

Congenital Anomalies/Anatomical Variants

A Congenital High Flow Arteriovenous Malformation of the Bladder Presenting With Polypoid Cystitis and Ureteral Obstruction

Eric A. Addo^{a,*}, Justin Emtage^b, Kamal Massis^a, David J. Hernandez^b^a University of South Florida, 2 Tampa General Circle, STC 7028, Tampa, FL 33606, USA^b University of South Florida, 2 Tampa General Circle, STC 6, Tampa, FL 33606, USA

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ABSTRACT

A 21-year-old male with a history of Down's syndrome presented with hematuria and right flank pain. Computed Tomography (CT) of the abdomen/pelvis revealed right hydronephrosis and a right-sided pelvic vascular abnormality. Angiography revealed an arteriovenous malformation (AVM) fed by the right superior and inferior vesical arteries and nephrostogram showed a long segment of obstructed distal right ureter. Cystoscopy revealed erythema and edema of the right hemi-bladder and biopsy diagnosed polypoid cystitis. Treatment was performed by transarterial embolization with ethylene vinyl alcohol copolymer. Follow up cystoscopy and retrograde pyelography at 3 months post-procedure showed resolution of all visible pathology.

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Introduction

This report highlights the presentation of the exceedingly rare phenomenon of a congenital arteriovenous malformation (AVM) of the male pelvis involving the superior and inferior vesical arteries. We discuss the nuances associated with diagnosis as well as options regarding management. There has only been a few documented case reports of ischemic injury to the bladder secondary to AVMs with pathological specimens demonstrating pseudocarcinomatous uroepithelial hyperplasia and polypoid cystitis.^{1,2} With resolution of the AVM, associated inflammatory changes also abated with no documented risk of uroepithelial malignancy. This case report is exempt from Institutional Review Board by meeting exemption criteria by the University of South Florida IRB.

Case presentation

A 21-year-old male with past medical history of Down's syndrome presented with generalized abdominal pain and gross hematuria for 5 days. There was no previous history of nephrolithiasis or coagulopathies. Physical examination was unremarkable, but a voided urine sample did show gross hematuria. Urinalysis was nitrite and leukocyte esterase negative. Additional

laboratory values were normal except for an elevated creatinine of 1.6 mg/dL. A computed tomography (CT) scan of the abdomen/pelvis with intravenous contrast revealed bladder wall thickening, right hydroureteronephrosis, and a delayed right nephrogram with poor contrast clearance. A vascular malformation was noted in the right pelvis adjacent to the area of bladder wall thickening (Fig. 1). Subsequent angiography was performed, which identified a high flow AVM in the right inferior aspect of the true pelvis originating from hypertrophied superior and inferior vesical artery branches with a nidus along the right posterior bladder wall (Fig. 2A and 2B). A right nephrostomy tube was placed and an antegrade nephrostogram revealed a high-grade obstruction of the right distal ureter with minimal contrast passage (Fig. 3A). Cystoscopy showed severe bullous edema and erythema along the right side of the trigone, right ureteral orifice, and right lateral bladder wall. Bladder washings demonstrated groups of atypical urothelial cells. Histological section from posterior bladder wall biopsies revealed polypoid bladder mucosa with lamina propria edema, vascular congestion, and chronic inflammation consistent with polypoid cystitis.

Given the high risk for surgical resection of the high flow AVM in the bladder, embolization was deemed the best treatment option. Angiography via the right femoral artery was performed followed by super-selective catheterization of both the right superior and inferior vesical arteries and embolization with ethylene vinyl alcohol copolymer, Onyx (ev3, Irvine, CA). Immediate post-embolization angiogram showed stasis of the AVM (Fig. 2C).

* Corresponding author.

E-mail address: eaddo@health.usf.edu (E.A. Addo).

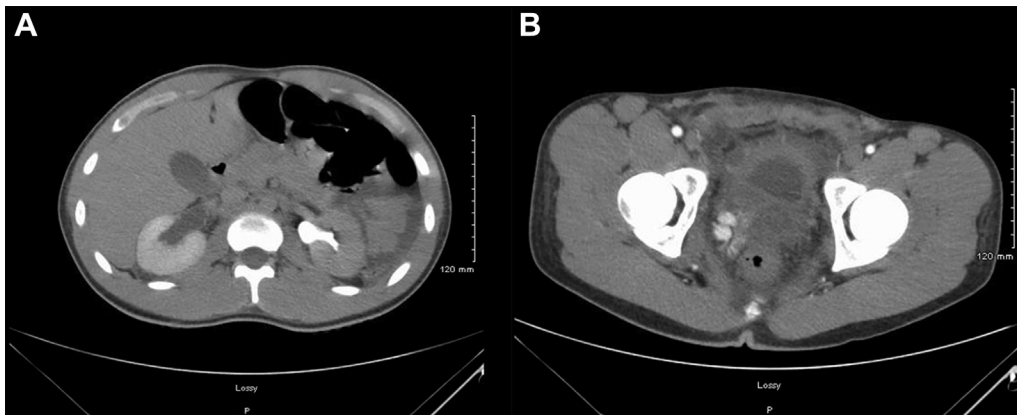


Figure 1. Intravenous contrast-enhanced Computed Tomography of the abdomen/pelvis showing: A. Excretory phase with right hydronephrosis and poor contrast clearance. B. Arterial phase with large right vesical arteriovenous malformation (AVM).

