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Short Communication

AN EXPERIENCE WITH FOUR CASES OF SPLIT NOTOCHORD SYNDROME AND REVIEW OF LITERATURE

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ABSTRACT:

Objectives: To evaluate the rarity of split notochord syndrome, to access frequency of various associated anomalies and to find out compatibility of this condition with survival.

Design: All those cases who presented with vertebral abnormalities and those who presented with spina bifida associated with gastrointestinal and urological abbarasion were further investigated for possible association of split notochord syndrome.

Place and duration: The study was conducted at the department of paediatric surgery the Children Hospital Pakistan Institute of Medical Sciences from 1987-2001.

Methods: All those children and aborted fetuses who had gross spinal cord defect, accessory limb, or teratoma like lesion associated with mylomeningocele or those who had gross duplication of pelvic genitourinary organs were further assessed by detailed examination of the local lesions and further radiological, contrast and histopathological studies.

Results: We could identify four such cases, each had completely different anatomical presentation. One was an aborted fetus of 24 weeks gestation. Another child died soon after the delivery. One had pelvic skeletal and organs duplication with enteric fistulae, double exstrophy of bladder, double penis, and split lumbosacral spine, and the fourth case had complete duplication of external genitourinary organs in the form of fully formed double vulvae and split spinal column in the region of sacrum and coccyx.

Conclusion: Split notochord syndrome is perhaps rare abnormality, but often it may not be identified especially if it is not specifically looked for. Fifty percent of our cases were either aborted or still-born, therefore to assess the actual incidence of split notochord syndrome it is suggested that study should be conducted by antenatal ultrasounds and all fetuses who show spinal column defects and aborted should be examined with detailed dissection and histopathology analysis of the tissues.

KEYWORDS: Split notochord, Rare occurrence,Variable presentation.

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INTRODUCTION

Split notochord has been defined with a cleft vertebral column (butterfly vertebrae) associated with gastrointestinal tract, central nervous system and genitourinary tract anomalies. It is a rare occurrence and only twenty cases seems to have been described in literature. It is considered to be due to the failure of fusion of lateral ossification centres of the vertebral bodies resulting in protusion of neural and gastrointestinal components. In majority the site of

occurrence is dorsolumbar region, although lumbo-sacral defect can also occur with other abnormalities like imperforate anus, enteric fistula, meningocele etc. A plain X-ray often will show double spine at the site of split. The defects and abnormalities that are associated with split notochord syndrome are so variable that high degree of clinical suspicion is required to avoid lapses in diagnosis of this condition. This variable nature of presentation is very well highlighted in the cases that we are reporting in this article.

CASE I:

On ultrasound at 24 weeks of gestation a fetus was noted to have a large dorsal spinal defect, it had solid and cystic eomponents with meningocele, the solid component were presumed to be a teratoma associated with mylomeningocele. Parents were counseled and explained the incompatible nature of pathology with unlikely useful survival. With their consent therapeutic abortion was performed. On examination products of abortion placenta and cord were noted to be of normal size, shape and gross appearance. The fetus however had several interesting features. There was a large spinal defect in the dorsal spine with large lobulated thin walled meningomylocele containing cerebrospinal fluid. Adjacent to the defect there were solid components containing brain like neural tissue, there was an area of scalp with full grown hairs. But the most interesting finding was a fully developed eye with complete eyeball, retina, iris, cornea, sclera and eyelashes. Fig. 1. An X-ray of this fetus showed complete duplication of thoracolumbar spine Fig. 2a and 2b. It was more in the nature of split notochord syndrome. On histopathology in addition to grossly identified tissues and organs mentioned above there were also intestinal components in the teratomatous mass.

CASE 2:

Another full term baby was born at home and died on arrival at the hospital. On detailed examination he had dorsolumbar skin covered meningocele mass, with an enteric fistula and a fully developed lower limb like structure arising from the surface of this mass. Fig 3. On further investigation with an X-Ray of the spine double split spine of the lower dorsal and entire lumbar area was noted. On dissection of the lump besides the splitting of the spinal cord there was an occult meningomylocele and a herniating intestinal loop with enteric fistula. This is yet another example of split notochord syndrome with associated organogenisis where parasitic twinning, fetus-in-fetu and teratoma like lesions are difficult to differentiate.

CASE 3:

An eight month old girl who had no symptoms, was brought to us because she had double vulva, on detailed examination she had fully developed perineal genitourinary organs. Fig.4, each vulva possessed anal canal, vagina, labias, urethra and clitoris. All these orifices were fully formed, and she passed urine and faeces from both urogenital organs. The plain X-ray of pelvis and spine revealed split vertebral column and sacrum and coccyx in the direction of each vulva. On further investigations with ultrasound and contrast studies, there were double bladders, double rectum and pelvic colons, independent set of uterus and one ovary on each side. Since she did not have any symptoms, it was decided not to undertake any form of surgical corrections because it might have done more harm than good. It might have resulted in gross functional compromise. On follow up she continues to be asymptomatic and parents have been reassured.

