Split Notochord Syndrome With Neuroenteric Fistula, A Rare Malformation

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Split Notochord Syndrome With Neuroenteric Fistula, A Rare Malformation

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Abstract

Split notochord syndrome (SNS) with neuroenteric fistula (NEF) is a very rare congenital malformation involving spinal cord, vertebral column and gastrointestinal tract (GIT). A 5-hour old male newborn was presented with imperforate anus and thoraco-lumbar myelomeningocele (MMC). On examination of the MMC an intestinal loop in the configuration of stoma was found in the center of MMC. The lesion was identified as split notochord syndrome with NEF. Cranial ultrasound revealed hydrocephalus. A multidisciplinary approach was advised but parents refused treatment and left against medical advice.

Introduction

SNS is a very rare congenital malformation of central nervous system involving the brain, spinal cord and vertebral column. The scarcity of the malformation can be appraised by the fact that less than 40 cases of SNS have been reported in literature. [1] In about half of the cases of SNS the malformation is also involving the GIT in the form of dorsal NEF. In a few number of cases it is also associated with other anomalies like imperforate anus, vesical extrophy, hypospadias, and so on. [2][3]

We present a case of SNS with dorsal NEF and imperforate anus. The extreme rarity of the condition appeals for its repeat reporting.

Case Report(s)

A 5-hour old newborn (male) was received in the nursery emergency department of our institution with complaints of imperforate anus and a lump at the thoraco-lumbar region. The baby was a product of consanguineous parents and delivered by spontaneous vaginal delivery, at term, in a village supervised by lady health visitor. The weight of the baby was 2.5kg and was active. The vitals of the baby at the time of presentation were normal. On examination the baby had no anal pit or visible meconium in the region of perineum and meatus. The spinal lump was a myelomeningocele having an intestinal loop in the configuration of stoma through which meconium was coming out. [Image 1]

Patient was admitted as a case of SNS with NEF and imperforate anus. Initial management was started in the form of oxygen inhalation, IV cannula passed, parental antibiotics and fluids started, injection Vitamin K given and anti-tetanus toxoid injected intramuscular. Cranial ultrasound of the patient revealed hydrocephalus. Abdominal radiograph confirmed the split spine. [Image 2]

Patient was further discussed with pediatric neurosurgeon and a multidisciplinary approach was planned for the patient. Parents were counseled about the disease and its outcome and our planning for the operation. The parents refused any advised treatment and left the hospital against medical advice.

Discussion

SNS is a very rare congenital malformation and involves central nervous system, vertebral column and sometimes GIT as well. It is thought to be a result of maldevelopment of notochord, neuroenteric canal and paraxial mesoderm; but the exact etiology is still unclaimed. [1]

SNS is very scarce condition and is associated with dorsal NEF in about 50% of cases. In about more than 1/3rd of cases, SNS is also associated with ARM especially imperforate anus. The other associated anomalies that have been reported are hydrocephalus with Arnold-chiari malformation, malrotation, congenital short colon, colonic duplication, bladder extrophy and other urogenital anomalies. [2][3]

The anomaly is usually associated with anterior and posterior clefts of the spine especially at the thoraco-lumbar region through which the intestinal fistula appears. Some case reports described the complete GIT evisceration through the defect in the vertebral column. The level of NEF that have been reported is small intestine, cecum, colon and rectum. The NEF of colon and rectum is mostly observed in the previously reported cases. [4][5]

In our case the SNS was associated with MMC, NEF, hydrocephalus and imperforate anus. We tried to incorporate pediatric neurosurgeon in the management of this complex group of anomalies and our planning was to separate the NEF, making of
colostomy, and repair of MMC, but the parents refused the surgery and left us against medical advice. The patient never returned to us.

Conclusion

SNS associated with NEF and other congenital anomalies is a very rare and fatal disease with very high morbidity and mortality. A multidisciplinary approach is recommended involving, pediatric surgeon, pediatric neurosurgeon, physiotherapist and social worker for the management of disabilities associated with this anomaly.

References

Illustrations

Illustration 1

Image showing gut loops in the meningomyelocele.

Illustration 2

Radiograph showing split spine with bowel gas shadows inside.
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