

Management of symptomatic adult penile urethral arteriovenous malformation

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ABSTRACT

We present the Case of a 23-year-old male with a history of recurrent spontaneous urethral bleeding due to an arteriovenous malformation (AVM) of his corpus spongiosum which abutted his penile urethra. AVMs are often congenital but can also be related to prior trauma. The literature on male genital AVMs is primarily limited to pediatric AVMs of the scrotum or glans penis with one report of adult urethral AVM in the setting of known trauma. We describe a novel presentation of atraumatic adult male genital AVM treated by surgical repair with resolution of bleeding.

Introduction

Arteriovenous malformations (AVM) are abnormal communications between high flow arterial vessels and low-pressure veins which circumvent the high resistance of capillary beds. The direct mechanism of AVM formation is unknown, yet most cases can be attributed to congenital anomalies or post-traumatic healing aberrations.¹ These malformations are identified by enlarged, tortuous, and dysplastic vessels exhibiting altered flow dynamics. They may present clinically with bleeding, thrombi, and even high-output cardiac failure.¹ In addition to their physiologic consequences, AVMs can also impart significant psychological distress to the affected patient especially in the setting of recurrent bleeding.

Due to the rarity of male genital AVMs there are no established guidelines for diagnosis and treatment, therefore, a patient-centered approach must be taken in order to address the unique challenges of each case. Diagnosis and treatment planning can be aided by radiologic modalities. Arteriograms provide high-fidelity representation of the involved vessels, doppler ultrasounds can elucidate high vs low flow malformations, and MRI can reveal adjacent tissue involvement in excellent detail.¹ Current treatments include sclerotherapy, embolization, surgical excision, cryotherapy, and laser therapy.^{2,3} Timing of therapy represents another factor that must be decided on a case-by-case basis. One school of thought recommends delaying definitive treatment of asymptomatic malformations in applicable cases until anatomical

maturity so that the structures are larger.⁴ However, earlier intervention could benefit those with particularly bulky AVMs, emotional distress, or symptomatic AVMs.

Presented is a case of penile urethra and corpus spongiosum arteriovenous malformation in an adult male with no history of genital trauma treated definitively with surgical excision.

Case presentation

A 23-year-old male presented with an eight-year history of intermittent episodes of bleeding per urethral meatus. The patient underwent numerous urethral catheters, cystoscopies and fulgurations for these episodes. His symptoms transiently resolved with endoscopic treatment, but multiple recurrences caused him severe emotional distress. He also required blood transfusions on a few of his admissions. The patient was subsequently referred to our center for further workup and exploration of definitive treatment options given his refractory disease.

The patient denied any pain or discomfort with sexual intercourse or urination. The bleeding episodes were described as spontaneous and were not associated with erections, voiding, or activity. Physical exam revealed a normal appearing circumcised penis and scrotum with mild mid-shaft urethral induration. Gross blood per meatus could be observed with gentle manual pressure on the penile shaft. An MRI revealed a 5.5cm long by 0.8cm wide AVM in the right corpus spongiosum of the proximal/midshaft penile urethra. The patient was initially referred to

Abbreviations: Arteriovenous malformation, AVM.

