

Squamous Cell Carcinoma in Bladder Extrophy : A Case Report and Literature Review

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Abstract

Background: Bladder extrophy is a rare congenital anomaly, it is now well reconized that an exstrophy of the bladder is associated with an increased risk of bladder cancer.

Case presentation: We report the case of 34-year-old male with squamous cell carcinoma of an unreconstructed exstrophic bladder. Because of the locally advanced disease, he received neoadjuvant chemotherapy with favorable response. Then, he underwent a total cystectomy with uretero-intestinal anastomosis.

Results: The patient progressed three months after the surgery. Chemotherapy was established and he received 2 cycles, however, he died 2 months later.

Conclusion: Squamous cell carcinoma associated with bladder extrophy remains relatively high. These tumors tend to be aggressive, follow-up is mandatory in these cases.

Keywords: Squamous Cell Carcinoma (SCC); Bladder extrophy

Background

Bladder extrophy is a rare congenital anomaly with an incidence of about 1 in 50,000 newborns [1,2]. It's a protrusion of the urinary bladder through a defect in the abdominal wall [3]. Bladder Exstrophy-Epispadias Complex (BEEC) describes a spectrum of birth defects involving failure of urogenital development during early gestation [4]. With the development of new repair techniques, the quality of life of patients has been greatly improved. However, it is now well recognized that exstrophy of the bladder is associated with an increased risk of bladder cancer [1]. Sixty percent of cancers in ectopic bladder occur during fourth and fifth decade of life, majority of them being adenocarcinoma (95%) [2]. We report here, a case of squamous cell carcinoma on a patient with neglected bladder extrophy.

Case Report

We report the case of a 34-year-old male patient, without medical history, who lived with an exstrophied bladder (exstrophy-epispadias complex). The patient noticed a progressive growth in his abdomen about 2-years prior to the presentation. By the time of diagnosis, it was ulcerated and surinfected. The examination of the lower abdomen revealed a classical exstrophy- epispadias complex in the suprapubic region of the anterior abdominal wall with noticeable growth in the bladder extrophy. A tumour measuring about 60 mm × 50 mm dominated the bladder area. The growth didn't extend into the urethra

and the scrotum was normal with normally descended testes. Rest of the systemic examination was within normal limits. Patient had no known comorbid illnesses. The lesion was biopsied and found to be invasive SCC (Figure 1). CT scan of the abdomen showed a classical Exstrophy-Epispadiasis complex with a mass in bladder extrophy invading the skin and the subcutaneous fat tissue, measuring 63 mm. The mass caused an obstruction of the bilateral ureteric orifices resulting in bilateral hydronephrosis, the right lumbar ureter measured 18 mm, remaining dilated until its delivery in the neo bladder, the same goes for the pelvic ureter measuring 20 mm. Many lymph nodes were involved: obturator, external iliacs, the largest one measuring 19 mm × 13 mm. Liver presented no lesions. Chest X-ray was normal.

The patient was planned for neoadjuvant chemotherapy and received three cycles with 5fluoro-uracil and Cisplatin. The reassessment showed a good local cutaneous response but no measurable tumour regression was shown in the CT scan. The patient was taken up for surgery. A wide excision of the tumour along with exstrophy-epispadias complex (entailing total cystectomy) was performed along with uretero-intestinal anastomosis using the Wallace method. The histopathology report showed a moderately differentiated squamous cell carcinoma (Figure 2) of size 58 mm × 55 mm invading the entire bladder wall (Figure 3), prostate (Figure 4), and the skin (Figure 5). The ureteral and uretral margin was not seen. Lymphovascular and perineural invasions were found. Three months after the surgery, a CT scan showed a local tumor progression. After the multidisciplinary consultation meeting: chemotherapy regimen 5FU-Cisplatin was established. The

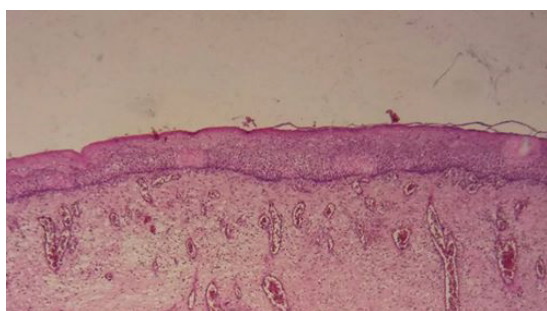


Figure 1: Mucosa of bladder extrophy in squamous metaplasia.

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Received May 16, 2017; **Accepted** June 22, 2017; **Published** June 24, 2017

Citation: Touil A, Loughlimi H, El-Abbassi S, Elouazzani H, Mahassini N, et al. (2017) Squamous Cell Carcinoma in Bladder Extrophy : A Case Report and Literature Review. J Clin Case Rep 7: 982. doi: [10.4172/2165-7920.1000982](https://doi.org/10.4172/2165-7920.1000982)

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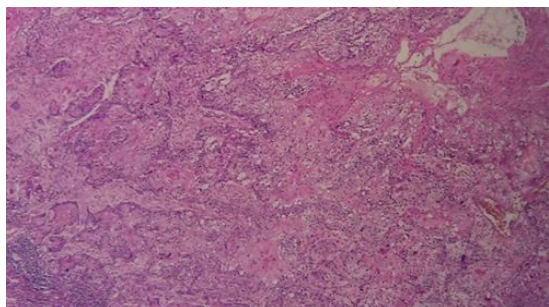


Figure 2: SCC: tumor's proliferation consists of nests and cords of tumoral cells, showing signs of keratinization with union bridge and horned globes.

