Urology Case Reports 6 (2016) 39–41

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Pediatrics

Profound Hematuria in a Toddler Yields an Unusual Diagnosis

Ezekiel E. Young [a,*], Nilay Gandhi [b,d], Peter Stuhldreher [a], Justin A. Bishop [c], Ming-Hsien Wang [a]

[a] Division of Pediatric Urology, The James Buchanan Brady Urological Institute, The Johns Hopkins School of Medicine, 1800 Orleans St., Suite 7308, Baltimore, MD 21287, USA
[b] The James Buchanan Brady Urological Institute, The Johns Hopkins School of Medicine, Baltimore, MD, USA
[c] Department of Pathology, The Johns Hopkins School of Medicine, 600 N. Wolfe Street/Carnegie 417, Baltimore, MD 21287, USA

ARTICLE INFO

Article history:
Received 4 March 2015
Received in revised form
11 February 2016
Accepted 12 February 2016

Keywords:
Hematuria
Lymphatic malformation
Partial cystectomy
Pediatric urology

ABSTRACT

Herein we present a rare case of profound recurrent gross hematuria in a young child with no known predisposing event. She was eventually diagnosed with a large lymphovascular malformation of the bladder. She underwent multiple unsuccessful attempts at embolization before eventual curative partial cystectomy.

Introduction

Gross hematuria in children is an uncommon condition, seen in approximately 1 of 1000 outpatient pediatrician visits.[1] The majority of these cases represent benign conditions attributed to renal disorders, urinary tract infections, nephrolithiasis, or trauma; however a significant portion will have no identifiable etiology. In addition to the common etiologies, one must also consider rare conditions such as systemic coagulopathies, bleeding diatheses, or vascular malformations.[2–4] Below we present the case of a 3-year-old female who presented with recurrent gross hematuria resulting in anemia and clot urinary retention who was ultimately diagnosed with a lymphovascular bladder malformation.

Case presentation

An 18-month-old otherwise healthy female presented to an outside hospital with painless gross hematuria with clots. She was taken for cystoscopy and bladder biopsy with pathology reportedly demonstrating an ill-defined vascular malformation. MRI demonstrated multiloculated cystic areas external to the anterior bladder wall extending toward the dome and right anterolateral bladder wall (Fig. 1A and B) suggestive of a mixed venous and lymphatic malformation. She underwent embolization with sodium tetradecyl sulfate (sotradecol) and was eventually discharged. The hematuria recurred several times and she eventually presented to our emergency department at 3 year of age and was found to be severely anemic (Hgb 4 g/dL). She was taken for cystoscopy with clot evacuation and then for selective embolization with doxycycline. MRI was again performed at that point and demonstrated a heterogeneous increased T2 signal infiltrating the bladder wall in the right lateral wall, dome, anterior and inferior aspect of the bladder (Fig. 1C and D). The radiologists felt these findings most consistent with a microcystic lymphatic malformation. The patient was discharged after a week of clear urine with stable Hgb. A week later, the patient returned with recurrent hematuria and clot retention requiring further transfusions. After ultimately failing several additional sessions of embolization the patient was taken for open exploration. Intraoperatively, the anterior bladder wall was noted to demonstrate extensive necrosis and the mucosa was noted to be erythematous. A partial cystectomy was performed uneventfully. The hematuria resolved and she was discharged on post-operative day 3. She has now been

* Corresponding author. Present address: Department of Urology, Stony Brook University School of Medicine, HSC 9-043, Stony Brook, NY 11794, USA. Tel.: +1 4015696913.
E-mail addresses: eyoung31@jhmi.edu, zekeyoung@gmail.com (E.E. Young).
[d] Present address: Potomac Urology, 4660 Kenmore Ave., Suite 1120, Alexandria, VA 22304, USA.

