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Inferior vesical fistula: A rare case report of exstrophy variant with literature review

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ABSTRACT

We present a rare case of exstrophy variant - inferior vesical fistula. A two-year-old boy with normal continence presented with a suprapubic pit and mucosal plate on the penile dorsum. Examination and cystoscopy revealed vesical fistula. The fistula was closed and the penis was reconstructed. Recognition of the exstrophy variant is important because the treatment and prognosis are very different.

Key Words: Bladder exstrophy, variants of exstrophy, vesical fistula.

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Introduction

The variants of the exstrophy complex are very rare developmental abnormalities with 8% of the total cases [1-6]. Inferior vesical fistula is also a variant, in which there is a tiny communication between the lower part of bladder and the exterior. To date, only one girl has been reported in the literature [6]. Herein, we present the first case of an inferior vesical fistula which is a very rare variant of exstrophy in a 2-year-old boy.

Case report

A 2-year-old boy was brought by his parents with the complaint of an opening in the suprapubic region. On the other hand, the patient was asymptomatic and passing urine in good stream from meatus at tip of penis. There was no discharge of urine from the opening. There was no abdominal pain or dysuria. In addition, there was no history of incontinence and fever or urinary tract infection. On physical examination, a 0.5x0.5cm opening was present at base of the penis dorsally with no discharge. A wide and pale mucosal plate was observed in the dorsum of the penile shaft (Fig. 1). The dorsal prepuce was deficient. The glans was conical and adequate. The meatus looked normal and was at the tip of the glans. The testes were bilaterally descended.

Abdominal and urinary ultrasonography (USG) was normal. A micturating urethrogram (MCU) was performed, and it showed normal bladder and urethra without vesicoureteral

reflux (VUR) (Fig. 2). Diagnostic cystoscopy through the normal meatus showed an opening in the anterior wall of the bladder just above bladder neck. Scopy done through the vesical fistula showed a 2 cm mucosa-lined channel between the bladder and skin. On complete distention of the bladder with dilute betadine, spill of the betadine from the site of the fistula was visualized (Fig. 3).

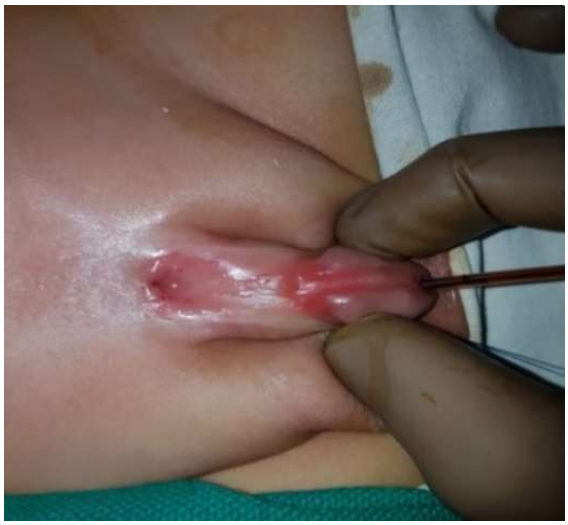

Fig. 1. Clinical image of the inferior vesical fistula.

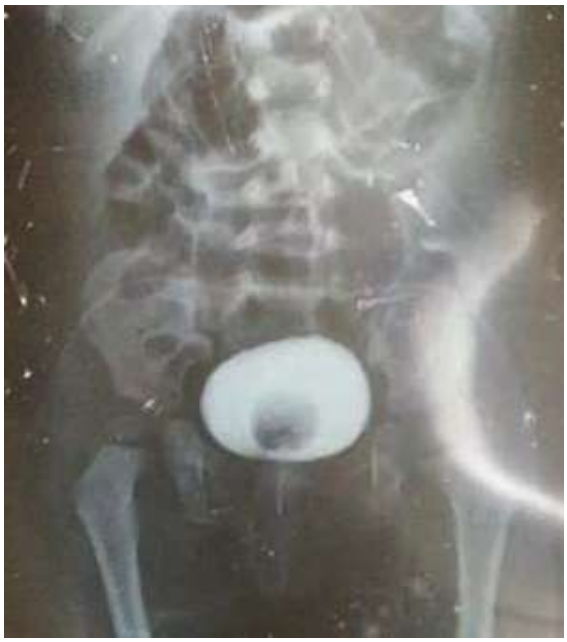

Fig. 2. A micturating urethrogram.

