

Squamous Cell Carcinoma Revealed by Bladder Exstrophy

F. Bakloul, A. Slaoui, N. Jakhlal, A. Elghazoui, T. Karmouni, K. El khader, A. Koutani and A. Ibnattya
Urology B, Ibn Sina Hospital Rabat, Morocco.

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ABSTRACT

Patients with bladder exstrophy have a high risk of bladder cancer, usually adenocarcinoma kind. The squamous cell carcinoma is rare, with only 12 cases reported in the literature. We report one case of squamous cell carcinoma of bladder exstrophy occurs unpaired child in a 34 years old man.

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Introduction

The bladder exstrophy is a rare congenital malformation of the urogenital tract, associated with a high incidence of adenocarcinoma. We report a case of squamous cell carcinoma of bladder exstrophy occurs unpaired child in a 34 years old man. Thus this we will discuss the characteristics of the bladder exstrophy, carcinogenesis mechanisms, treatment modalities and prognosis of these tumors.

Observation

A 34 years old man, with an unpaired bladder exstrophy was admitted for management of cauliflower burgeoning mass. (Figure 1), gradually increasing in volume last year, and becoming painful since 4 months.



Figure 1. Cauliflower burgeoning mass at the bladder plate.

Biopsy returned for a moderately differentiated squamous cell carcinoma. Before being sent to our training, here received 3 courses of 5-fluorouracil and cisplatin. The abdominopelvic CT shows bilateral ureterohydronephrosis, also a suprapubic process with lymphadenopathy in internal iliac and obturator chains. Renal function was normal. He under

went cystectomy with Wallace kind skin urethrostomy. The second operating time was the parietal repair done by plastic surgeons using a double-sided plate with flapstems or fasciae latae (Figure 2).

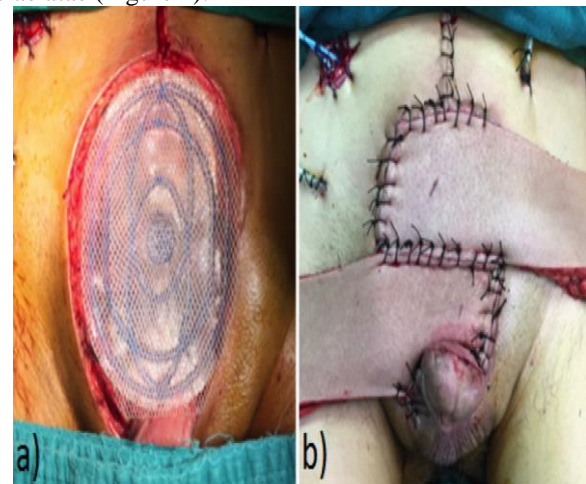


Figure 2. The parietal defect repair : a) plate b) and shreds tensor fascia lata

The study pathological revealed a moderately differentiated squamous carcinoma and in filtrating the entire wall of the bladder, prostate and hypodermis. The suites were marked by a wall of infection requiring removal of the plate.

Discussion

The bladder exstrophy is a rare malformation of the urogenital tract, with an incidence of 1 in every 10.000 to 40.000 births. It is more common in males. It is characterized by dehiscence of the abdominal wall below the umbilicus ; pubis and the rectus muscles are separated ; the anterior surface of the bladder, cervix and urethra is absent. Indeed, the lining of the posterior surface of the bladder and cervix comes to protrude between the right and pubic muscles. The malformation could also reached genitalia : for males the penis appears short , spread , curled up with an epispadias ; for girls the clitoris is bifurcated and the vaginal opening is narrow. By the way, other defects may be associated with this disease namely inguinal hernia and cryptorchidism in boys. [1]

Tele:

E-mail address: fouadbakloul1984@gmail.com

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Figure 3

The Exstrophy increases the risk of developing a bladder tumor by 694 times compared to the general population [2]. Chronic irritation and infection lead to metaplastic transformation of urothelium resulting in degeneration. This is most likely the etiopathogenesis. Adenocarcinoma is the most common type in cases of bladder exstrophy, which represents 95% of cases, while squamous cell carcinoma accounts for only 3% to 5% of cases [3, 4]. To date, only 13 cases of squamous cell carcinoma of bladder exstrophy have been reported in the literature, including the present case [5, 6, 7, 8, 9]. Five of them suffered in their childhood repair of bladder exstrophy: cystectomy with diversion or enterocystoplasty. Coffey kind of increase were carried out for each 2 patients [7, 8, 10, 11]. The fifth patient had no documents about the procedure made [9]. The age at diagnosis of squamous cell carcinoma is 52.6 years (range 34-78), with a sex ratio of 3 men and women: 1. The average duration of symptoms was 18.6 months (range 4-60). The appearance of a new growth on the exposed mucosa is the most frequently reported symptom (92.3%). Two patients presented inguinal lymphadenopathy (15.3%), but none of the patients had systemic metastases. In our patient, the scanner has objectified internal iliac lymph nodes and obturator.

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Complete tumor resection is the only therapeutic option. The chemotherapy's and radiotherapy's role in this disease can not be assessed due to the small number of patients. It is important to be aware that the simple cystectomy may not prevent recurrence, especially among men. In a group of 65 patients (45 men, 16 women) with bladder exstrophy,

Smeulders and Woodhouse found the occurrence of bladder cancer on tissue remnants in four male patients after a median follow up of 34 years (range 28-48). The authors concluded that the male anatomy would not allow a complete cystectomy as in women and/or genital tract secretions maculins could be carcinogenic. [2]

In fact, the repair of the defect in the abdominal wall created is also a problem when treating such cases because the repair power can lead to wound dehiscence and added morbidity. Using a plate for repair has been well described. Another method is the use of flaps tensor fascia lata [12]. Also, the assistance of our plastic surgery colleagues may be requested for the parietal closure as in our case.

Prognosis of squamous cell carcinoma of the bladder was poor describes at advanced stage of the diagnosis. Nevertheless, Kassouf et al. [13] reported an overall survival of 47.6% at 2 years with a median recurrence-free survival of 5.1 months.

Conclusion

The bladder exstrophy is a predisposing factor for the development of malignant tumors of the bladder. In such cases, Adenocarcinoma is the histological usual type. The squamous cell carcinoma contributes only a minority. The treatment of these tumors is to cystectomy associated with a reconstruction of the wall. Better results can be achieved in patients with localized disease, and especially when a multidisciplinary approach was adopted

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