



Squamous Cell Carcinoma on the Site of Bladder Exstrophy: A Case Report and Review of the Literature

Harrison Sumba*, Tidiani Bagayogo Kariba, Fouad Bakloul, Hamza Lamchahab, Tariq Karmouni, Kadir El, Khader, Abdelatif Koutani and Ahmed Ibn Attiya Andaloussi
Department Of Urology « B » Ibn Sina University Hospital Rabat Morocco.

ARTICLE INFO

Article history:

Received: 22 February 2016;

Received in revised form:

12 April 2016;

Accepted: 18 April 2016;

Keywords

Bladder exstrophy, squamous cell carcinoma, abdominal wall reconstruction.

ABSTRACT

Bladder exstrophy is a rare congenital anomaly. There are very few documented cases of adults presenting with bladder exstrophy in literature as well as its management which is poor and limited to retrospective studies of small number of cases. Malignant potential of the exstrophied bladder mucosa is well known. Sixty percent of cancers in ectopic bladder occur during fourth and fifth decade of life, majority being adenocarcinoma. This disorder is not only a psychological, social and professional handicap in adults but surgical difficulties also top-up its particular aspect owing to difficulty in parietal closure with frequent use of osteotomy, and rare chances of bladder preservation. We report herein a case of squamous cell carcinoma of exstrophied bladder in a 34-year-old male patient whom due to poor response neoadjuvant chemotherapy underwent radical cystoprostatectomy, urinary diversion with a cutaneous ureterostomy and reconstruction of hypogastric region. He is on a frequent follow up. The case is being reported on account of its rarity. Pertinent literature is being reviewed.

Résumé

Exstrophie vésicale est une anomalie congénitale rare. Il y a très peu de cas documentés dans la littérature d'adultes présentant une extrophie vésicale ainsi que sa prise en charge qui est pauvre et limitée à des études rétrospectives de petit nombre de cas. La malignité potentielle de la muqueuse de l'exstrophie vésicale est bien connue. Soixante pourcent des cancers sur une vessie ectopique surviennent pendant la quatrième et cinquième décennie de la vie, la majorité étant adénocarcinome. Ce trouble est non seulement un handicap psychologique, social et professionnel mais les difficultés chirurgicales aussi se surajoutent pour en faire de son aspect particulier du fait de la difficulté de fermeture pariétale avec l'utilisation fréquente de l'ostéotomie, et rares chances de préserver la vessie. Nous rapportons un cas de carcinome épidermoïde sur extrophie vésicale chez un homme de 34 ans dont due à une mauvaise réponse à la chimiothérapie néoadjuvante a subi une cystoprostatectomie radicale, avec une dérivation urinaire type urétérostomie cutanée, reconstruction de la région hypogastrique, il est surveillé régulièrement. Le cas est rapporté vu sa rareté. Une revue pertinente de la littérature.

© 2016 Elixir all rights reserved.

Introduction

Exstrophy of the bladder is a rare congenital anomaly with an incidence of about 1 per 50000 newborns. Malignant potential is high; 95% are adeno-carcinomas; 3 to 5% are squamous cell carcinomas [1,2]

To date, one hundred and ten cases of malignant neoplasms arising from untreated bladder exstrophy have been reported and early vesical reconstruction is the best oncologic prevention [3].

This disorder is of particular aspect in adults, who consult late, with reasons for not having approached for treatment

earlier include ignorance and poverty mostly in third world countries. This significantly alters their quality of life from a psychological, social and professional aspect.

Surgical management of this malformation disease particularly in adults is not easy, in view to abdominal wall closure.

We describe a case of uncorrected bladder exstrophy with squamous cell carcinoma discovered in a man in mid thirties.

Case Report

A 34-year old male, who lived with an exstrophied bladder, had noticed a growth-like change in the bladder,

progressive in nature about a year prior to presentation, with pain; and mild bleeding.

Examination revealed a relatively well-built man with a classical exstrophy-epispadias complex presenting a fierce looking growth measuring about 54 mm x 34 mm. Ureteric orifices were not discernibly visible. There was spillage of urine in between the growth. Scrotum was normal with normally descended testes (Figure 1). Digital rectal examination revealed no pelvic masses. The rest of the systemic examination was within normal limits. Patient had no known comorbid illnesses.

Contrast enhanced CT scan showed classical deformities of bladder exstrophy with a tumour invading deep muscles causing bilateral ureteric obstruction that resulted to bilateral hydronephrosis. Presence of lymph nodes in inguinal, iliac region. Absence of lesion in liver and lungs (Figure 2 and 3).

