

## Case Report


# A Rare Case of an Adult with Untreated Bladder Exstrophy Presenting with Adenocarcinoma of Urinary Bladder

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## Abstract

Untreated bladder exstrophy in an adult is rare, as the defect is obvious and primary reconstruction is usually done in infancy. There are less than 90 reported cases of primary adenocarcinoma in an untreated bladder exstrophy in literature and only two such case reports from India. We report the case of adenocarcinoma in a 29 year old male with untreated bladder exstrophy to highlight the extreme rarity, yet distinct possibility and challenges faced in surgical management of such cases.

## Key words

Bladder exstrophy, Adenocarcinoma, Urinary bladder.

## Introduction

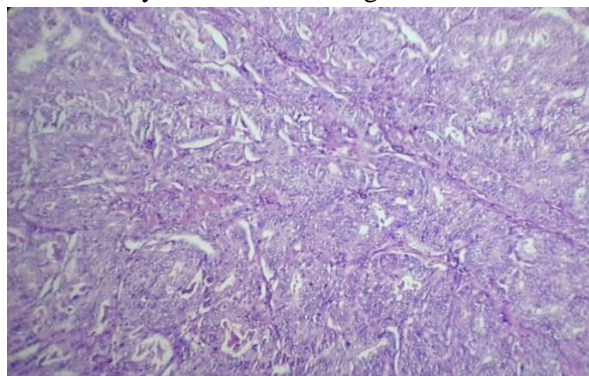
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## Case report

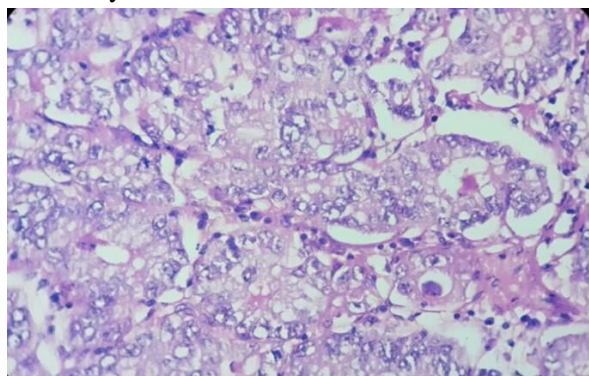
A 29 year-old male came with chief complaints of serosanguinous discharge from a growth in lower abdomen and associated right flank pain since four months. He gave history suggestive of bladder exstrophy since birth for which he had not sought any treatment. On examination, there was a 7 cm X 6 cm sized non-tender, exophytic

mass involving nearly entire exposed surface of bladder. The pubic bones were widely separated with divercation of recti. His hematological and biochemical parameters were within normal limits. Histopathological examination of the exstrophy bladder grossly shows bladder mucosal growth along with skin measuring 9cm x 8cm x 6.5cm. The bladder mucosa measures 5.5cmx3.5cm and was seen protruding outside the skin surface. Ureter was identified measuring 3.5 cm in length and on cutting lumen was patent. Microscopy was suggestive of moderately differentiated adenocarcinoma of urinary bladder which shows infiltration to the depth of 1cm (**Figure – 1** and **Figure - 2**). The skin margins, base and ureter were free from tumor invasion.

**Figure – 1:** 10X view of H&E stained section from urinary bladder mucosal growth.



**Figure – 2:** 40X view of H&E stained section show moderately differentiated adenocarcinoma of urinary bladder.



## Discussion

Bladder exstrophy is a rare disease with an incidence of 2.15 per 100,000 births [1]. With the defect being obvious, almost all patients are

treated soon after birth. Studies have found that such patients, even after reconstruction, are prone to malignancies. According to Smeulders, et al., risk of neoplasia in treated exstrophy patients is 17.5% [2]. The risk of bladder neoplasia in treated exstrophy patients is 4%, that is, 700 fold higher risk than in normal population [2].

In 1955, McIntosh and Worley recognized the risk of carcinoma in untreated exstrophy of bladder [2, 3]. Of the 40 cases reviewed by him, two-thirds were men and the average age at diagnosis was 44 years (the youngest was 21 years). A total of 82.5% cases were adenocarcinoma, 12.5% were Squamous Cell Carcinoma (SCC), 5% were of unknown type. The histology of the bladder mucosa in untreated exstrophy has been described in detail by Smeulders, et al. in 2001 [2]. The trigone is covered by transitional cell epithelium. The epithelium at the top is characterized by squamous metaplasia merging into normal skin in the middle of the bladder, the epithelium shows glandular metaplasia. Considering the range of epithelia lining the untreated exstrophy bladder, various tumors are possible in such patients. The two most common malignancies are adenocarcinoma (75-85%) and SCC (5%) [3, 4]. In contrast, only 0.5-2% of bladder cancers in normal population are adenocarcinoma [2, 4]. Histological subtypes of adenocarcinoma of bladder can be nonspecific, enteric, mucinous, signet-ring cell, or mixed [5]. McIntosh and Worley considered that exposure of the exstrophied bladder to the environment and constant infection resulted in glandular metaplasia [3], possibly to produce protective mucus, and that this was the site of malignant change. It is also suggested that adenocarcinoma in exstrophic bladder originates from the colonic epithelium covering the mucosa of the organ. However, accurate prospective studies are lacking to confirm these theories. Furthermore, rarity of such cases makes a prospective study quite difficult. Smeulders concluded that chronic irritation and infection leading to metaplastic transformation of urothelium resulting in

malignant changes is the most likely possibility [2].

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