Late presentation of ectopia vesica with malignant transformation. A case report and review

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1. Introduction

Exstrophy of the bladder (ectopia vesicae) is a rare congenital anomaly with an incidence of about 1 per 50,000. Nowadays it is usually surgically corrected in the neonatal period thus presentation in the adult period is a rarity. The malignant potential is considerable and most cases are adenocarcinomas but squamous carcinomas do occur [1]. We are reporting a patient with Ectopia Vesicae who had never sought medical advice regarding his condition and presented for the first time in his 7th decade with irreducible inguinal hernia. By that time he was discovered to have malignant transformation in the unreconstructed bladder extrophy. This work has been reported in line with the SCARE criteria [2].
ureter. Retrograde pyelography confirmed the ultrasound finding and showed a distal ureteric stricture (Fig. 2). A week was spent for temporal renal dialysis and optimizing his general condition, after which he underwent excision of Ectopia Vesicae under general anesthesia and urinary diversion via ureterosigmoidostomy which was chosen over ileal conduit because it is internal diversion with no ileostomy, moreover the risk of malignancy associated with ureterosigmoidostomy will be insignificant considering patient age. Mitrofanoff operation was a difficult option in the presence of irreducible hernia, which was found to be incarcerated omentum, small bowel and caecum. It was repaired using Bassini technique as the priority was to remove the tumor, urinary diversion and later on to have a second reconstructing surgery in a better set-up. Inguinal lymph node biopsy was taken. He went through an uneventful postoperative course and the histopathology showed metaplastic squamous mucosa and a moderately differentiated mucinous adenocarcinoma with lymph node involvement. After 3 weeks he had good fecal continence, normal renal functions, minimal residual hydro-ureter and hydro-nephrosis (Fig. 3) with a midline incisional hernia (Fig. 4). He was referred to the distant National Cancer Institute, for further management where he received chemo-radiation. He was on follow up there till he passed away nine months later.

3. Discussion

Exstrophy of the bladder (ectopia vesicae) is a rare congenital anomaly with an incidence of about 1 per 50,000. It is defined as an incomplete fusion of the mesoderm, which forms the tubercle genitalia, anterior wall of the bladder and inferior portion of the anterior abdominal wall. This incomplete fusion will manifest as rectus muscle diastasis, symphysis pubis separation and eversion of the posterior bladder wall into the anterior abdominal wall with separated scrotum/labia and divided penis/clitoris [1].

The deformity is usually treated in the neonatal period. Although some authors reported that 66–67% of unreconstructed ectopia vesica are dead by their third decade [3], and others documented almost normal life expectancy with reconstructed bladder extrophy and follow up [4], nevertheless it is unusual to come across a case of unreconstructed Ectopia Vesicae in late adulthood. Lack of awareness, ignorance, social embarrassment or even lack of appropriate facilities might be implicated in such delayed presentation. Even more it is uncommon to see a case of extrophy complicated by carcinoma. Here, we report a case who presented in his 7th decade, who had never sought medical advice regarding his

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**Fig. 1.** Fungating tumor in the ectopia vesica with the irreducible right inguinal hernia.

**Fig. 2.** Descending pyelogram showing a distal ureteric stricture, hydroureter, and hydronephrosis.

**Fig. 3.** Postoperative IVU: Resolved obstruction and functioning ureterosigmoidostomy.
Exstrophy or it is urological complications. He presented due to irreducible right inguinal hernia and was discovered to have adenocarcinoma in an unreconstructed ectopic urinary bladder.

Inguinal hernia incidence in ectopia vesicae was reported to be 86% in boys and 15% in girls with (78%) of cases being bilateral [5]. This may be due pubic bone separation and patent processus vaginalis, in our case hernia was the actual presenting complain of the patient.