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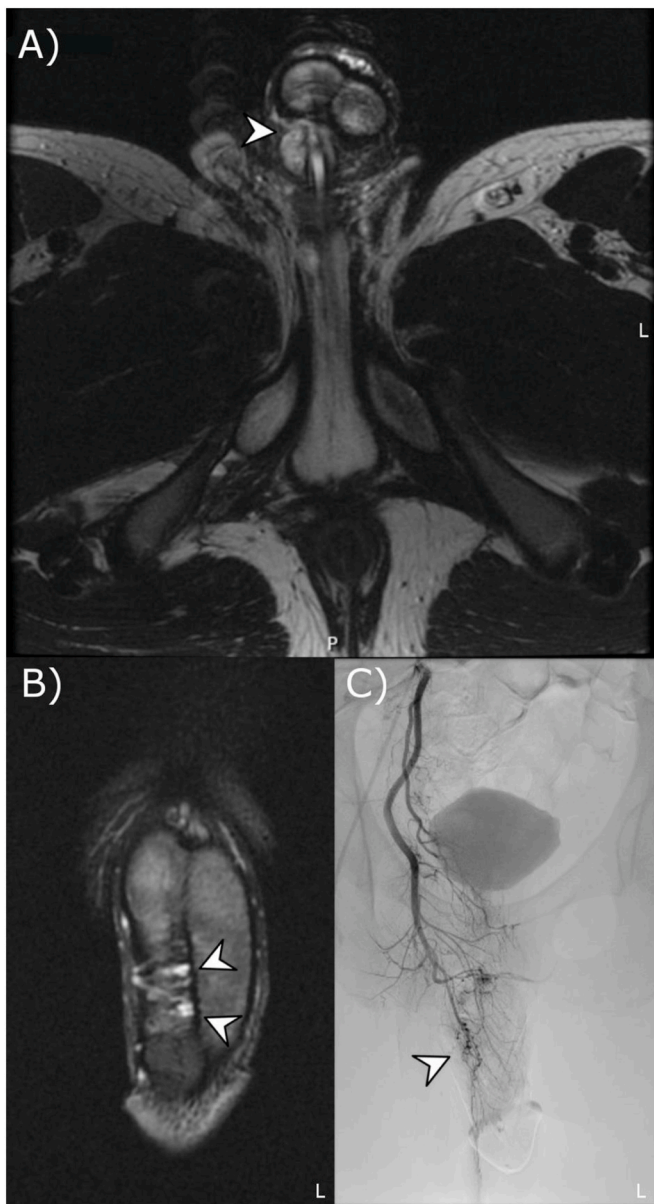


Fig. 1. A) Axial MRI of the defect showing encroachment on the penile urethra. B) Coronal MRI showing increased signal intensity adjacent the right corpus spongiosum and midshaft penile urethra. C) Angiography showing serpentine, dilated blood vessels enhancing avidly post IV contrast administration.

IR for angiogram with intention to treat, however the lesion was not adequately identifiable on angiogram or ultrasound (Fig. 1). After further counseling, surgical excision was then offered and agreed upon.

We began our surgery with cystourethroscopy which demonstrated erythematous changes in the penile urethra consistent with the AVM location on MRI (Fig. 2). A catheter was then placed, and a ventral longitudinal penile incision was made. Sharp and blunt dissection was carried to the urethra and the malformation was visualized. Doppler ultrasound confirmed venous flow through the AVM. The tunica of the corpus spongiosum was opened longitudinally at the site of the malformation which was palpable and visible due to its purple discoloration. A scalpel was used to dissect and excise the vascular malformation which extended down to the urethral lumen (Fig. 3). After incising we closed the resulting small urethrotomy and confirmed our location once again with cystoscopy. The resulting defect was then oversewn in a running fashion with a fine absorbable suture followed by closure of the corpus spongiosum, overlying Dartos fascia, and skin. Total EBL was

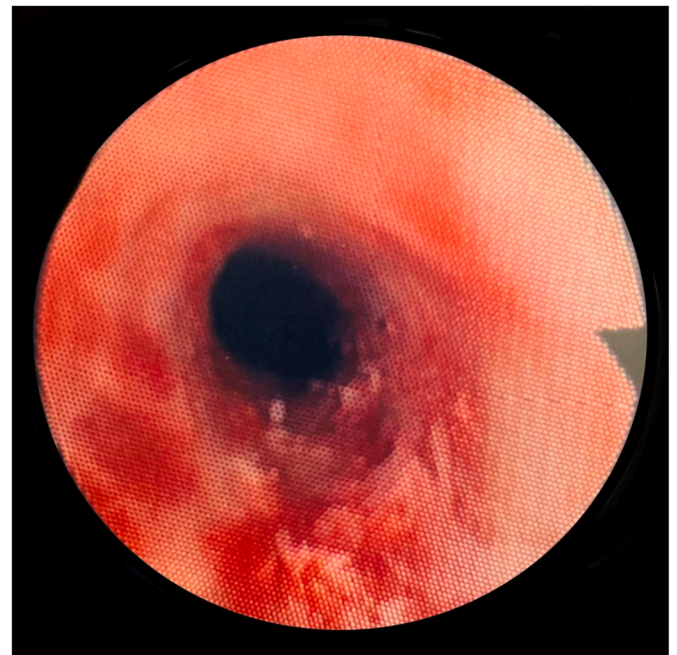


Fig. 2. Intraoperative cystourethroscopy photo showing erythematous changes of urethra.

