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

Although bladder extrophy 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 extrophy 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.

**References**