Follow up cystoscopy at 3 months post-procedure showed a significant decrease in the bladder edema and erythema. Retrograde pyelogram revealed a patent distal right ureter (Fig. 3B). The nephrostomy tube was removed after determination of distal ureter

patency. Patient was asymptomatic at time of follow up. Based on prior studies showing a risk for recurrence, we plan to perform serial cystoscopy and radiologic evaluation periodically to ensure continued resolution.³

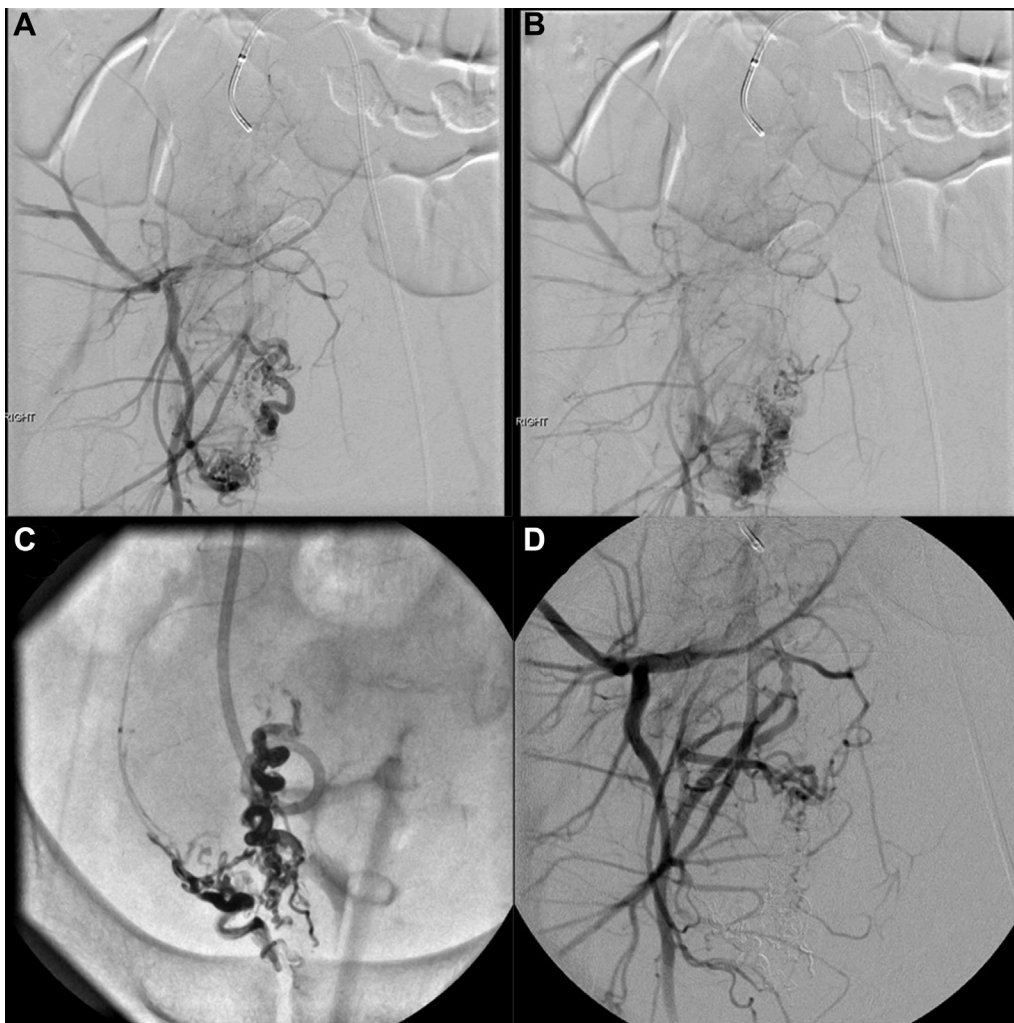


Figure 2. Angiography showing A/B Large right vesical AVM C. Embolization with ethylene vinyl alcohol copolymer D. Repeat arteriogram showing complete resolution of the AVM.

