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Cloaca with duplicated phallus – A case report

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ABSTRACT

A child born with atypical genitalia constitutes a medico-social emergency, a multidisciplinary team constituting a pediatric endocrinologist, pediatric surgeon and a psychologist has to be convened. A newborn with abnormal genitalia, referred for sex assignment and further management soon after birth. Baby had atypical genitalia and absent anal opening not fitting into any type of the described Disorders Sexual Develpoment or Anorectal malformation. Child had cloaca and in addition there was male looking phallus with hemiscrotum like structure without gonads.

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XX DSD has female internal genitalia with 46XX karyotype and variable degree of masculinization, anywhere between Prader stages 1–5 [1].

Association of cloacal abnormality with XX DSD with or without accessory urethra is known but very uncommon and only a few cases are reported in the literature [2,3].

We report a rare case of persistent cloaca with duplicated phallus like structure by the side of genitalia with accessory urethra extending up to the tip of duplicated phallus.

1. Case report

A full term neonate presented with atypical genitalia. There was a single perineal opening which resembled female cloaca and in addition there was a duplicated phallus with urethra up to tip, with hemiscrotum like pad of fat giving appearance of male genitalia (Fig. 1). Bilateral gonads were not palpable.

On day 4 of life, after initial VACTERL workup cloacoscopy and diverting transverse colostomy was done in view of a long common channel of 3.5 cm. Child was investigated at 3 months of age. The karyotype was 46 XX.

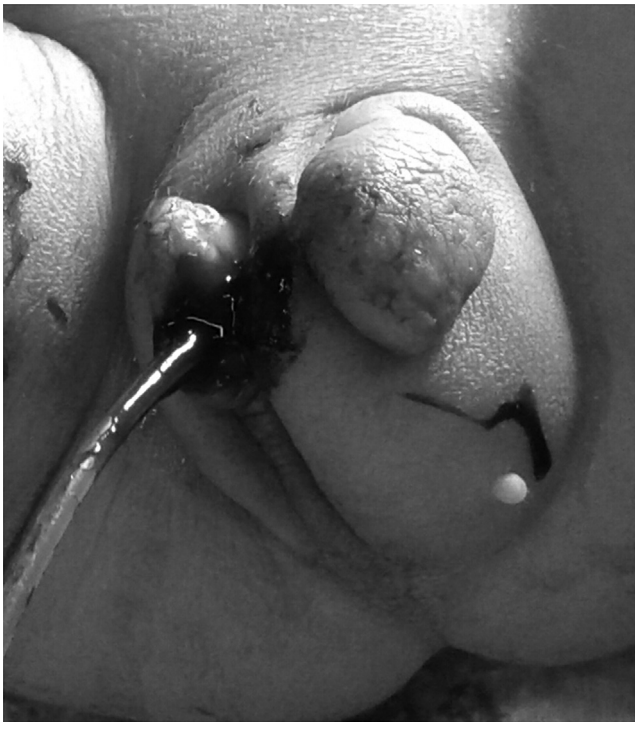

Fig. 1. Perineum with cloaca and accessory phallus.

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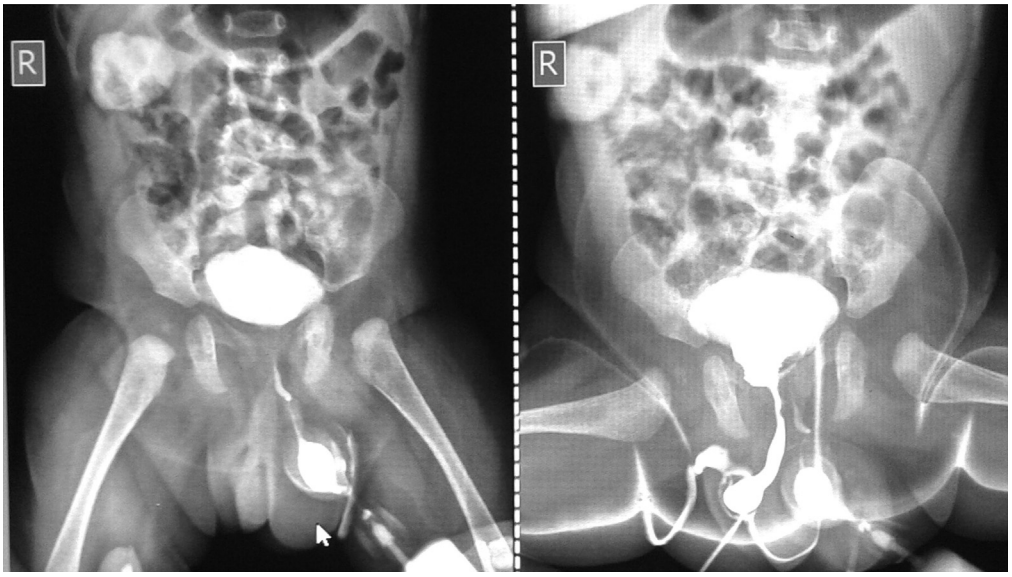

Fig. 2. Contrast study through the two urethral openings with arrow pointing to course of urethra.

Internal organs were female, uterus and ovaries were normal, External genitalia–duplicated accessory phallus has rudimentary hypo plastic corpora, glans was ill developed, hemiscrotum like tissue has only pad of fat with no gonads palpable.

Female looking genitalia on medial aspect of duplicated phallus were like typical long channel cloaca. Contrast study revealed accessory rudimentary phallic urethra draining in bladder (Fig. 2).

Decision was taken to excise male looking genitalia as female sex was assigned.

