Caudal Duplication

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Abstract
A two year old girl was admitted with an abnormal mass in the perineum, which had two rudimentary feet between two normally functioning lower limbs. She had two introituses, one on each side of the mass, from the right she passed urine and rarely stools and from the left she passed only stools. She was continent for urine and stools. MRI revealed a bifid spine below L1 and a bifid sacrum, duplicated colon, single right kidney, absent mullerian structures and a solid mass with incompletely formed feet with cartilaginous structures.

Introduction
Caudal duplication is a very rare entity in which structures derived from the embryonic cloaca and notochord are duplicated to various extent. The term encompasses a spectrum and often is quoted as one type of incomplete separation of monoovular twins and can be considered to be a rare type of conjoined twinning. It is associated with other congenital malformations of the genitourinary and gastrointestinal tract. Dominguez et al introduced the term caudal duplication syndrome for multiple anomalies and duplication of the distal organs derived from the hindgut, neural tube and adjacent mesoderm.

Case Report
A 2 year old girl was admitted with an abnormal mass in the perineum which had two rudimentary feet between two normally functioning lower limbs. She had two introituses, one on each side of the mass. From the right introitus she passed urine and rarely stools, from the left introitus she passed only stools. She was continent for urine and stools. MRI revealed a bifid spine below L1, a bifid sacrum, duplicated colon, single right kidney, absent mullerian structures and a solid mass with incompletely formed feet with cartilaginous structures. Patient underwent staged

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Fig. 1: A 2 year old female with abnormal mass in the perineum with two rudimentary feet between two normally functioning lower limbs. She had two introituses, one on each side of the mass. From the right she passed urine and rarely stools and from the left she passed only stools.
excision of the mass with left colon resection and external genitalia reconstruction.

Pathological Findings

Gross: Received specimen of a triangular non ulcerated skin flap with underlying adipose tissue. Protruding from the skin flap were two malformed feet like structures.

Microscopy: Sections reveal keratinized epidermis, underlying dermis, subcutaneous tissue, muscle and bony tissue showing no e/o any significant pathology.

Discussion

Caudal duplication is a rare type of conjoined twinning, in which structures derived from the embryonic cloaca and notochord are duplicated to various extent. The term encompasses a spectrum and is often quoted as one type of incomplete separation of monoovular twins.

It is associated with other congenital malformations of the genitourinary and gastrointestinal tract. Dominguez et al introduced the term caudal duplication syndrome for multiple anomalies and duplication of the distal organs derived from the hindgut, neural tube and adjacent mesoderm.

Aetio-pathogenesis

The pathogenesis is unclear. Polytopic primary developmental field defect or a disruption sequence or somatic or germ line mutations in certain developmental genes could be involved. The insult to the caudal cell mass and hindgut occurs at approximately 23rd-25th day of gestation. Partial or complete duplication of the organizing centre within a single embryonic disc may increase the risk of mesodermal insufficiency and thus account for the failure of complete development of the cloacal membrane and consequent exstrophy or other aberrations.

Our patient was a 2 year old female, literature reports occurrence in a 19 weeks female foetus, a 21 weeks male foetus, a 1 month infant, and even in an adult a 23 year old male.

Our patient had an abnormal perineal mass with two rudimentary feet, the other associated anomalies were two introituses, one on each side of the mass, from the right side she passed urine and rarely stools, from the left side she passed only stools. She was continent for urine and stools. She also had a bifid spine below L1 and a bifid sacrum, duplicated colon, single right kidney, absent mullerian structures and a double vulva. Other reports in the literature, include duplication of the distal spine from L4, left double ureter, duplication of vagina and cervix.
and distal colon, cloacal extrophy and demised co twin with lower abdominal wall defect, extended intestinal tract, absent external genitalia and imperforate anus as well as a case of mucosa lined left hemiperineal defect associated with penoscrotal hypospadias, penoscrotal transposition, and an overhanging caudal skin, covered by a soft tissue flap, resembling a caudal appendage. A caudal duplication that had ruptured in utero through the hemiperineum, could explain the anomaly. The adult case in the literature had diphallus and anorectal duplication. Yet another case of six lower extremities, double external male genitalia, mega bladder, three dilated ureters, polycystic-right and left kidney has been reported. An extra lower limb with 14 digits attached to an accessory parasitic pelvis in the middle suprapubic area, has also been reported, as well as two well formed lower limbs and pelvis with extra male genitalia attached to the epigastrium, attached to the thorax xiphisternum, with exomphalous major and talipes equino varus as well as omphalocoele, secondary pelvis with two limbs, accessory pelvis, kidney and two bladders.

In our case, a staged excision of the mass, left colon resection and external genitalia reconstruction was done.

References

SALE OF INFANT COUGH MEDICINES SPLUTTERS TO A HALT
The UK Medicine and Healthcare products Regulatory Agency (MHRA) announced last week that over-the-counter cough medicines are no longer suitable for children younger than 2 year. A selection of antihistamines, anti-tussives, expectorants, and decongestants found in cough medicines are associated with a risk of serious adverse events caused by dosing errors in children younger than 2 years due to their small size.

The fact that the UK and USA are still deliberating whether these medicines are suitable for older children is remarkable, considering that the January, 2008, Cochrane Review of randomised controlled trials spanning the past 40 years, concluded that “there is no good evidence for or against the effectiveness of over-the-counter medicines in acute cough”. The MHRA calls itself a leading contributor in improving the availability of medicines suitable for children, but it has disappointingly failed to act in this case.

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