CAUDAL DUPLICATION SYNDROME WITH CAUDAL REGRESSION SYNDROME

Liyakat Ali Chowdhury, Suadipta Saha, Surajit Das, Samiran Samanta

1 Department of Radiodiagnosis, Institute of Post-Graduate Medical Education and Research and Seth Sukhlal Karnani Memorial (IPGME &R and SSKM) Hospital, Kolkata, West Bengal, India
2 Department of Radiodiagnosis, Midnapur Medical College and Hospital, Midnapur, India.


ABSTRACT

Caudal Duplication Syndrome (CDS) is a rare congenital anomaly and has an association between malformations and duplications of the gastrointestinal and genitourinary systems and neural tube defects. We report a rare case of caudal duplication syndrome, a disorder associating partial or complete duplication of the spine or spinal cord and of other caudal structures, such as urogenital tract and variable degrees of neurological dysfunction. Also associated with caudal regression in the form of sacro-coccygeal agenesis.

Key Words: Rachischisis, Myelocystocele, Dorsal dermal sinus, Patent urachus.

Introduction

The child was delivered uneventfully. There was no history of congenital abnormality in the family. The mother did not experience any illnesses during pregnancy. There was no history of exposure to teratogenic or mutagenic agents during pregnancy. The patient had a birth weight of 2.68 Kg. The child cried normally after birth. On physical examination there were two vulvae consisting of three labia majorae, the central one dividing two independent parallel vaginal introitus, each with a pair of labia minora, a clitoris and a central urinary meatus.

X-ray lumbar sacral spine (Fig. 1) revealed duplication of vertebral body from L-1 level downward. Investigation of the spine was done with magnetic resonance imaging (MRI), which disclosed complex malformations of the thoracic and lumbar sacral spine. A T-12 hemivertebra was present. From L-1 level there was complete duplication of the vertebral bodies (Fig. 2) extending down along with rachischisis (Fig. 7) and terminal myelocystocele (Fig. 4). The sacral and coccygeal vertebrae were absent. The spinal cord was duplicated from level T-12 downwards. A fluid filled sinus tract extending from the spinal...
canal and ending in the subcutaneous tissue of the back, may represent a variant of dorsal dermal sinus (Fig. 5). Double urinary bladder with patent urachus and covered extrophy (Fig. 4), double uterus, vagina and rectum (Fig. 6), and umbilical hernia were visualised. There was sacral agenesis. Right kidney measured 50.3 mm while left kidney measured 36.6 mm. Right kidney was malrotated as well.
Discussion

Caudal duplication syndrome (CDS) was described as such for the first time by Dominguez et al in 1993 even though Ravitch et al reviewed the pathology in 1953 by collecting 20 different cases of colon duplication and genital and urinary organ anomalies since the year 1876, from which at least 12 were consistent with the requirements of caudal duplication. Dominguez et al in 1993 defined CDS as the association between malformations and duplications of the gastrointestinal and genitourinary systems and neural tube defects. Spinal and spinal cord duplicity (diasplasia) malformations span a wide spectrum of anomalies, ranging from a simple fibrous band splitting the cord in two halves to complete duplication of the spine and spinal cord. They are usually associated with other systemic malformations, including duplication of vascular structures, of the distal gastrointestinal and urogenital tracts (as in the present case), and possibly limb malformations. The term caudal duplication syndrome has been applied to those instances. Pang et al advanced a unified theory for the spinal cord duplication disorders, suggesting that all result from abnormal adherence between ectoderm and endoderm. In the view of Dominguez et al, these anomalies originate from damage to the mass formed by caudal cells and posterior gut at approximately 25 days of pregnancy. Pang et al classified spinal cord duplication anomalies into types I and II. The first is characterized by two hemicords, each contained within its own dural sac, and separated by an osteocartilaginous septum. Type II is defined by two hemicords in the same dural sac, separated by a fibrous septum. The case we report may be classified as type I. Most cases present with severe neurological impairment, although a few patients with mild or absent neurological dysfunction are on record.

Conclusion

Skeletal, gastrointestinal and genitourinary tract abnormality should be sought for in a case of caudal duplication syndrome with caudal regression syndrome. It is one of the rarest cases with practically no differentials.

References