Biopsy of this growth revealed a squamous cell carcinoma.

In view of locally advanced disease, he received neoadjuvant chemotherapy consisting of 3 cycles of Cisplatin and 5FU. However, a reassessment after chemotherapy did not show a measurable tumour regression. He was taken up for surgery. He underwent a radical cystoprostatectomy with en block bilateral pelvic lymphadenectomy, urinary diversion with a cutaneous ureterostomy.

Reconstruction of the lower anterior abdominal fascia defect was undertaken with Prolene mesh with tensor fascial lata flaps without pubic approximation (Figure 4, 5, 6). We could achieve a tension-free closure of the anterior abdominal wall defect without any complication (Figure 7). Post operatively the patient had an uneventful recovery.

Histopathology report showed a moderately differentiated squamous cell tumour with invasion of the entire bladder wall, the prostate (pT4). Presence of polyhedral cells. No keratinocytes revealed. The cut margins of both ureters were free.

Post operative US Scan of the abdomen showed a marked reduction in the hydronephrosis. Renal function parameters were within normal range. The patient is on regular follow up, with improved self-esteem and social interaction.



Figure 1. Adult exstrophy before surgery; notice the fierce looking growth with urine spillage

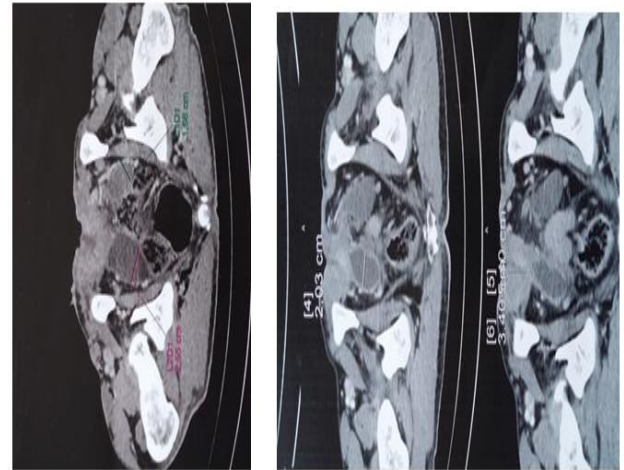


Figure 2. CT scan images of exstrophy bladder.

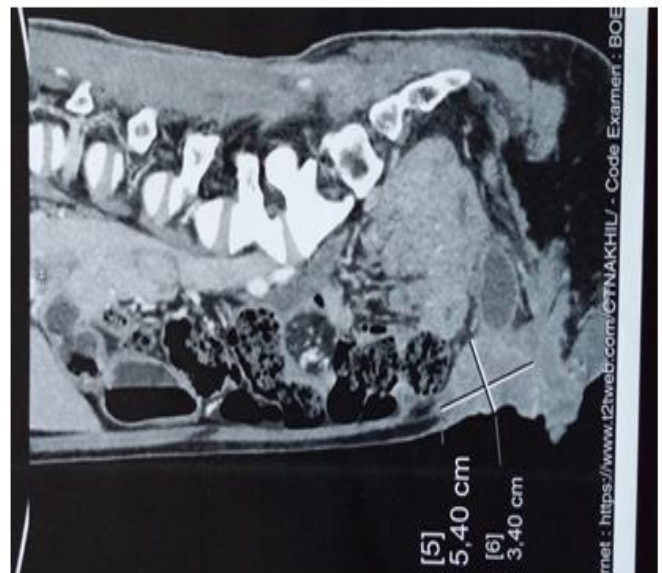


Figure 3. CT scan image of bladder exstrophy



Figure 4. Excision of bladder exstrophy.



Figure 5. Tensor fascia lata flaps prior to lower abdominal wall reconstruction.

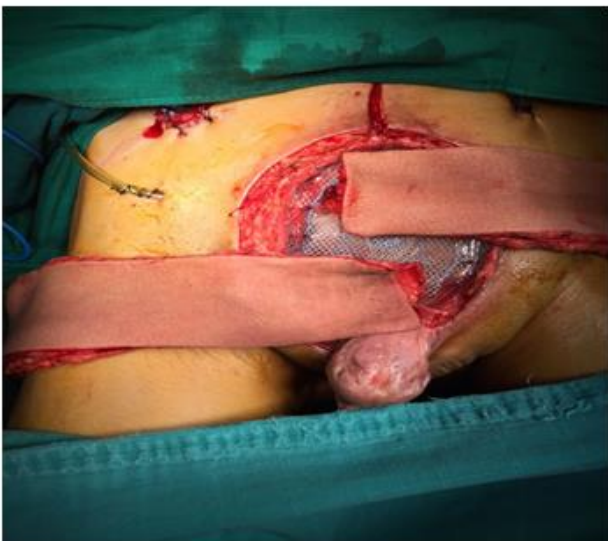


Figure 6. Prolene mesh in place to be covered by tensor fascia lata flaps.