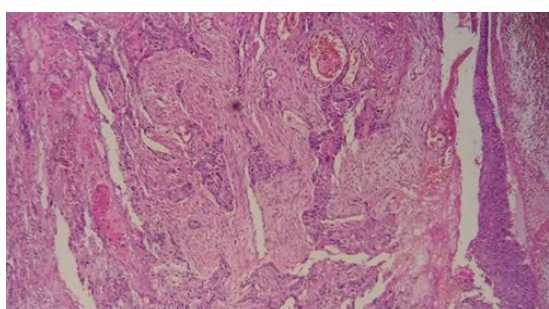


Figure 3: SCC infiltrating the bladder wall, the mucosa appears in the right of the image.

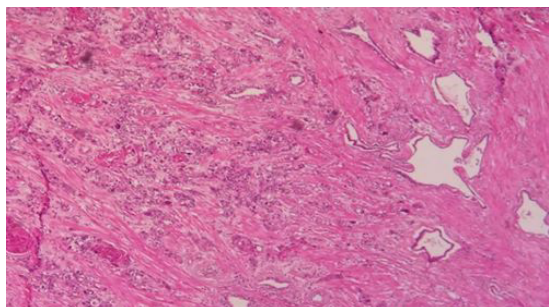


Figure 4: SCC infiltrating the prostate, the prostatic channels are crushed in the right of the image.

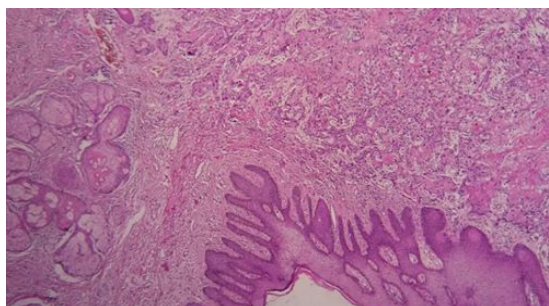


Figure 5: SCC invading the skin.

patient received 2 cycles of chemotherapy, however, the patient died 2 months later.

Discussion

Bladder exstrophy is a rare congenital anomaly [4,5]. Bladder

exstrophy-epispadias complex (BEEC) describes a spectrum of birth defects involving failure of urogenital development during early gestation [2,4,5]. Its incidence is estimated at 2.7 per 100,000 live births. Its etiology is attributed to abnormal partitioning of the cloacal membrane, which prevents migration of mesenchymal tissue for proper lower abdominal wall development [4]. It is more commonly diagnosed in males (Male: female - 5:1 to 6:1) [1]. The malignant potential of exstrophied bladder mucosa is well known [1,2], 95% are adenocarcinoma; and 3% to 5% are squamous cell carcinomas [1,2,5-7]. Majority (60%) of the malignant tumours in ectopic bladders occur during the fourth and fifth decade of life and out of the remaining (about 20%) occur after 60 years and before 40 years. [1,2,5,6].

Chronic irritation and infection leading to metaplastic transformation of the urothelium resulting in malignant changes is the most likely possibility [1,2,4,7]. Malignant degeneration of embryonic rests of gastrointestinal tissue can give rise to adenocarcinoma. It is also suggested that adenocarcinoma in exstrophic bladder originates from the colonic epithelium covering the mucosa of the organ [2].

Currently, exstrophy of the bladder can be diagnosed antenatally with the use of high-resolution real-time ultrasound [1]. With advances in pediatric medicine and reconstructive surgery, the morbidity and mortality of this disease has decreased drastically. In the developing world, however, one may come across these cases in older patients often because of a delay in seeking medical advice [1]. This was the case with our patient. The current golden standard for managing bladder exstrophies is surgical reconstruction [2]. Urinary diversion with removal of the bladder plate, along-with ureterosigmoidostomy, is an alternative, but carries a higher risk of malignancy [4]. The treatment of squamous cell carcinoma of the bladder remains similar to the treatment of urothelial cell carcinoma [5]. Meanwhile, the golden standard of treatment of squamous cell carcinoma of the bladder is radical cystectomy, with no established guidelines for any adjuvant or neoadjuvant therapy. Radiation therapy is offered to inoperable cases or those who refuse surgery [1,2].

There are no specific guidelines for follow-up in cases of squamous cell carcinoma. The guidelines for follow-up of bladder cancer in general are applied also for patients with squamous cell carcinoma [1].

The prognosis of SCC of the bladder is poor due to its advanced stage at diagnosis [1,2,5]. Kassouf et al. reported a median recurrence-free survival of 5.1 months [1,5]. Till date, no data is available to assess the differential prognosis of SCC and adenocarcinoma with regard to the exstrophy patients as most data is case reports [5].

Despite bladder closure or diversion surgery within the first few years of life, patients with exstrophy have an almost 700-fold greater incidence of carcinoma of the bladder than the age-matched general population. Early cystectomy is not protective [8]. Currently, no guidelines for screening of bladder carcinoma in patients with history of exstrophy with cystectomy have been established [4].

Conclusion

The development of reconstruction techniques improved the quality of life of patients with bladder exstrophy, however the risk of malignant degeneration remains high. Squamous cell carcinoma remains a rare entity. These tumors tend to be aggressive, good follow-up is highly recommended in these patients.

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing Interests

The authors declare that they have no competing interests.

Funding

Special thanks are due to the Faculty of Medicine and Pharmacy of Rabat; the source(s) of funding for all authors

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