http://dx.doi.org/10.1016/j.eucr.2016.02.009

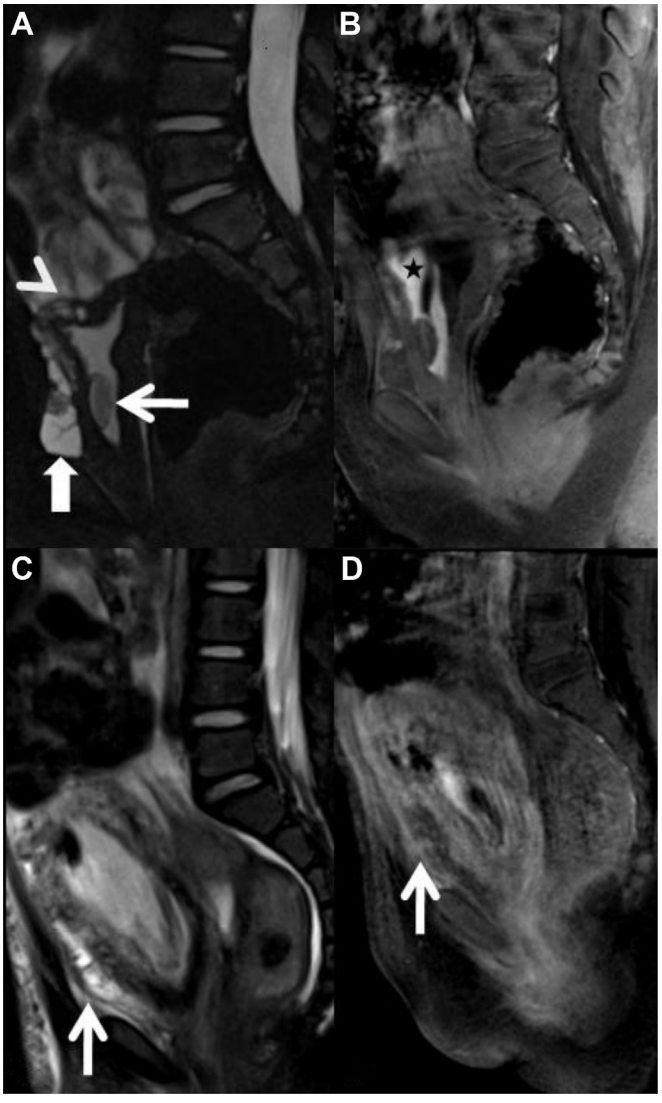

Figure 1. 3-year-old-girl with history of a bladder vascular malformation had undergone MRI of the pelvis for further characterization; (A) sagittal T2 fat-saturated image shows a lobulated predominantly hyperintense lesions adherent to the anterior bladder (block arrow), intraluminal (arrow) and intramural (arrowhead) in location; (B) sagittal T1-post contrast image shows no significant enhancement of the three components and gadolinium contrast in the bladder (black asterisk), findings consistent with a lymphatic malformation. MRI of the pelvis performed status-post partial sodium tetradecyl sulfate embolization and clinical presentation of hematuria; (C) sagittal T2 fat-saturated image shows decrease in size of extrinsic lobulated component (arrow) and stable appearance of intramural component; (D) sagittal T1-post contrast image shows no enhancement (arrow), again confirming the imaging features of a lymphatic malformation.

entirely asymptomatic with 12 months of follow-up. Her final pathology demonstrated infarcted lymphovascular proliferation (Figs. 2 and 3) Given the infarcted nature of the tissue and the subtle histologic distinctions between the various lymphovascular proliferations found in the bladder, the pathologists were unable to offer a more specific diagnosis.

Discussion

Lymphovascular malformations of the bladder are extremely rare, and have only been reported in a handful of patients in the medical literature.[5–10] While four of these previous six patients were children, this case represents the youngest known child to

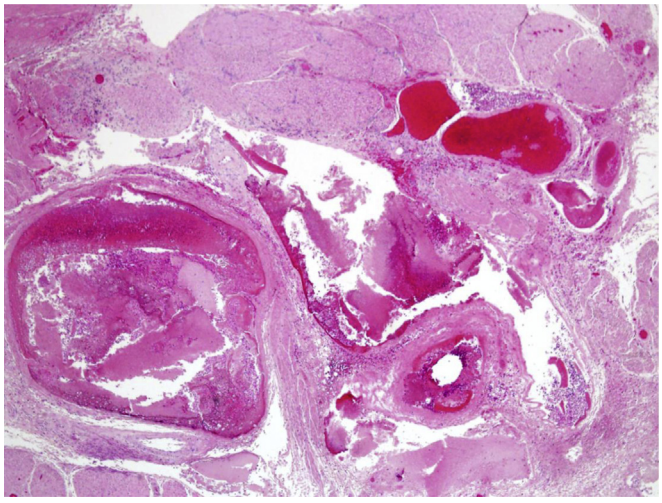

Figure 2. The lesion consisted of a collection of dilated vessels of varying sizes, randomly distributed within the detrusor muscle.

have been affected. Interestingly, the current patient also appears to have suffered from one of the gravest cases thus far.

Initial treatment efforts, utilizing embolization with sodium tetradecyl sulfate, aimed to maximally preserve bladder and to avoid open surgery, but did not have lasting effect. The interventional radiologists at our institution believed that doxycycline would be a more effective sclerosing agent. Accordingly the lesion was embolized with doxycycline over the course of three separate sessions, each time with fleeting, if any, improvement in the degree of hematuria. Although there is no literature specifically reporting on the embolization of lymphovascular malformations of the bladder, it may be possible to extrapolate from other anatomic locations. The vast majority of lymphatic malformations occur in the neck and axillary regions and can be problematic cosmetically and from their mass effect, but do not generally bleed. While the goal of treatment of such lesions is to decrease their size, rather than to stop bleeding, it has been well documented that macrocystic lesions respond much better to percutaneous approaches than microcystic ones. Rozman et al, for example, reported minimal reduction in the size of microcystic lesions, with all such patients instead requiring eventual

