

Trauma and reconstruction

Penile fracture in a patient with Ehlers-Danlos syndrome: A case report

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ABSTRACT

The association of penile fracture with Ehlers-Danlos syndrome (EDS) has never been described. Our patient is a 41-year-old male with EDS who presented with a traumatic penile fracture, ten days following sexual intercourse. This report recognizes the possible heightened risk of structural penile injury in patients with connective tissue disorders such as EDS and emphasizes a need for a high index of suspicion of occult urethral injury and special considerations in surgical management of these complex cases.

Introduction

Penile fracture is an uncommon urological problem, with an incidence of 1 in 175,000 in general population.¹ It occurs when rapid blunt force is applied to an erect penis and involves the rupture of the tunica albuginea layer investing the corpora cavernosa. The current standard of treatment involves immediate surgical exploration, repair of the defect, and investigation of occult urethral injury.²

Ehlers-Danlos syndrome (EDS) is a rare group of inherited connective tissue disorders, affecting roughly 1 in every 5000–10,000 people globally.³ Patients afflicted by EDS present with numerous systemic symptoms such as hyper-extensible skin, hypermobile joints, tissue fragility, vascular aneurysms, bruising, and delayed wound healing. Although EDS-related pathologic sequelae of the genitourinary tract have been described, to our knowledge, this is the first reported case of penile fracture in a patient with EDS.

Case presentation

A 41-year-old man with EDS presented to our emergency department with penile shaft pain and swelling, ten days following sexual intercourse. He delayed his visit to the emergency department, hoping for the injury to “heal itself”.

The patient reported an emergent surgical repair of his penile fracture about five months prior, followed by an uncomplicated post-operative course. His urologic history is otherwise negative.

On exam, he had ecchymosis limited to his suprapubic region and a palpable hematoma at the left base of his penile shaft. His meatus was orthotopic and testes bilaterally descended. He endorsed tenderness on palpation, which was limited to the area of the hematoma. His urinalysis

was negative, and a penile and scrotal ultrasound study demonstrated a 3.6 cm × 2.4 cm × 2.9 cm heterogeneous fluid collection adjacent to the left base of the penile shaft (Fig. 1). Based on his presentation and prior history of penile fracture, a decision was made to proceed with an operative penile exploration and repair of suspected recurrent penile fracture.

Intraoperatively, a large hematoma was observed overlying a 5 cm tear in the ventro-lateral aspect of the left proximal corporal body. Following hematoma evacuation, tunical defect was repaired in a multi-layer fashion. Flexible cystoscopy performed at conclusion of the case, did not reveal unusual findings. A Foley catheter was placed uneventfully.

On the first post-operative day, the patient's Foley catheter was removed, and following a successful voiding trial, he was discharged to home. Two weeks later, the patient reported voiding through “two separate streams”, and based on this and direct observation, he was taken to the operating room for further evaluation and surgical repair. An on-the-table pressurized retrograde urethrogram (RUG) confirmed two distinct UC fistulous tracts. These were repaired with sequential tension-free, non-overlapping layers. At time of his last follow up, his wounds were fully healed, and he was voiding without difficulty.

Discussion

We describe a novel risk factor for penile fracture, connective tissue disorder, which possibly predisposes the occurrence of corporal or urethral rupture. Despite EDS being a rare disease, urologists must be aware of the challenges that this patient population presents. This case highlights the importance of prompt diagnostic evaluation and surgical management of any patient with penile fracture, with a much greater

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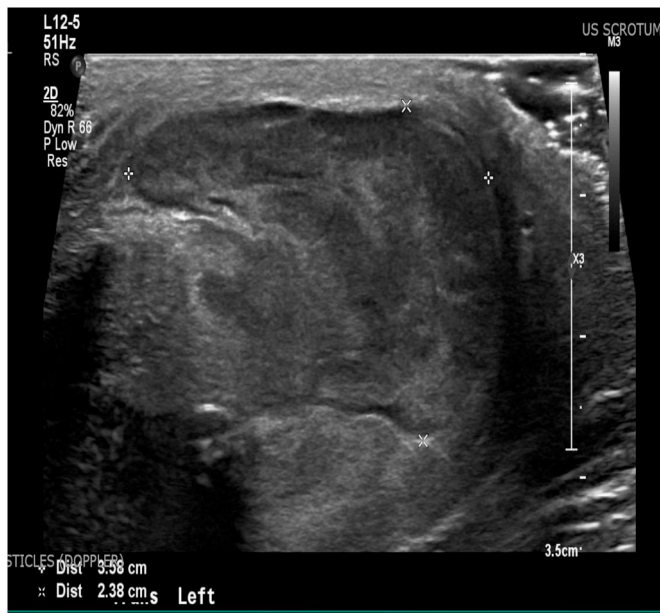


Fig. 1. Penile and scrotal ultrasound demonstrating heterogeneous fluid collection adjacent to the left base of the penile shaft.

index of suspicion in patients affected by concurrent tissue disorders. Based on this case, we propose that men with EDS, or any other impairment in connective tissue structure, may be at a much higher risk of penile fracture. We recommend a full evaluation of the urethra with a formal pre-operative retrograde urethrogram (RUG), or a pressurized intra-operative RUG, even if the presenting symptoms are not consistent with a suspicion of urethral injury.

Although patients seldom report recurrent penile fractures, our patient presented with a history of penile fracture and its repair five months prior. Ultimately, it is unclear whether the patient sustained his subsequent penile fracture due to erections weakened by the initial injury, or aberrant tissue healing caused by his EDS.

Adjustments to the surgical technique in patients with known or suspected diagnosis of EDS are recommended by many surgeons.^{4,5} In

our patient, his recurrent penile injury, re-operative surgical field, as well as the delayed presentation, were all likely contributors to his poor healing and subsequent UC fistula formation.

We acknowledge our case presentation does not include a long-term follow up evaluation, such that it is presently unknown whether he may develop subsequent ED or penile curvature.

Conclusion

We present the first case of penile fracture and its recurrence, in a patient with EDS. Although urologic involvement is either rare or under-recognized in this patient population, this complication has not been previously reported in the literature. Our report highlights special considerations needed for surgical management of these patients with a challenging condition.

Conflicts of interest

None.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.eucr.2019.101011>.

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