Case Report

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Covered and Duplicate Exstrophy with Duplication of Bladder, Urethra, Vagina and Dextrocardia: A Case Report

S.		Mullick
B.B.		Jolly

Department of Paediatric Surgery, Safdarjung Hospital, New Delhi, India

Key Words

Exstrophy, covered
Ectopia vesicae
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Urethra duplication
Vagina duplication

Abstract

Variants of classical exstrophy are exceedingly rare. We report a case of covered and duplicate exstrophy with duplication of the bladder, urethra and genital tract with dextrocardia.

Dr. S. Mullick, MCh, D II/141, West Kidwai Nagar, New Delhi 110023 (India)

Introduction

Classical exstrophy of the bladder is not as rare as its variants. The usual problems of exstrophy are not encountered in covered and duplicate exstrophy because of nearly complete skin cover and the relatively well-developed bladder, urethra and external genitalia. We describe a patient with covered and duplicate exstrophy with duplication of the bladder, urethra and genital tract with dextrocardia. We could not find a similar case in the presently existing literature.

Case Report

A 1-year-old female presented with double stream of urine since birth. She was the first born and weighed 3 kg at birth; delivery was full-term and uncomplicated. There was no history of drug intake, fever or exposure to radiation in the antenatal period. Physical examination revealed two paramedian swellings between the umbilicus and pubis. The right one had a normal skin covering, but the left one showed a red surface at the centre. The clitoris was bifid and the labia were widely separated. Four openings were presented between the labia, two on either side of the midline (fig. 1). Urine emanated from the upper two orifices. Plain radiograph of the abdomen revealed wide pubic diastasis and a liver shadow on the right side. A chest radiograph showed dextrocardia (fig. 2). Excretory urogram revealed normal upper tracts; both the ureters opened into separate bladders.

Cystogram showed two bladders and urethrae separately (fig. 3). Cavitogram through the lower openings showed separate filling of the vaginae. Cystoscopy and colposcopy confirmed the above findings. Barium enema excluded the possibility of colonic duplication. No attempt was made to excise the patch of exstrophic bladder mucosa, it was left to undergo metaplasia and epithelization. The patient was lost to follow-up.

Discussion

Classical bladder exstrophy occurs in 1 in 50,000 live births [1]. Variants of exstrophy, like covered exstrophy, duplicate exstrophy, superior vesical fissure, and pseu-doexstrophy, are very rare. These variants have musculoskeletal findings of classic exstrophy, including divergent recti and spread symphysis pubis, but the bladder is closed and a varying degree of skin cover is present. This bladder covering may consist of skin alone or skin with a patch of ectopic vesical mucosa. In our case the right bladder was covered with skin alone, while the left one was covered with skin having a patch of ectopic vesical mucosa. These exstrophy variants are attributable to a delay in the mesodermal progression. If, however, there is early dehiscence of the cloacal membrane as well as a delay in the mesodermal ingrowth, duplicate exstrophy may oc-

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Fig. 1. External genitalia revealing bifid clitoris, two vaginae, and two urethral openings.

Fig. 2. Radiograph of chest revealing dextrocardia.

Fig. 3. Cystogram revealing duplication of bladder urethra (a) and pubic diastasis (b).

cur. Depending on the stage of development in the region of cloaca, duplication may affect the bladder and urethra alone, the vagina and uterus alone, the rectum and colon alone or a combination of these [2]. Complete duplication of the bladder and urethra is rare. Only 45 cases

have been reported so far [3]. This anomaly is more common in males than in females. Duplication of the lower urinary tract and genitalia with exstrophy is exceedingly rare, only 3 cases have been reported so far [3]. To the best of our knowledge a combination of these abnormalities with dextrocardia has not been reported.

Associated congenital anomalies of other organ systems are present in the majority of cases. In 40 cases reviewed by Kossow and Morales [4] 90% had some type of duplication of external genitalia and 42% had duplication of the lower gastrointestinal tract. Spinal duplication and fistula between the rectum, vagina and urethra were other associated abnormalities [4]. These patients may

present with incontinence, double urethral stream and infection. These are the symptoms that warrant surgical intervention. In duplicate exstrophy, treatment is gratifying because a normal bladder and urethra are present in most cases [5].

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