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CASE REPORT

Split Notochord Syndrome With Dorsal Enteric Fistula In A Male Neonate: A Case Report

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ABSTRACT

Split notochord syndrome (SNS) is an extremely rare congenital anomaly and 33 such cases have been reported in literature, till date. Of these 33 cases of SNS; 20 presented with associated dorsal enteric fistula. We are reporting here a 2.5 kg male neonate, born normally at hospital, to a 24 yrs old Gravida II Para I mother. Clinical examination and investigations revealed SNS associated with hydrocephalus (Arnold Chiari malformation), thoracic meningomyelocele and dorsal enteric fistula just below meningomyelocele. He also had associated imperforate anus, reducible right inguinal hernia and penoscrotal transposition. Staged correction of the disease was planned. Right inguinal herniotomy and sigmoid loop colostomy was done on 14th day of age as a first stage of the operation. His father was reluctant for further operation at present therefore he was discharged on request after a week of operation. The reason for reporting the case is its rarity.

Key Words: Anorectal malformation ,Dorsal enteric fistula ,Neural tube defects ,Spinal dysraphism ,Split notochord syndrome

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Introduction

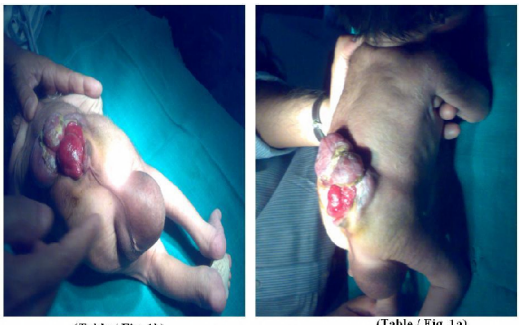
Split notochord syndrome is an extremely rare congenital malformation associated with anomalies of the vertebral column, gastrointestinal tract and central nervous system [1],[2],[3]. Twenty cases of SNS associated with dorsal enteric fistula have been reported in literature till date [10]. Present report describes a newborn male of SNS associated with hydrocephalus, thoracic meningomyelocele, dorsal enteric (colonic) fistula, imperforate anus, right inguinal hernia and penoscrotal transposition.

Case Report

One day old, 2.5 kg, male child was admitted in the department of paediatric surgery with multiple congenital anomalies. He was delivered normally at district hospital to a Gravida II Para I, 24 years old mother on April 30, 2009. The pregnancy & hospital delivery were uneventful. There was no history of infection, intake of any teratogens, drugs or exposure to radiation etc in antenatal period. Clinical examination revealed hydrocephalus, thoracic meningomyelocele, dorsal enteric fistula just below the meningomyelocele and fistula was draining faeces There was associated reducible right inguinal hernia, imperforate anus, and transposition of the scrotum[Table/Fig 1] (Table / fig 1a, 1b & 1c). He is moving his both lower limbs normally and clinically no neurological deficit detected. He was also passing urine well in stream.

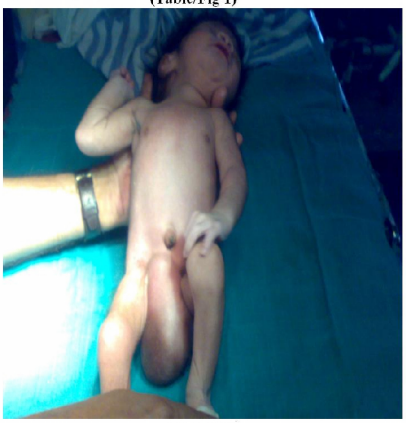
Plain x ray of spine suggests complete splitting of lumber vertebrae and sacrum and it also shows gas filled bowel loops in the right inguino-scrotal area (right inguinal hernia) [Table/Fig 2]. Ultrasonography of head showed hydrocephalus and Arnold Chiari malformation II, which was also confirmed on computed tomography (CT) scan head. CT

scan of abdomen and spine showed complete splitting of all lumber vertebrae and sacrum with complete duplication of notochord as well [Table/Fig 3]. The kidney, liver, spleen etc were all in normal position. Fistulography was done through dorsal enteric stoma to delineate the fistula which confirmed the communication with the colon without any connection to the bladder, etc. The dye containing large intestine was also seen in the inguino scrotal area on the right side [Table/Fig 4] (Table / fig 4a, 4b). This dye study films also very well delineates the complete splitting of the lumber vertebrae and the sacrum. We planned for correction of the disease in stages. As first stage; right inguinal herniotomy and diverting sigmoid loop colostomy was done on 14th day of age. He developed para stomal herniation of bowel on 4th postoperative, needed operative intervention and there after he did well. We planned for ventriculo-peritoneal shunt and meningomyelocele excision and repair as second stage of operation before discharge but parents were reluctant for further operation at present so he was discharged on request after a week.



(Table / Fig 1b) (Table / Fig 1a)

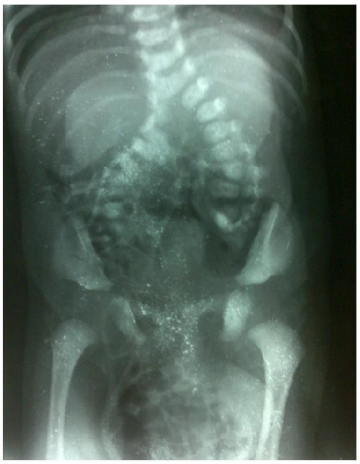
(Table/Fig 1)



(Table/Fig 1c)