Figure 7. Adult exstrophy after surgery: lower abdominal wall reconstruction by tensor fascia lata flaps and continent diversion.

Discussion

Bladder exstrophy is a serious malformation of lower abdominal wall with pubic diastasis and lateralized rectus muscle that causes difficulty in abdominal wall closure in surgery.

It is a rare congenital anomaly with well-known malignant potential if left untreated [4].

This bladder malformation is best managed in the neonatal stage with good results. Defect is obvious and definitive treatment with primary bladder wall reconstruction is possible in infancy.

Adults presenting with exstrophy is very rare and its incidence at this stage is not documented adequately in literature. Patients with untreated bladder exstrophy are exposed to significant social constraints: they have difficulties to socialize, carry out their professional activities within the population because of the stench of urine [5, 6], and main concern of consultation being urinary incontinence, urinary smell, and social ostracism.

Some of these patients show no interest regarding treatment primarily because of ignorance, financial constraint or poverty in developing countries.

Debate on management of bladder exstrophy in adults remains open, given the limited number of published studies [7-10].

Ureterosigmoidostomy was the first form of diversion to be popularized for patients with exstrophy. Although the initial series were associated with multiple metabolic problems, results improved markedly with newer techniques of reimplantation.

Difficulty of anterior lower abdominal wall closure also poses a peculiarly adult surgical problem. It resorts frequently to osteotomy. In our case, we did not have to resort to it. Mansour et al. reported the use of patch in bladder closure [7], while Pathak et al. reported the use of transposition grafts [8]. We used tensor fascia lata flaps, leading to satisfactory results.

The use of a rectus abdominis myocutaneous flap for closing exstrophy is more complex than the tensor fascia lata flap [11, 12]. Rectus fascial flaps have been used in abdominal repair because lack of support predisposes the bladder to reopening as well as fistula formation [13]. Marlex mesh [polyethylene] and Teflon mesh are expensive and not always available. We have used polypropylene [Prolene TM] mesh which is cheap and readily available in most of the centers.

Even though squamous cell carcinoma [SCC] of urinary bladder is considered as a separate entity, treatment remains similar to transitional cell carcinoma [TCC] [14].

Systemic chemotherapy regimens used to treat transitional cell carcinoma are generally ineffective for tumours with non-urothelial histology such as adenocarcinoma or SCC [15, 16]. However overall experiences with chemotherapy in non-urothelial carcinomas are limited.

Hayashi et al in a report of 18 cases of SCC bladder observed that radiation and cisplatin-based chemotherapy were effective supportive therapy to improve the prognosis [17]. Chemotherapy was given due to locally advanced disease T2b with ureteric involvement together with unwillingness on the part of patient to accept surgery.

About 115 cases of cancer in exstrophic bladder have been reported [18, 19]. There are no comprehensive guidelines on role of adjuvant or neoadjuvant chemotherapy.

The gold standard of treatment of SCC of bladder is radical cystectomy with no established guidelines for any adjuvant or neoadjuvant therapy. Radiation is used for cases found inoperable or where patient is reluctant for surgery [15].

In our case the choice of Gemcitabine & Cisplatin was guided by available literature reports for palliative chemotherapy in bladder cancer [15, 16, 17]. In view of poor response, and after discussion with patient and his consent, he underwent surgery.

He is on a frequent follow up with no renal insufficiency, improved self-esteem, social integration which has made the whole exercise meaningful.

Conclusion

Adult bladder exstrophy correction is a very rare practice in urology due the fact that this congenital disease is diagnosed and corrected in neonates. Patients with bladder exstrophy presenting in adulthood should not be denied surgical correction. Malignant potential is high in adults; we advocate the radical surgical management, after exclusion of any primary malignant sites related to the gastrointestinal tract. The abdominal-wall defect in bladder exstrophy is often large and accompanied by a wide detachment of the pubic bone. Prolene mesh closure and various transposition grafts technique are possible; we used tensor lata flaps technique in our case with satisfactory results. Surgical correction in adults with exstrophy of the bladder greatly improves self-esteem, confidence, and social relationship.

