genitourinary involvement is rare outside the kidney, and almost all of these cases occur in patients with previously diagnosed active disease.[2] Presented here is the unusual case of a young man whose WG first presented with a penile lesion. The diagnosis was subsequently made after follow up in hospital revealed granulomatous vasculitis of the lungs and nasal cavity.

Case report

A 24 year-old man of East Indian decent presented with a 1 month history of painful enlargement of his penis with discharge. There was no history of sexual contact, and he was unresponsive to a course of antibiotics. The patient was not on any medications, and was a lifelong non-smoker and non-drinker. His

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medical history was insignificant other than a complaint of occasional arthralgia and several months of nasal stuffiness.

Penile examination revealed a grossly swollen glans with a hard corona that was extremely sensitive to contact and a purulent urethral discharge. The edematous prepuce was confluent with the lesion and could not be retracted. It was difficult to observe the full extent of penile involvement.

The remainder of the physical examination was normal including the chest, abdomen and genitalia. There was no inguinal adenopathy. The provisional diagnosis was penile cancer and the patient was scheduled for an immediate biopsy and possible excision.

At the time of procedure 2/3 of the penile shaft and the entire glans were noted to be covered by a verrucous appearing, fungating lesion. There appeared to be invasion through Buck's fascia and there was almost no visible normal distal penis. Biopsy of a portion of the lesion revealed a granulomatous inflammatory reaction with numerous stellate abscesses Figure 1. Areas of necrosis were surrounded by palisading histiocytes with giant cells. No neoplastic changes were noted. Stains for acid fast bacilli, fungi, bacteria, elementary bodies and Donovan bodies were negative.

Five days later a debridement was carried out. Over the next few days the patient experienced hemoptysis and difficulty breathing. Chest x-ray revealed multiple parenchymal nodules and cavitating granulomatous lesions in both lungs. Examination of the nasal passages revealed numerous lesions. Biopsy of these lesions showed a granulomatous inflammatory pattern with heavy infiltrate of neutrophils, histiocytes, multinucleated giant cells, lymphocytes and plasma cells Figure 2. Necrotizing vasculitis was also noted in variously sized vessels. At this point a clinical diagnosis of WG was confirmed by a positive C-ANCA serology.

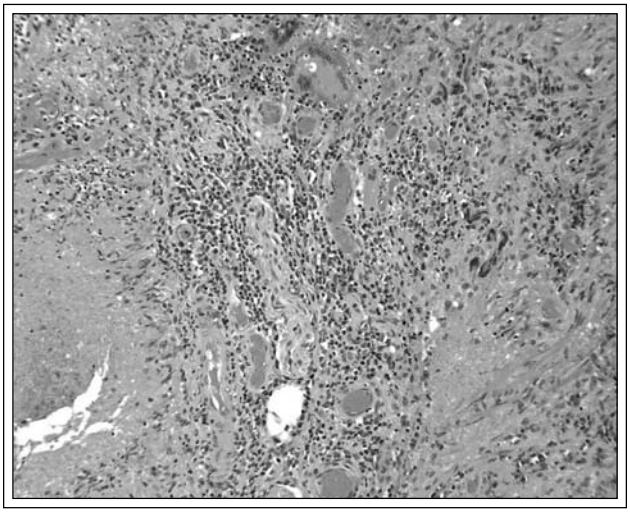

Figure 1. An H&E stained section of penis showing necrotizing granulomatous inflammation with multinucleated giant cells and histiocytes. X100.

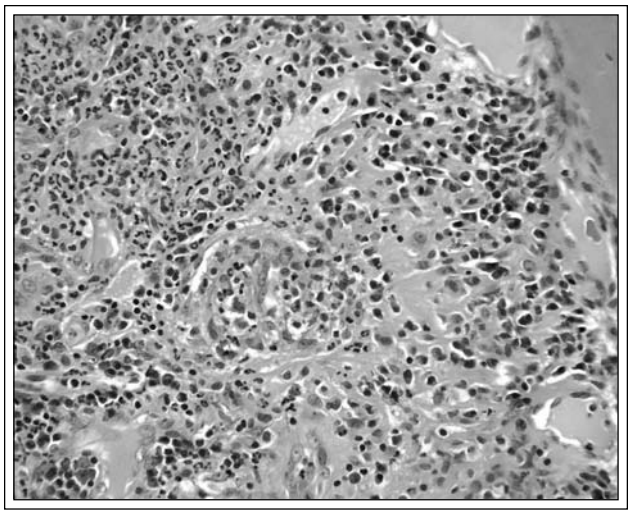

Figure 2. An H&E stained section of the nasal passage showing typical inflammatory vasculitis. X200.

The patient was started on cyclophosphamide and prednisone. The cavitary lesions in his lungs slowly resolved over one month and the patient was discharged.

The remaining penile tissue healed, leaving the patient with a penile stump. He is contemplating further reconstructive surgery.

Discussion

This case report provides several important learning points regarding genitourinary manifestations and management of WG. Wegener's granulomatosis classically affects the respiratory tract and kidney, but atypical systemic manifestations have been described.[3] Genitourinary reports include prostatic, ureteral, bladder and perianal involvement; however, involvement of the penis has only been reported eight times in the literature.[4,5] Our case is one of only four reported in the literature where penile involvement was the initial presentation of the disease. Despite this, it has been suggested that WG should be considered in the differential diagnosis when lesions histologically indistinguishable from those expected in the respiratory tract in WG are found in the penis. If this occurs, in order to start cytotoxic and/or

immunosuppressive therapy as early as possible, a diagnosis of WG should be considered and appropriate investigations carried out.[6] We therefore suggest that future patients who present with limited WG of the penis be treated medically before surgical intervention.

Huong et al report that seven of eight cases with penile involvement showed dramatic improvement with medical therapy. In two other case reports of penile involvement in WG, an early diagnosis in one case resulted in prompt immunosuppressive therapy with good effect, whereas a delayed diagnosis in the other case led to penile loss.[2]

The differential diagnosis of urogenital granulomatous inflammation includes several infectious etiologies such as *Mycobacterium tuberculosis*, non-infectious etiologies such as sarcoidosis, polyarteritis nodosa and Churg-Strauss syndrome and foreign body reactions.

Conclusion

Wegener's granulomatosis of the penis is a rare manifestation of this disease. Treatment with cyclophosphamide and steroids should precede surgical intervention, as this may lead to resolution of the lesions, and maximise recovery of viable tissue. □

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