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CASE REPORT

Bladder exstrophy: An atypical journey of a rare case of an adult

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Abstract

Bladder exstrophy is a rare and complex malformation. Often associated with diverse deformations, an accurate diagnosis is a must for adequate management. In the African setting, especially in a remote area, delivery rarely occurs within a healthcare structure, thus reducing the chance of an early diagnosis. Due to a low density of specialists, people in rural areas refer to traditional healers for healthcare problems, the thing that delays the time to diagnosis. We report, according to the CARE guidelines (https://www.equator-network.org/reporting-guidelines/care/), a case of an 18-year-old male patient who was transferred to us for better management of a reddish hypogastric mass leaking urine. Initially considered as a congenital wound, his parents applied traditional products to heal it. Following the poor outcomes, the parents will consult several general physicians; unfortunately, no one gave a clear diagnosis. The clinical examination on admission revealed a good general condition and noted the presence of a reddish, ovoid structure of about 9 centimeters of the minor axis and 11 centimeters of the major axis in the hypogastric region allowing urine to flow in its upper part. The external genitalia examination revealed a retracted and short penis with urine exit at its base through the reddish structure. The patient was transferred outside the country for better management due to a lack of dedicated equipment. Although vesical exstrophy is a rare disease, an early diagnosis allows adequate management and good outcome. Thus, a well-performed neonatal examination is required to avoid later diagnosis and complications.

KEYWORDS

bladder exstrophy, democratic republic of congo, urinary incontinence, urological malformation

1 | INTRODUCTION

In African communities, the delivery of a child with deformities is experienced as a real tragedy due to the mysticoreligious considerations that surround it and the financial burden that constitute such a situation for the families.

Bladder exstrophy is a rare and complex malformation, which is characterized by the absence of closure of the entire anterior medial abdominal wall under the umbilical and the anterior wall of the bladder, also affecting the urethra, the pelvic girdle, the perineum, and the external genitalia. It presents as a reddish, vulturous plaque that bulges under the abdominal thrust and is an integral part of this wall called the bladder plate.[1,2,3] It is associated with several deformities. Among them, the most found are those of the urinary tract (epispadias, genital tract (micropenis), and osteoarticular system (diastasis of the pubic symphysis)).[3]

Bladder exstrophy remains one of the most challenging conditions managed by pediatric urologists. The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000 live births. Reconstructive management of exstrophy bladder is the best started in the neonatal period with good results.[4] When left untreated, exstrophy has malignant potential. It is associated with an increased risk of bladder cancer, with 95% of the arising tumors being adenocarcinomas and 3%–5% being squamous cell carcinomas.[5] We report a case that was brought to us for management of a hypogastric wound from birth, later diagnosed as bladder exstrophy.

2 | CASE PRESENTATION

This is an 18-year-old patient who was transferred to us for better management of a reddish hypogastric mass leaking urine. This mass was taken as a (congenital) wound by the parents who used to apply traditional products to heal it.

In his prenatal history, his father reported that no prenatal monitoring had been performed; his parents were illiterate and lived in a village located 250 km from the city of Kindu, in the eastern part of the Democratic Republic of Congo. We did not notice any consanguinity history. No regular child care was provided. The patient underwent several clinical examinations, but no one of them was performed by a urologist.

The clinical examination shows a patient with appropriate weight and height with a good functional status. In the hypogastrium, we noted the presence of a reddish, ovoid structure of about 9 centimeters of the minor axis and 11 centimeters of the major axis allowing urine to flow in its upper part (Figure 1). The examination of the external genitalia notes a retracted and short penis with urine exit at its base through the reddish structure (Figure 2). The patient had a characteristic "ducking" gait, and abdominal and pelvic ultrasound showed the absence of anterior abdominal wall muscles, but the bladder was not identified. Intravenous urography was not performed due to a lack of resources, and the other laboratory tests (blood and urine) were within the norms. The psychiatric examination conducted noted a psychologically unstable patient.