15ml, with a total OR time of 173 minutes. Dressings were applied and the patient was discharged the same day with a foley in place for two weeks. The patient has remained asymptomatic at 3 months follow up, showing no signs of symptom recurrence and with great urinary flow.

Discussion

Male genitalia AVMs are a rare urological finding primarily confined to pediatric cases of the scrotum and glans penis.^{1,3,4} Upon review of the current literature, we believe this case represents the first described example of an idiopathic adult penile urethra and corpus spongiosum AVM. The only other reported case of an adult AVM, reported by White JT et al., describes a urethral AVM arising in the setting of past trauma.¹ Due to the rarity of male genitalia AVMs, each case presents a novel diagnostic and therapeutic challenge.

Our patient's extensive history of endoscopic biopsies and fulgurations attests to the complexity of diagnosis and treatment of these rare lesions. Initial testing with MRI aided in anatomical definition of the lesion. Subsequent arteriography is considered essential before proceeding with treatment in order to precisely evaluate feeding arteries and draining veins although in this case, angiography was not therapeutic.⁵

Several treatment options for AVMs have been utilized in other cases. Surgical excision, laser treatment, cryotherapy, embolization, and sclerotherapy have all been described.¹⁻⁵ Minimally invasive approaches, such as sclerotherapy and embolization, can be primary treatment or be implemented as adjunct therapies before surgical excision.¹⁻³ However, certain vasculature architecture and shunt types may lend to incomplete therapeutic outcomes with these modalities. In these cases, recurrence of symptoms, expansion of the AVM, and vessel collateralization can occur.² AVMs are notorious for recurrence, but surgical excision has been shown to minimize these complications and is a viable definitive therapy.⁵ The importance of complete ligation and obliteration of aberrant vasculature is essential as any remaining AVM has the potential to revascularize with surrounding tissue.^{2,4}

Our decision to proceed with surgical excision was based on the following factors: patient preference, insufficient angiographic characterization for therapy, encroachment of the penile urethra, concern for

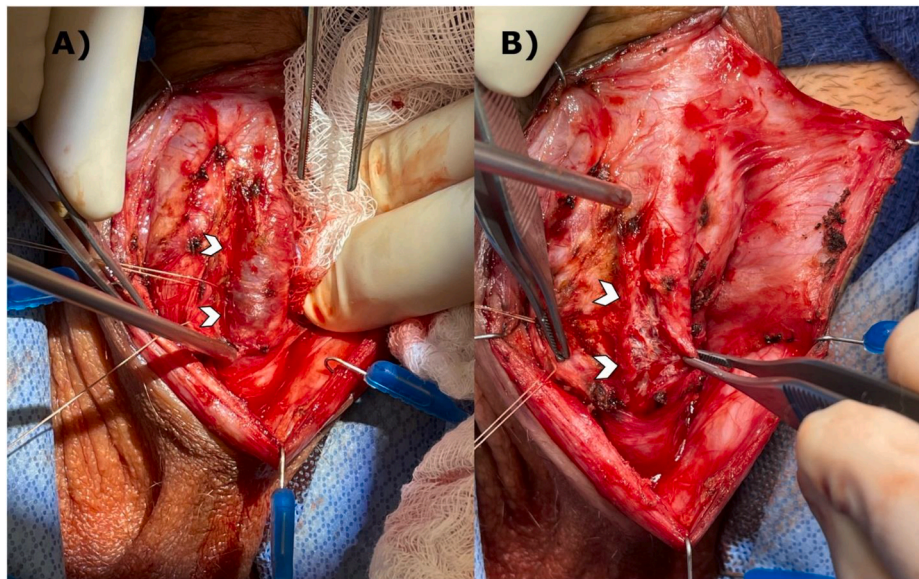


Fig. 3. A) Intraoperative photo showing corpus spongiosum with bulging and discoloration at site of AVM. B) Intraoperative photo showing incised corpus spongiosum at site of AVM.

future stricture formation, and non-diffuse localization of the lesion. The patient's post-operative course has been unremarkable and three month follow up has shown complete absence of any symptom recurrence or sequelae.

Conclusion

Surgical excision of penile urethra and corpus spongiosum arteriovenous malformations is an excellent definitive modality for managing recurrent penile urethral AVMs where embolization may not be indicated or straightforward to perform.

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