CASE REPORT

Primarily Closed Bladder Exstrophy in a Female Patient Complicated by a Bladder Calculus and Squamous Cell Carcinoma-A Rare Presentation

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Abstract

Bladder exstrophy is a rare congenital anomaly with an estimated incidence of 1 in 10,000 and 1 in 50,000 live births. It is even rarer in females with a male-to-female ratio being 5:1 to 6:1.We report a 19yrs old female who had primary closure of exostrophy at the age of 6months and now presented with vesical calculus and squamous cell carcinoma of bladder..

Key Words

Bladder Exstrophy, Congenital Anomaly, Squamous Cell Carcinoma

Introduction

Bladder exstrophy is a rare congenital anomaly (1), it is even rarer in females with two series reporting a 5:1 to 6:1 male-to-female ratio (2,3) . We report a case of 19years old female patient who gave history of primary exstrophy repair at age of 6months and now presented with vesical calculus and squamous cell carcinoma of bladder.

Case Report

A19years old female with primary bladder exstrophy closure at the age of 6 months presented with obstructive urinary flow and frequency of micturition. Patient was evaluated elsewhere with ultrasound abdomen that revealed bilateral mild to moderate hydroureteronephrosis with a large vesical calculus. X-ray KUB (*Fig.1*) confirmed the calculus. Her serum chemistry was within normal limits. Urine culture showed E.coli. After receiving culture specific antibiotics, she underwent open cystolithotomy. During the procedure, an induration was felt at the bladder base, which was biopsied. Biopsy revealed squamous cell carcinoma. Her metastatic work up was negative. CECT abdomen and pelvis was suggestive of localized disease. She underwent radical cystectomy; bilateral extended lymphadenectomy with an ileal conduit. The suprapubic wound closure required a faciocutaneous thigh flap that was transferred to the suprapubic region through a subcutaneous tunnel (*Fig 2*). The resultant raw area on the left upper thigh was covered using a split thickness skin graft. Postoperative period was uneventful. Final histopathology was suggestive of a localised well-differentiated squamous cell carcinoma. All lymphnodes were negative for malignancy. Patient was doing well at 24 months of follow-up.

Fig1. X-Ray of KUB Region Showing A Large Vesical Calculus

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Fig 2. Post Operative Picture Showing Suprapubic Gap Closed using Fasciocutaneous Thigh Flap

Discussion

Currently, the bladder exstrophy can be diagnosed antenatally with the use of high-resolution real-time ultrasound. However, in our country we come across these patients at an older age, as there is often a delay before they seek medical advice. Moreover, ensuring follow-up in our patients is also difficult due to ignorance and other socio-economic problems.

We report a case of exstrophy in a female patient who after the primary closure did well for 19 years but had never gone back for follow up. She had now presented with a vesical calculus and its inherent complications i.e infection, long-standing irritation, and malignancy (squamous cell carcinoma). The most common complication after reconstruction for exstrophy-epispadias complex is stone formation ranging from 26 to 18% (4,5).Silver *et al* (6) in their series of 530 patients of exstrophy-epispadias complex observed stone formation in 15% of cases. In their series most of the stones formed in the bladder and the risk of stone formation was associated with augmentation ($p<0.001$) and bladder neck procedures to increase outlet resistance ($p<0.001$).Other risk factors included urinary tract infections, foreign bodies (remnant suture, squamous epithelium), vesicoureteral reflux and urinary stasis. Rub *et.al.*(7) reported a case of a large bladder stone with right-sided hydronephrosis in a 3-year-old child who underwent exstrophy repair at the age of 7 months. No. 1 braided, polyester, non-absorbable suture used to close the pubic bones during the repair, was responsible for stone formation. However, stones following primary closure are rare.

In our case, stone formation can be hypothesized on the pretext that the initial ultrasound revealed bilateral hydroureteronephrosis with a vesical calculus. She may have had associated vesicoureteral reflux resulting in pseudo-residue causing infection and stone formation.

Although bladder exstrophy is associated with increased incidence of adenocarcinoma there are case reports of primary squamous cell carcinoma in unreconstructed exstrophic bladder (8). Our case had squamous cell carcinoma in a reconstructed bladder, this may be due to the long-standing stone, which had resulted in mucosal irritation, metaplasia, and malignancy (squamous cell carcinoma) which is a known complication in 2-10% of patients with chronic catheters or stones (9,10).

Conclusion

It is prudent to inform exstrophy patients regarding the need for a regular long-term follow-up. This has been well summarized in Robert Jeffs thoughts 'Proper surgical execution and follow-up, follow-up, follow-up-the recipe for success'. In addition, long-standing stone disease should make one suspicious of malignancy especially if overt mucosal changes are noted intraoperatively.

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