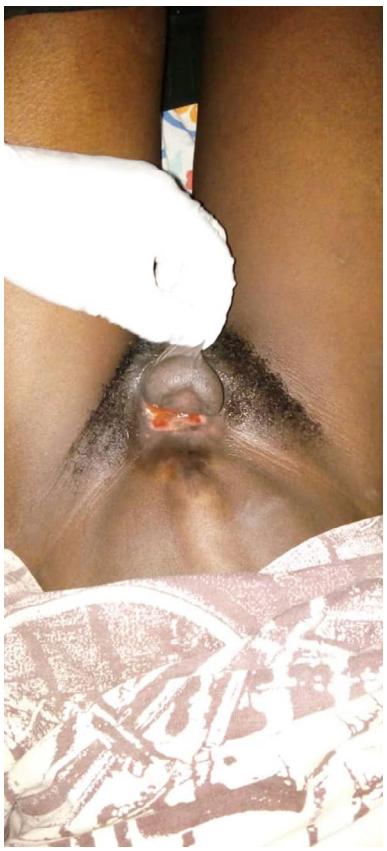

FIGURE 1 An 18-year-old patient with bladder exstrophy: Front view

Due to a lack of adequate equipment for his care, the patient was transferred outside the country.

3 | DISCUSSION

Bladder exstrophy results from an abnormality of the cloacal membrane that forms the abdominal wall under the umbilical cord during the first weeks of embryonic life.[6] The diagnosis is based on a nonvisualization of the bladder theoretically possible from the ultrasound of the first trimester, and once diagnosed, it constitutes a cause of medical interruption of the pregnancy.[7] In

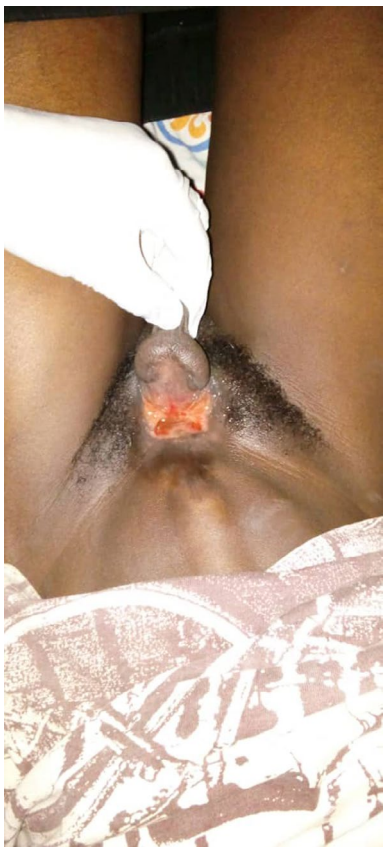

FIGURE 2 An 18-year-old patient with bladder exstrophy: Superior view

the African context in general, and in the Democratic Republic of Congo, in particular, prenatal follow-up, accurate examination of the newborn at birth, and postnatal follow-up are often inexistent in the remote areas, thus precociously excluding the possibility to diagnose the malformation.

The spontaneous evolution is often marked by an alteration of the ureters and the kidneys by ascending infection and stenosis. Besides that, carcinogenesis is 200 times higher in these patients compared to the general population. When diagnosed during the neonatal period, several treatments can be proposed to improve the quality of life, including perfect parietal reconstruction and urinary diversion by urostomy, which is a heavy and complex surgery[8] that is difficult or impossible to perform in our environment due to the lack of necessary equipment.

Also, previous studies noted that the treatment must be accompanied by psychological assistance, as the majority of these patients are unstable.[9] Hence, the importance of an awareness campaign for pregnant women in low-income countries (LICs), such as the Democratic Republic of Congo, to attend antenatal visits for an early diagnosis of congenital deformities.

4 | CONCLUSIONS

The lack of follow-up of antenatal visits during pregnancy explains the diagnosis of several deformities including postnatal bladder exstrophy in some remote areas of the country where its management is impossible.

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CONFLICTS OF INTEREST
None.

AUTHOR CONTRIBUTIONS
JTM, GBM, and DSN have designed, conceptualized the study, and written the first draft under the supervision of LMM. GMB, PMB, and FGC contributed to the drafting of the paper. All authors have reviewed and approved the final manuscript.

ETHICAL STATEMENT
This case report received ethical clearance from the Ethical Committee of the Catholic University of Bukavu.

CONSENT
Written informed consent was signed by the patient prior to the publication of this paper.

DATA AVAILABILITY STATEMENT
All the materials used in this study are available on request.

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