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

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ABSTRACT

INTRODUCTION: Exstrophy of the bladder is a rare congenital anomaly usually treated in neonatal or childhood period. When combined with renal agenesis and presents for the first time in the adulthood with malignant transformation, is an extreme rarity.

CASE PRESENTATION: We present a case of 65 years single male who presented with a right irreducible inguinal hernia and an unreconstructed Ectopia Vesicae with fungating tumor. He was anemic with impaired renal function, left renal agenesis and right sided hydronephrosis, hydroureter and distal ureteric stricture. He underwent palliative excision of Ectopia Vesicae and urinary diversion via ureterosegmoidostomy after his condition was optimized. Histopathology showed metaplastic squamous mucosa and a moderately differentiated mucinous adenocarcinoma. Three weeks later he had good continence and normal renal function. He was sent to a distant radio-oncology center for further management.

DISCUSSION: In 1851 the first ureterosegmoidostomy for ectopia vesicae was done. Later on it becomes more popular. Some people preferred deferring it until the age of 4 years while others advocates earlier reconstruction. Plastic operation, during neonatal life was also described. In the majority of cases, the fibrotic nature of the bladder and the absence of the sphincter make the reconstruction almost impossible. We performed the only possible option in our setting as our patient had a complex congenital anomalies which present late in life complicated with advanced malignant transformation.

CONCLUSION: Despite the plethora of congenital malformation and advanced malignancy, surgical excision and diversion with adjuvant chemo-radiation provided a good palliation for this patient.

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 exstrophy. This work has been reported in line with the SCARE criteria [2].

2. Case report

We are reporting a 65 years old single male farmer who was brought to our hospital, which is the main community hospital in the state, with a right irreducible inguinal hernia without symptoms of bowel obstruction. On clinical examination he was found to have a right side irreducible inguinal hernia, non-repaired ectopia vesica with fungating mass and multiple bilateral lymphadenopathy (Fig. 1). His brother informed our team that wasn't seen by any health personnel after being born at home or during his infancy and childhood. Later on in his adult hood, their low socioeconomical status and social embarrassment prevent him from seeking medical advice. He tried to keep himself dry by wearing a lot of cloths, perfumes and always spreading incense around to flush the odor.

Upon assessment, he was anemic with impaired renal function, his serum creatinine level was twice the normal value (Risk according to RIFLE classification) but we were not able to determine the progression of his disease due lack of previous medical records. The rest of blood tests were within normal limits. Ultrasound revealed left renal agenesis and right sided hydro-nephrosis, hydro-

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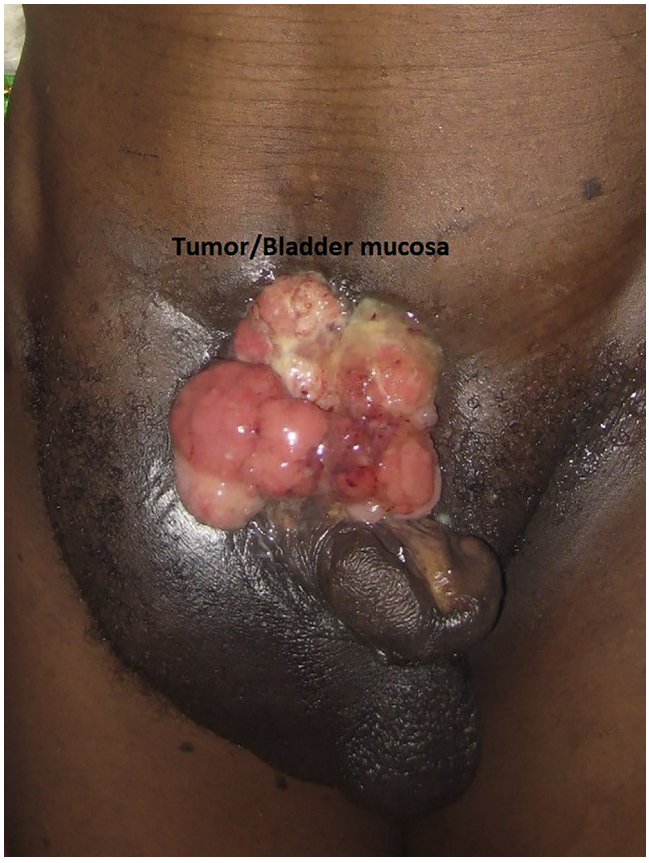

Fig. 1. Fungating tumor in the ectopia vesica with the irreducible right inguinal hernia.