The first of a case of carcinoma of the unreconstructed bladder extrophy was reported in 1895 [6]. Nielsen and Nielsen [7] reviewed 81 cases in 1983 and until now a total of 119 cases have been reported. Adenocarcinomas of the bladder is an uncommon malignant neoplasm and account for less than 2% of all bladder cancers [8]. However, the reported incidence among exstrophy patients varies from 3.3% to 7.5% [9,10]. Among patients with unreconstructed bladder extrophy, approximately 90% of malignancies were adenocarcinomas while 5% were squamous cell carcinomas [9,10,11,12]. These cancers tend to be aggressive [9,10].

The cause of the carcinogenesis in bladder extrophy is unknown yet, although there are many postulations. The epithelium of the extrophic bladder shows glandular metaplasia in the middle of the bladder and squamous metaplasia at the top of the trigon emerging into normal skin [13]. These metaplastic epithelium may have changed into malignancy in our case. Mechanical irritation on the bladder mucosa is another factor in carcinogenesis. Embryological origin such as misplaced rectal epithelium during the division of the cloaca is postulated.

There appears to be an increased risk of bladder cancer associated with HPV infection, which has been documented by the results of meta-analyses, although they failed to show a connection between HPV types and bladder cancer histology [14,15]. Alten et al recently implicate the presence of high risk HPV types as a strong carcinogenesis at least in the SCC component of the tumor [16].

Surgical diversion of urine into the bowel as a treatment for ectopia vesica was firstly performed by Simon in 1851 [17]. Later on ureterosigmoidostomy becomes more popular [18]. Some people preferred deferring operation of ureterosigmoidostomy until the age of 3 or 4 years, while others advocates reconstruction in the first instance [19]. Plastic operation, performed during the first few days of life and transplantation of the ureters into an isolated ileal loop was also described [17].

The strategy in the management of bladder extrophy in neonates, infants and children is to convert the extrophied bladder into a continent reservoir which can be periodically emptied either spontaneously or with assistance and at the same time to preserve the upper tracts. This can be achieved in one or multiple stages. The procedures available for the reconstruction are: bladder closure, bladder neck repair, epispidias repair, ureteric reimplantation, bladder augmentation, bladder neck division and a catheterizable continent stoma. These procedures are fully capable of achieving the goals of management, with a little help from medication to relax the detrusor [20].

In adult cases the fibrotic nature of the bladder wall and the absence of the sphincter at the bladder neck make the reconstruction of a distensible and continent bladder almost impossible. In our case we performed the only possible option in our limited set-up which was radical removal of the bladder with diversion by ureterosigmoidostomy (Fig. 4). The patient present later with incisional hernia which is attributed to tension repair since pelvic osteotomies was not done. We recommend abdominal wall closure using fasciocutaneous plasty, flaps or mesh, this was not done as we lack the mesh and the expertise in our remote setting.

Although bladder extrophy occurs with other variants of congenital anomalies, it is combination with renal agenesis was described as a rare condition in the literature [21,22,23], to the best of our knowledge no case was reported to survive long enough up into the 7th decade with unreconstructed deformity and renal agenesis without seeking medical care for either his malformation or it is urological complications.

4. Conclusion

Despite being so unfortunate to have the extremely rare condition of ectopia vesica and renal agenesis complicated with advanced malignancy, the surgical excision and diversion kept this old man satisfied and dry. It is remarkable that he survived long years without any medical care, however efforts should be made to pick up and treat such anomalies much earlier.

Conflicts of interest

Nothing to disclose.

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Ethical approval

Ethical approval obtained from: Gadarif University – Scientific Research Ethical Committee.

Consent

Written and signed consent is obtained.

Author contribution

1-Sami Eldirdiri: The main author and general surgeon whom the patient belongs to and did the operation in a team with.
2-Rehab M. Elmushly : Plastic surgeon working in the same team.
3-Sami G. Elazhary: General and colorectal surgeon who used to work in our team and participates in the writing.

Registration of research studies

This is a case report.

Guarantor

Sami Eldirdiri.

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