



Contents lists available at ScienceDirect

## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

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

Sami Eldirdiri<sup>a,\*</sup>, Rehab M. Elmushly<sup>a</sup>, Sami G. Elazhary<sup>b</sup><sup>a</sup> University of Gadarif, Sudan<sup>b</sup> Ibrahim Malik Hospital, Sudan

## ARTICLE INFO

## Article history:

Received 7 March 2018

Received in revised form 10 April 2018

Accepted 30 April 2018

Available online 10 May 2018

## Keywords:

Case report

Exstrophy

Ectopia vesicae

Renal agenesis

Ureterosegmoidostomy

Malignant transformation

## 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.

© 2018 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 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 socioeconomic 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-

\* Corresponding author at: Gadarif University, Faculty of Medicine & Health Science, P. O. Box 449, Gadarif, Sudan.

E-mail address: [samielirdiri@gaduniv.edu.sd](mailto:samielirdiri@gaduniv.edu.sd) (S. Eldirdiri).



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

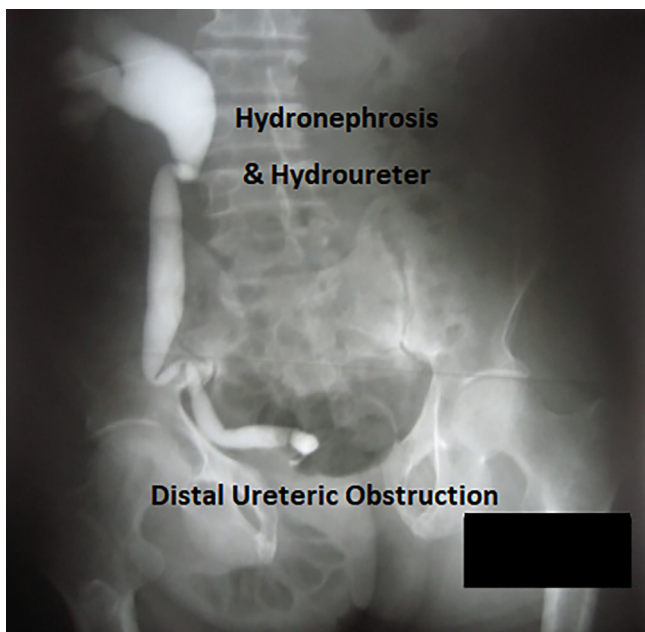


Fig. 2. Descending pyelogram showing a distal ureteric stricture, hydroureter, and hydronephrosis.

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

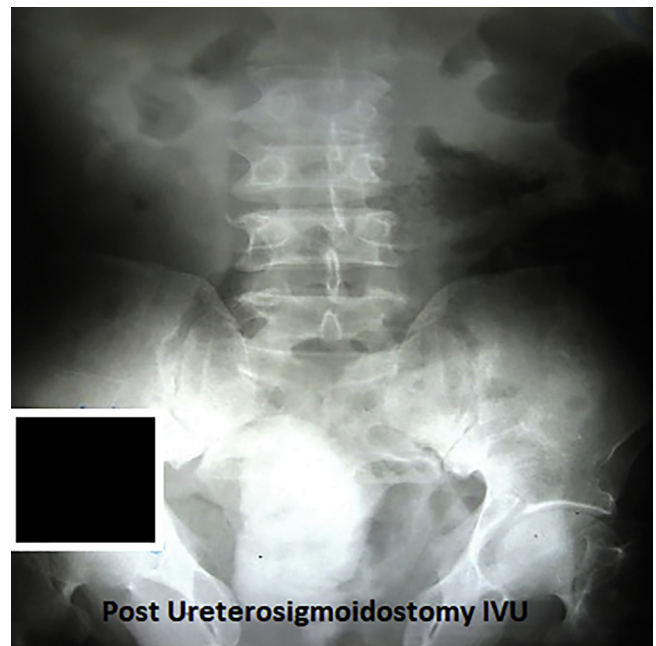


Fig. 3. Postoperative IVU: Resolved obstruction and functioning ureterosegmoidostomy.

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 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

Download English Version:

<https://daneshyari.com/en/article/8832558>

Download Persian Version:

<https://daneshyari.com/article/8832558>

[Daneshyari.com](https://daneshyari.com)