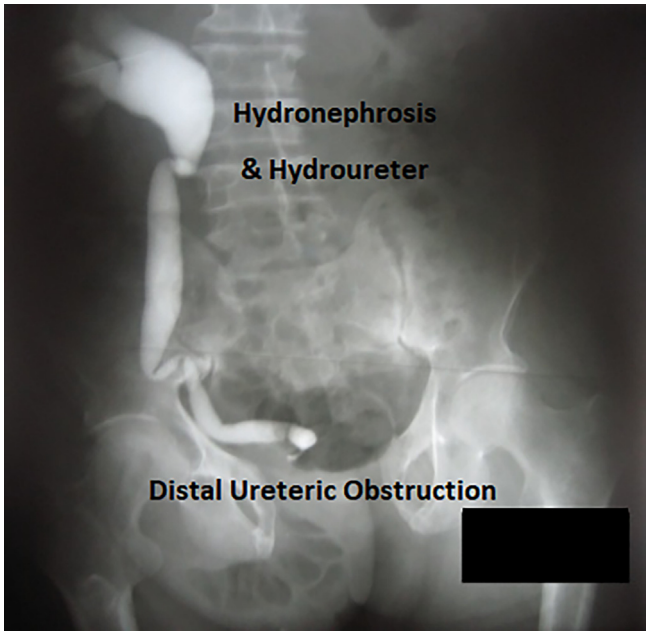

Fig. 2. Descending pyelogram showing a distal ureteric stricture, hydroureter, and hydronephrosis.

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 ureterosegmoidostomy

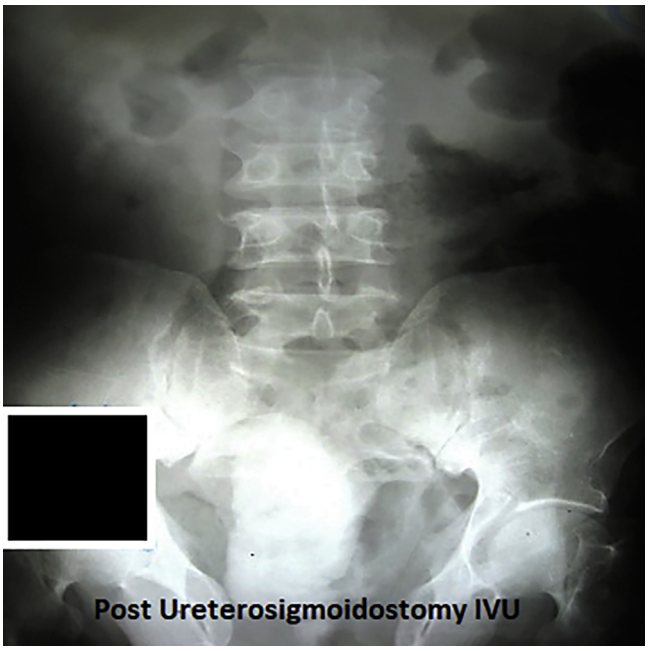

Fig. 3. Postoperative IVU: Resolved obstruction and functioning ureterosigmoidostomy.

which was chosen over ileal conduit because it is internal diversion with no ileostomy, moreover the risk of malignancy associated with ureterosegmoidostomy 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 exstrophy and follow up [4], nevertheless it is unusual to come across a case of unconstructed 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 is to see a case of exstrophy complicated by carcinoma. Here, we report a case who presented in his 7th decade, who had never sought medical advice regarding his

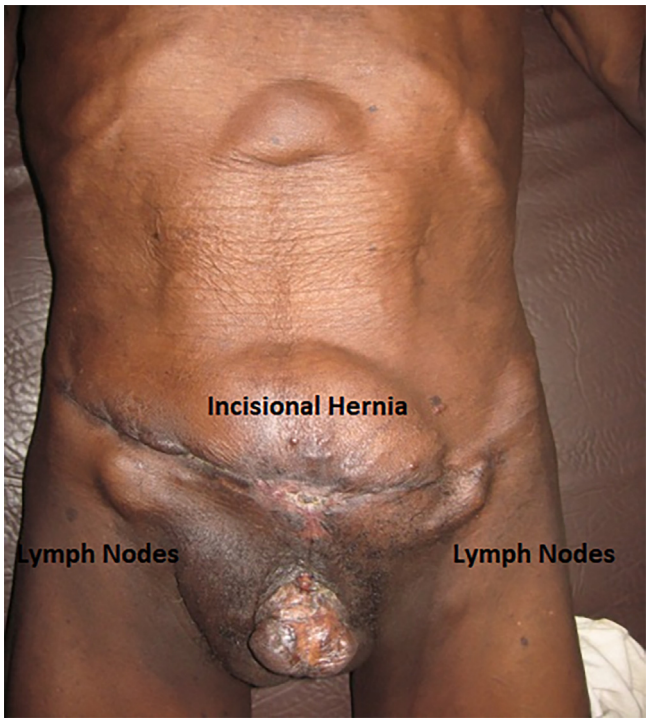

Fig. 4. Postoperative.

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 exstrophy 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 exstrophy, 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 exstrophy is unknown yet, although there are many postulations. The epithelium of the exstrophic 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 ureterosegmoidostomy becomes more popular [18]. Some people preferred deferring operation of ureterosegmoidostomy 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 exstrophy in neonates, infants and children is to convert the exstrophied 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, epispadias 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 ureterosegmoidostomy (Fig. 4). The patient present later with incisional hernia which is attributed to tension repair since pelvic osteotomy 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 exstrophy 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 Eldiridiri.

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