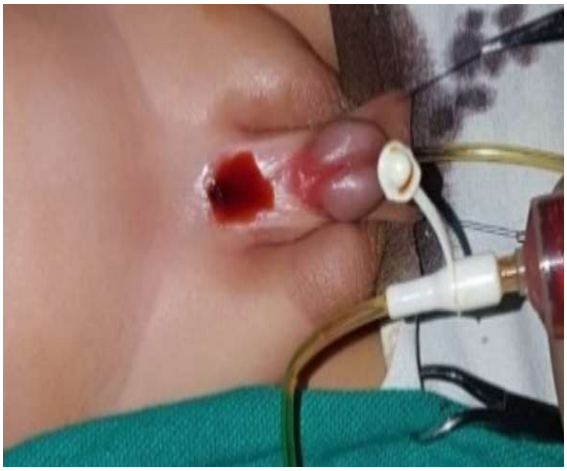

Fig. 3. View of betadine leak from the inferior vesical fistula.

At surgery, the vesical fistula was dissected all around and was found to pass suprapubically in the midline above the two corporal bodies (Fig. 4). Double ligation of the fistulous tract was done. The mucosal plate was excised and the penile skin was reconstructed.

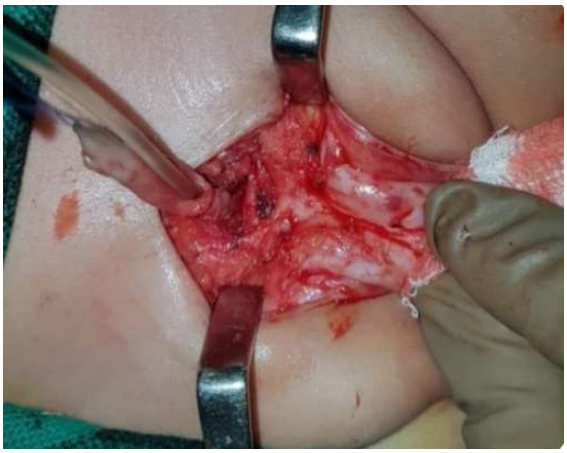

Fig. 4. Intraoperative view of the fistula tract.

Postoperative period was uneventful. Histopathology was consistent with vesicocutaneous fistula.

Discussion

Bladder exstrophy is a rare anomaly, with an incidence of 1 in 50000. Bladder exstrophy variants constitute 8% of these [1]. Variants

tend to occur more commonly in females, in contrast to the usual male predominance in typical bladder exstrophy [2]. Variant lesions have all the usual musculoskeletal findings of classic bladder exstrophy, but the bladder is closed with varying degrees of skin and subcutaneous cover and the urethra and sphincter mechanism may be intact [3].

Classic vesical exstrophy is thought to result from persistence or overdevelopment of an abnormal infraumbilical cloacal membrane that acts as a wedge, keeping the lateral mesoderm from progressing medially between its ectodermal and endodermal layers. Lacking mesodermal support, this membrane ruptures, resulting in an absent lower abdominal wall and exposed bladder. Variants are explained by incomplete rupture and/or persistence of the abnormal cloacal membrane [3]. The size, position and timing of rupture of cloacal membrane will determine the type of variant that results. In inferior vesical fistula, the presence of a normal bladder neck and urethra suggests a higher position of the cloacal membrane before its rupture. Variants were first classified by I. D. Williams as superior vesical fissure, vesical fistula and duplex bladder [4]. Other variants were subsequently elaborated by other authors. A complete classification is difficult because of varying features of each case. However, variants are mostly classified into pseudoexstrophy, covered exstrophy, superior vesical fissure and fistula, duplicate exstrophy, inferior vesical fissure and fistula. In pseudoexstrophy, symphysis pubis is widened but there is a normal anterior abdominal wall, bladder and urinary tract [5]. It is the mildest form.

Covered exstrophy variant is classic exstrophy with all of the accompanying defects but with a bladder that is closed and covered only by a thin, often translucent membrane of skin without muscle or fascial layer. In superior vesical fissure, pelvic deformity occurs, but only the upper bladder is open near the umbilicus. This is the most commonly described variant in literature (>30). In superior vesical fistula, defect and communication are small. Duplicate exstrophy is a rare variant in which patient presents with a patch of exstrophied bladder, a normal appearing abdominal bladder, diastasis of the symphysis pubis and rectus muscles and a form of epispadias.

Inferior vesical variants of exstrophy involve the lower part of bladder above the bladder neck. These can either be a fissure or a fistula; the fissure has a limited eventration of the bladder, whereas the fistula is a tiny communication between the bladder and the exterior [6]. These are extremely rare. Mahajan et al described a case of inferior vesical fistula in a 20 month old female who presented with continuous dribbling of urine since birth from a small opening situated just above base of clitoris. Urethra was normal. Simple closure of the fistula was followed by a normal micturition pattern [6].

Conclusion

Recognition of exstrophy variant is important because the treatment and prognosis are very different. Management of all variants consists of restoring the bladder integrity and fashioning of a good urethroplasty. For inferior vesical fistula, simple closure of fistula is the only treatment required and the prognosis with regards to urinary incontinence is good.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

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