Accessory phalus was dissected upto symphysis, urethra in it transfixed and divided as high as possible and hemiscrotum like pad of fat excised (Fig. 3).

The result was almost normal looking female genitalia with cloacal maformation (Fig. 4). Histopathology revealed atrophic corpora (Fig. 5). Cloaca was repaired by initially doing high sigmoid divided stoma and later posterior saggital anorectoplasty. The child had no other associated renal or spinal anomaly.

Child is now 3 years old on follow up for kidney status, continent for unine and on enemas regimen for bowel management.

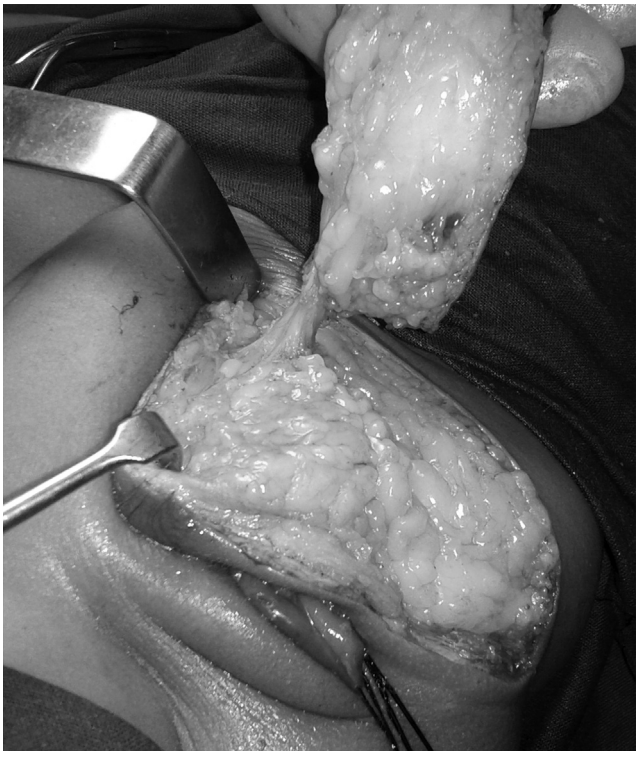

Fig. 3. Intraoperative photo of fat pad below accessory phallus.

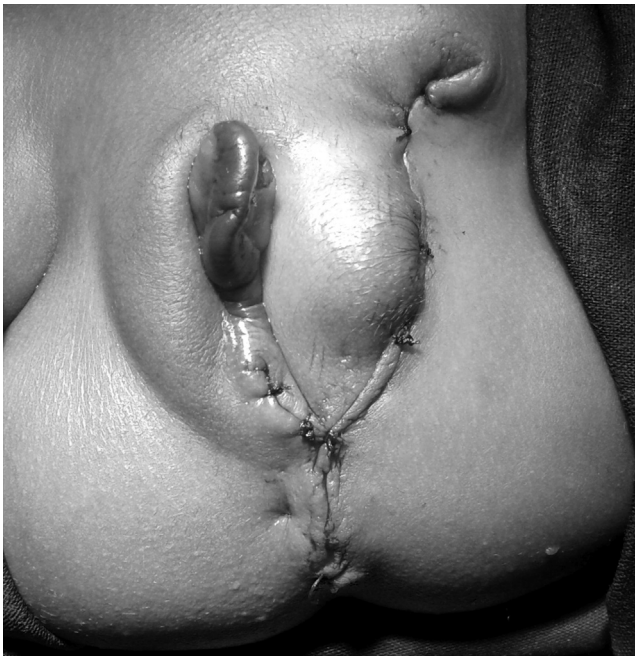

Fig. 4. Reconstructed labia majora.

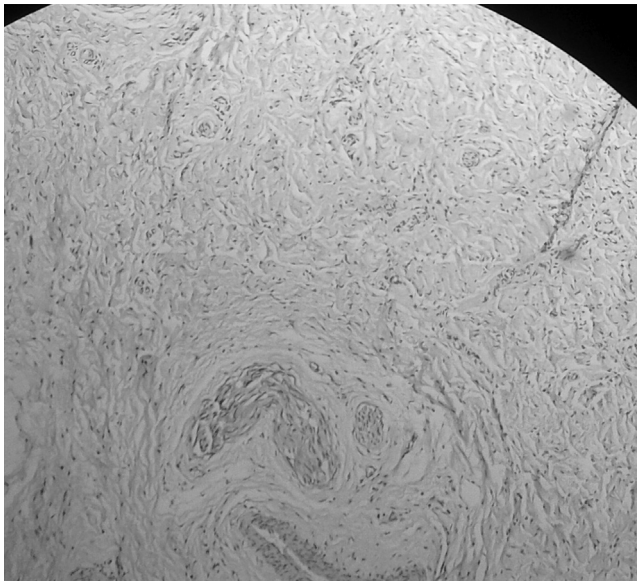

Fig. 5. HPE showing atrophic corpora.

2. Discussion

Isolated cloacal malformations as well as XX DSDS are well understood entities. Association of Cloaca with XX DSD is also known though rare. Both the anomalies have varied spectrum of severity along with various permutations when they occur together. Accessory phallic urethra has been documented in this association but presence of duplicated phallus with urethra and hemiscrotum like tissue cannot be explained [4–6]. Histopthologic demonstration of corpora confirmed that it was phallus and not just skin tag. We are not able to justify association of male looking genitalia and female looking genitalia together. Even though we are applying theory of abortive twinning or caudal duplication it is difficult to explain isolated incomplete duplication only of genitalia [7,8].

Inspite of advances in knowledge, science and technology, few anomalies cannot be hypothesized as regards their embryogenesis. It won't be wrong to say that nature defeats science.

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