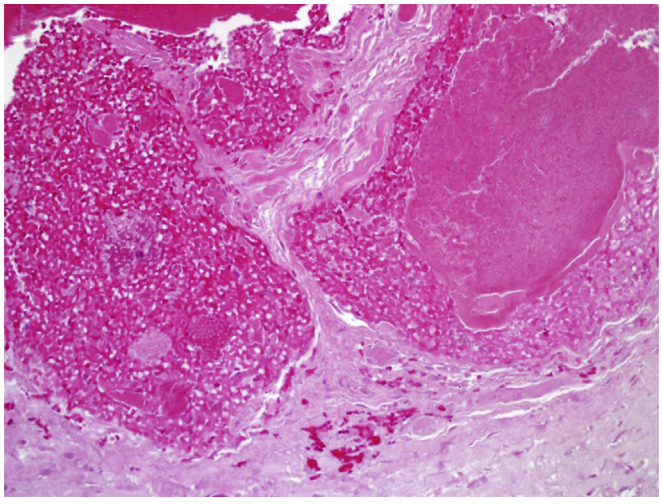

Figure 3. On high power, the lining endothelial cells were brightly eosinophilic with no residual nuclei, consistent with infarction.

surgery.[11] This makes sense as the larger cysts can be accessed and individually sclerosed using a needle whereas the microcysts are inherently too small to be individually accessed percutaneously. Based on this reasoning we suspect our patient originally had a mixed lesion made up of both macrocystic and microcystic components. Each of the embolizations may have effectively sclerosed some of the macrocysts, causing those portions of the lesion to become necrotic, but ultimately leaving the microcystic portion viable to continue bleeding. Based on this experience we would recommend against percutaneous treatment of lymphovascular malformations of the bladder that contain any significant microcystic component, as noted on imaging. Such treatments appear relatively ineffective and may have a deleterious effect on the quality of the bladder tissue, making the eventual surgery more difficult, potentially increasing the chance of post-operative bladder leak or other complications, and also preventing the pathologists from being able to provide a definitive histologic diagnosis. Instead prompt partial cystectomy would be advised for such lesions.

Conclusion

Lymphovascular malformations of the bladder are quite rare but can cause life-threatening hematuria. Cross-sectional imaging is recommended when this diagnosis is being considered. If imaging demonstrates a significant microcystic component of the lesion then attempts at embolization are less likely to be successful and prompt open exploration with partial cystectomy is warranted.

Conflicts of interest

There is no conflict of interest.

References

1. Greenfield SP, Williot P, Kaplan D. Gross hematuria in children: a ten-year review. *Urology*. 2007;69(1):166–169. http://dx.doi.org/10.1016/j.urology.2006.10.018.
2. Patel H, Bissler J. Hematuria in children. *Pediatr Clin North Am*. 2001;48(6):1519–1537.
3. Gordon C, Stapleton FB. Hematuria in adolescents. *Adolesc Med Clin*. 2005;16(1):229–239. http://dx.doi.org/10.1016/j.admecli.2004.09.005.
4. Pan CG. Evaluation of gross hematuria. *Pediatr Clin North Am*. 2006;53(3):401–412. http://dx.doi.org/10.1016/j.pcl.2006.03.002. vi.
5. Bolkier M, Ginesin Y, Lichtig C, Levin D. Lymphangioma of bladder. *J Urol*. 1983;129(5):1049–1050.
6. Wyler SF, Bachmann A, Singer G, et al. First case of lymphangioma of the bladder in an adult. *Urol Int*. 2004;73(4):374–375. http://dx.doi.org/10.1159/000082242.
7. Niu Z Bin, Hou Y, Sun RG, et al. Cystic lymphatic malformation of bladder presenting as a pelvic mass. *J Pediatr Surg*. 2011;46(6):1284–1287. http://dx.doi.org/10.1016/j.jpedsurg.2011.03.016.
8. Niu Z Bin, Yang Y, Hou Y, et al. Lymphangioma of bladder. *Urology*. 2010;76(4):955–957.
9. Seyam R, Alzahrani HM, Alkhudair WK, et al. Robotic partial cystectomy for lymphangioma of the urinary bladder in an adult woman. *Can Urol Assoc J*. 2012;6(1):E8–E10. http://dx.doi.org/10.5489/cuaj.10103.
10. Pratap A, Tiwari A, Pandey SR, et al. Giant cavernous hemangiolymphangioma of the bladder without cutaneous hemangiomatosis causing massive hematuria in a child. *J Pediatr Urol*. 2007;3(4):326–329. http://dx.doi.org/10.1016/j.jpurol.2006.10.010.
11. Rozman Z, Thambidorai RR, Zaleha AM, et al. Lymphangioma: is intralesional bleomycin sclerotherapy effective? *Biomed Imaging Interv J*. 2011;7(3):e18. http://dx.doi.org/10.2349/biij.7.3.e18.