Financial or Other Competing Interests: The authors declare no financial or competing interest.

Authors Contribution: All mentioned authors have contributed to the elaboration and development of this manuscript.

References

- [1].de Riese WD, Warmbold H: Adenocarcinoma in exstrophy of the bladder, a case report and review of the literature. *Int Urol Nephrol* 1986;18 (2):159–162.
- [2].Gupta S, Gupta IM: Ectopia vesica complicated by Squamous cell carcinoma; *Br J Urol* 1976; 48:224.
- [3].Justrabo E, Poulard G, Arnould L, Pluot M, Zalmai I. [Invasive adenocarcinoma with epidermoid carcinoma on the site of bladder exstrophy. Histochemical and immunocytochemical study]. *Arch Anat Cytol Pathol*. 1991;39 (5-6):223-7.
- [4].Engel RM. Bladder exstrophy: Vesicoplasty or urinary diversion? *Urology*. 1973; 2:20-4.[PubMed]
- [5].El Khader K, El Fassi J, Koutani A, Ibn Attya A, Hachimi M, Lakrissa A. Psychological, sexual and social aspects of bladder exstrophy in adulthood. Analysis of 3 cases. *Ann Urol (Paris)*. 2003 Jun; 37(3):113-6.
- [6].Lee C, Reutter HM, Grässer MF, Fisch M, Noeker M. Gender-associated differences in the psychosocial and

developmental outcome in patients affected with the bladder exstrophy-epispadias complex. *BJU Int*. 2006 Feb; 97(2): 349-53.

[7].Mansour AM, Sarhan OM, Helmy TE, Awad B, Dawaba MS, Ghali AM. Management of bladder exstrophy epispadias complex in adults: is abdominal closure possible without osteotomy? *World J Urol* (2010) 28:199–204.

[8].Pathak HR, Mahajan R, Ali NI, Kaul S, Andankar MG. Bladder preservation in adult classic exstrophy: early results of four patients. *Urology*. 2001 May; 57(5):906-10.

[9].Shoukry AI, Shoukry I, Management of bladder exstrophy in adulthood: Report of 5 cases, *Journal of Pediatric Urology* (2012), <http://dx.doi.org/10.1016/j.jpuro.2012.06.004>

[10].Nerli RB, Kamat GV, Alur SS, Koura A, Prabha V, Amarkhed SS. Bladder exstrophy in adulthood. *Indian J Urol*. 2008 Apr; 24(2):164-8.

[11].O'Hare PM, Leonard AG. Reconstruction of major abdominal wall defects using the tensor fascia lata myocutaneous flap. *Br J Plast Surg* 1982; 35: 361 – 366

[12].Skef Z, Bellinger M, Bloman E, Ballantine T. Use of inferior rectus myocutaneous flap for coverage of bladder exstrophy defect. *J Pediat Surg* 1982; 17: 718 – 720

[13].Hosseini S, Sabet B, Zarenezhad M. Abdominal wall closure in bladder exstrophy complex repair by rectus flap. *Ann Afr Med* 2011; 10: 243 – 245

[14].Serreta V; Pomara G; Piazza F; Gange E: Pure squamous cell carcinoma of the bladder in western countries. *Eur Urol* 2000; 37 (1):85-9.

[15].Montie JE, Eisenberger MA, et al. NCCN Clinical Practice Guidelines in Oncology. Bladder Cancer (including Upper tract tumours and Urothelial Carcinoma of Prostate); V.2.2008. Available online at http://www.nccn.org/professionals/physician_gls/PDF/bladder.pdf

[16].Seifker-Ratke AO, Dinney CPN, Czerniak B, Milikan RE. Bladder Cancer: In MD Anderson Manual of Medical Oncology. Kantarjian HM, Wolf RA, Koler CA editors. 1st Edition. Tata McGraw Hill Publishers, New Delhi; 2007:739-756.

[17].Hayashi N, Asano K, Furuta A, et. al. Invasive squamous cell carcinoma of the bladder: report of 18 cases and review of literature. *Nippon Hinyokika Gakkai Zasshi* 2004; 95 (5): 711-7.

[18].Rieder JM, Parsons JK, Gearhart JP, Schoenberg M Primary squamous cell carcinoma in unreconstructed exstrophic bladder. *Urology* 2006; 67 (1):199-200.

[19].Ribeiro JC, Silva C, Sousa L, García P, Santos A. Squamous cell carcinoma in bladder exstrophy. *Actas Urol Esp* 2005; 29 (1):110-2.