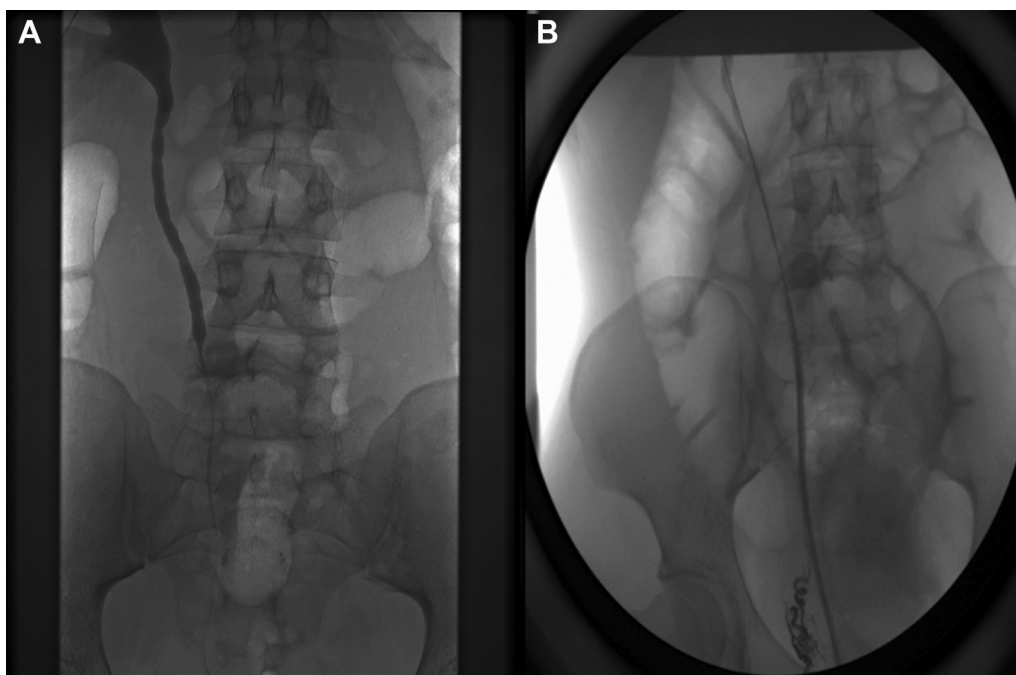


Figure 3. Fluoroscopic images showing A. Long-segment distal right ureteral obstruction on antegrade nephrostogram B. Resolution of ureteral stricture on retrograde pyelography 3 months post-embolization.

Discussion

Congenital pelvic AVMs are considered rare vascular anomalies and less than 20 cases involving the vesical arteries have been documented in the literature.^{1,2,4} Most of these cases have been reported in younger males given the association with embryologic dysfunction.⁴ These AVMs form when multiple dysplastic feeding arteries are shunted into veins without associated capillary network. The high-pressure system associated with the AVM, along with the ischemia in the specific end-organ can lead to hemorrhage. Bladder vascular malformations are located roughly 50% of the time at the base of the bladder with 25% of these lesions involving the bladder neck.¹ There has been some work by prior authors delineating the vascular congestion resulting from these AVMs as the source of conditions such as pseudocarcinomatous urothelial hyperplasia and polypoid cystitis. The previously mentioned reports did show that the associated inflammatory changes also abated with no documented risk of urothelial malignancy.^{1,2} At presentation, the patient had right ureteral obstruction that mimicked a long segment ureteral stricture. However, the obstruction was almost certainly due to edema and vascular congestion in the ureter (ureteritis) similar to seen in the bladder. Ureteroscopy was performed at 3 months after the embolization and showed no abnormalities.

Pelvic AVMs can usually be detected using pelvic ultrasound, CT, or magnetic resonance imaging. Confirmatory test is angiography, which aids in planning therapy and assessing the extent of the disease.⁴ Optimal treatment of these lesions continues to be controversial, but includes either surgical resection or embolization.^{3,5} Asymptomatic or mildly symptomatic lesions do not need treatment. Surgical treatment employs complete ligation of feeding vessels, but often collaterals develop and bypass the ligation site.⁴ Surgical intervention with transurethral resection carries an increased risk of intraoperative hemorrhage, incomplete removal of the AVM nidus, surrounding organ injury, and increased recurrence rates. Rapidly enlarging or symptomatic lesions should be embolized

preoperatively if surgical excision is deemed necessary. Endovascular therapy through percutaneous embolization has been advocated by some as the preferred treatment and also used if the lesion is deemed inoperable.³ Various embolic agents can be employed including coils, sclerosants, rapidly polymerizing acrylic adhesives, polyvinyl alcohol foam particles, and ethylene-vinyl alcohol copolymers (Onyx). Combined embolosclectomy has been utilized with strong efficacy in pelvic AVMs with the use of coils and ethanol helping in achieving complete remission. Often times, lesions begin to recanalize and multiple therapeutic endovascular interventions are required.^{3,5}

Conclusion

Congenital pelvic AVMs are rare, but a serious cause of massive hematuria. The resultant vascular congestion can cause pathologic changes in bladder mucosa such as polypoid cystitis. Long term surveillance is required to ensure no recurrence of the AVM.^{3,5}

Informed consent

Written consent was obtained by the authors from the legal guardian of the patient prior to submission.

Conflict of interest

The authors declare no conflict of interest.

Acknowledgments

None.

Supplementary data

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.eucr.2015.07.004>.

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