Agenesis of penis

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INTRODUCTION

Agenesis of penis is a very rare developmental anomaly, which is usually associated with scrotal hypoplasia or other urological developmental anomalies. Here we present such a case who presented to us with stenosis of newly constructed urethra.

Keywords: Agenesis, neo-urethra, penis

CASE REPORT

A 32-year-old male presented with the complaints of the absence of penis since birth and difficulty of micturition through the perineal site since 2 years. His old medical records suggests that he had his urethral opening in the anus at the time of his birth for which, he underwent some sort of surgery at the 1 year of age, when his urethral opening was made at the perineum. He had a complaint of recurrent urinary tract infection in his childhood for which he was evaluated and found to be having right sided gross hydroureteronephrosis and complete duplex system, for which he underwent right nephroureterectomy at his 2 years of age. On histopathological examination, it was found to be having dysgenetic right kidney. In our first visit, on examination, the patient had absent penis with normal looking scrotum and bilateral testes. The scrotum, testes, and testicular function are usually normal.

Agenesis of penis is a very rare developmental anomaly with the reported incidence of 1 in 30 million births worldwide. It is believed to result from either the absence of the genital tubercle, or its failure to develop. Absences of corpora cavernosa and corpora spongiosum are prerequisite for this developmental anomaly as claimed by several investigators. Except for the reported XX-XY mosaic, patients have 46 XY karyotypes. More than half of these have associated anomalies, including developmental defects of the caudal axis, genitourinary, and gastrointestinal tract anomalies. The scrotum, testes, and testicular function are usually normal.

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and ureter) and are normally placed in their respective position. Ascending urethrogram was done, which showed to be having meatal stenosis and stenosis of distal urethra [Figure 3]. Meatal dilatation and cystoscopy were done under spinal anesthesia. Cystoscopy suggested as normal urethra, verumontanum, prostate with slightly elevated bladder neck, urinary bladder was normal with only left ureteric opening.

**DISCUSSION**

The diagnosis of penile agenesis requires the absence of corpora cavernosa and copora spongiosum with urethra opening at any point on the perineum in midline, over pubis, anterior aspect of the scrotum, or, most frequently just anterior to the anus and anterior wall of the rectum.\(^6\) Embryological basis of this rare anomaly has been described as a result of failure of genital tubercle to develop with incomplete separation of the urogenital sinus from the hindgut by the urorectal septum. This rare entity should be differentiated from concealed penis, rudimentary penis, micropenis, epispadias, hypospadias pseudohermaphroditism, and intrauterine amputation of penis.\(^7,8\) Anorectal anomalies such as imperforate anus, congenital rectal strictures and rectovesical fistula, cryptorchid testis, hydrocele, hernia, renal dysplasia, horseshoe kidneys, and agenesis of prostate could be associated malformations.\(^4\)

Delay of diagnostic and therapeutic measures resulting in male gender assignment has led to severe psychological and anatomic problems because of difficulties in constructing functioning phallus.\(^9\) In contrast, it is easier to establish normal female appearance. However, no reports concerning long-term physiological and psychological result sexist in immediately gender reassigned patients.

There is a consensus that when the patients with PA are diagnosed in infancy, they should undergo early gender assignment and be raised as girls, despite male karyotype.\(^9\) However, its relevance to the Indian society is still debated. In Indian villages and towns, it would be easier to live as a sexually incompetent male in the society rather than a single unmarried girl.\(^10\)

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.
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