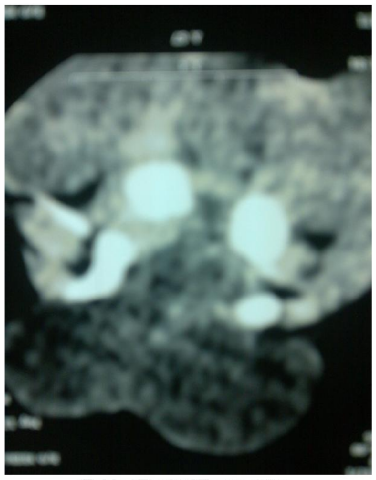
(Table / Fig 1a, 1b) Clinical photograph of patient (Prone & lateral view) Showing: Meningomyelocele, dorsal enteric fistula, imperforate anus & right inguinal hernia

(Table / Fig 1c)Clinical photograph of patient (Supine view) Showing: Large right inguinal hernia

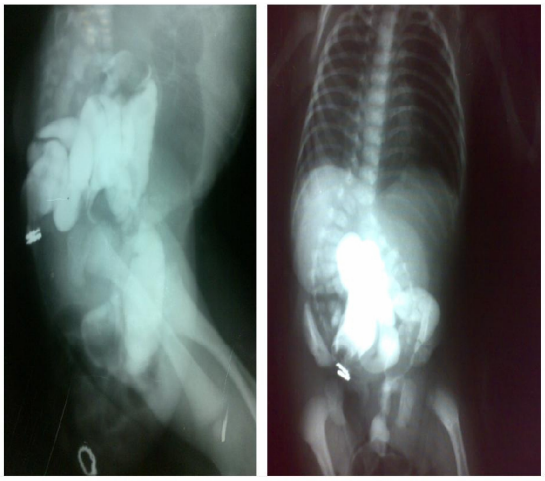


(Table / Fig 2) Plain X ray spine

Showing: Complete splitting of lumber vertebrae and sacrum and gas filled bowel loops in right inguino-scrotal area



(Table / Fig 3) CT scan spine
Showing: Complete splitting of lumber vertebra, spinal cord with spinal canal and menigomyelocele



(Table / Fig 4a) (Table/Fig 4) (Table / Fig 4b)

(Table / Fig 4a & 4b) fistulography // Dye study of colon (lateral and AP view)
Showing: Dorsal enteric fistulous connection with large bowel and part of colon also seen in right inguino-scrotal area and complete splitting of lumber vertebrae and sacrum also seen

Photographs: All Photographs displayed in this article were obtained after informed consent by the parents of the child.

Discussion

Complete cleft of the vertebral column associated with gastrointestinal tract and central nervous system anomalies is known as split notochord syndrome and is an extremely rare form of spinal dysraphism [1],[2],[3]. The name SNS was proposed by Bentley and Smith in 1960 and is also known as posterior spina bifida, combined spina bifida, neurenteric fistula, and dorsal enteric fistula [3]. Jesus LS and Franca CG (2004) were able to search out only 25 cases of SNS in literature [3]. Internet search revealed 8 more case report of SNS in literature making total of 33 till July 2009, excluding present case [3], ,[4],[5],[6],[7],[8,[9],[10]. Only one case was discovered in an adult and others were in newborn and young children and reported both in males as well in females [1],[2],[3],[4],[5],[6],[7],[8,[9],[10],[11].

SNS is frequently associated with vertebral anomalies (anterior and posterior spina bifida, butterfly vertebrae, complete duplication), central nervous system anomalies (meningocele, meningomyelocele, hydrocephalus, neurenteric cysts, duplicated spine or spinal canal sometimes with duplicated spinal cords), gastrointestinal tract anomalies (dorsal enteric fistula, imperforate anus, diverticulum, enteric cysts, atresia, intestinal duplication), and genitourinary system anomalies (penoscrotal transposition, epispadias, exstrophy of bladder, etc) [2],[3],[4], [7].

Twenty cases of SNS associated with dorsal enteric fistula have been described in the literature [1], [3], [4], [10], [12]. Nine of 33 cases of SNS also had associated imperforate anus / anorectal malformation, excluding our case [3,] [4], [10]. The location of the intestinal fistula vary from case to case and may be found in the distal ileum, caecum, or colon but most frequently located in large intestine [3], [4], [12]. Our case also had imperforate anus and the location of dorsal enteric fistula was in the large intestine.

Antenatal sonographic findings depend on the lesion and the associated anomalies and may suggest prenatal diagnosis of split notochord syndrome in some cases [2, 4, 13]. Pre-operative investigations are needed to identify and delineate extend of the disease and the system involved for the particular case. It includes plain x–ray of the spine, ultrasonography and CT scan of the abdomen, head and spine. A magnetic resonance imaging study may be needed for further evaluation of the case. Other investigations like fistulography; MCU, etc are needed in some case depending upon the need [3],[4],[5],[6], [10].

Management of a case of SNS associated with dorsal enteric fistula vary from case to case as the associated anomalies and system involved varies; therefore management must be

individualised. It is also considered that staging procedure is needed for proper correction of the anomalies [3], [4], [10], [14].

In our case we planned for correction of the disease in stages as; (a) sigmoid colostomy and right inguinal herniotomy, (b) ventriculo-peritoneal shunt for hydrocephalus, meningomyelocele excision and repair, (c) disconnection of the dorsal enteric fistula, (d) definitive procedure for anorectal malformation (imperforate anus), followed by colostomy closure and (e) correction of penoscrotal transposition. The first stage of operation; sigmoid colostomy and right inguinal herniotomy was done in our case on 14^{th} day of age. As the parents were very much reluctant for more operation at present so he was discharged on request.

Few survivals have been reported of cases with SNS, but the over all prognosis of a patient with SNS depends on the associated anomalies and extend of the lesions [2],[3],[4], [10], [14].

Consent

All Photographs displayed in this article were obtained after informed consent by the parents of the child.

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