CASE 4:

Newborn baby boy was brought with gross malformation of perineal and genitourinary malformations. On close examination he was seen to have double exstrophy of bladder, double penis, double urethra, an interpositional malformed skin covered mass between these divided organs and an omphalocele. Fig.5. He

had a vestigial enteric fistula on left side just under the penis through which exuding meconium could be seen. A plane X-ray revealed a completely split coccyx, sacrum and first lumbar vertebra with wide pubic symphysial diastasis. Fig 6. On exploration there were two colons upto proximal pelvic colon. On the right side there was anorectal agenesis and on the left side there was perineal fistula. There were double exstrophied bladders both with one ureter, a proximal colostomy and repair of omphalocele was performed. But patient has been lost to follow-up.

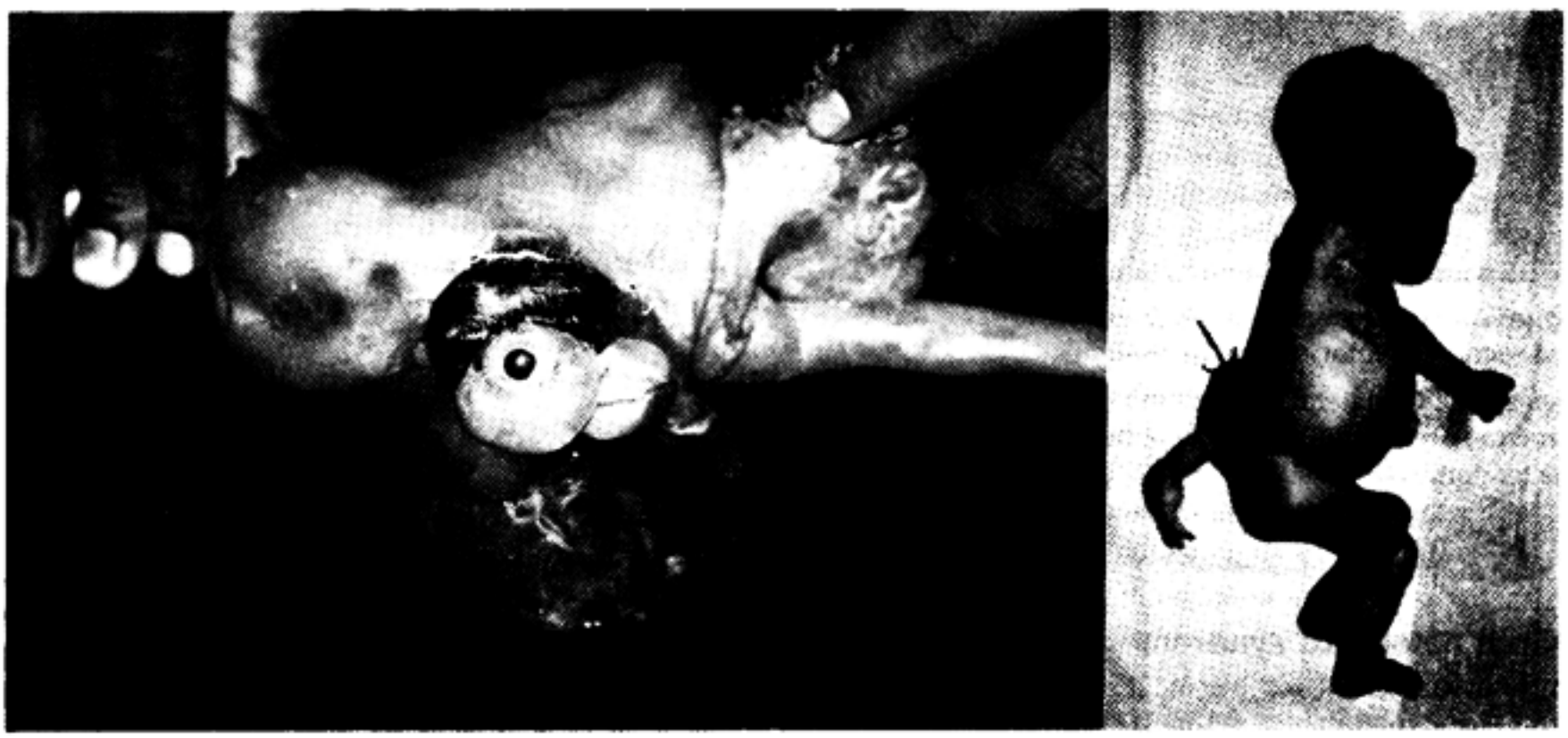

Figure 1: An interesting case of split notochord syndrome in a 24 week gestation fetus, associated with large multilobulated meningomylocele, brain substance and area of scalp and a fully formed eye, with scelra, eyelids, eyelashes, iris, cornea and retinal structures. Head is represented by only an area of scalp with hairs.

Figure 3: Full term baby who died soon after birth with a third lower limb jutting out of lumbosacral mass of covered meningomylocele. An enteric fistula can also be seen (➡) through which the meconium was coming out.

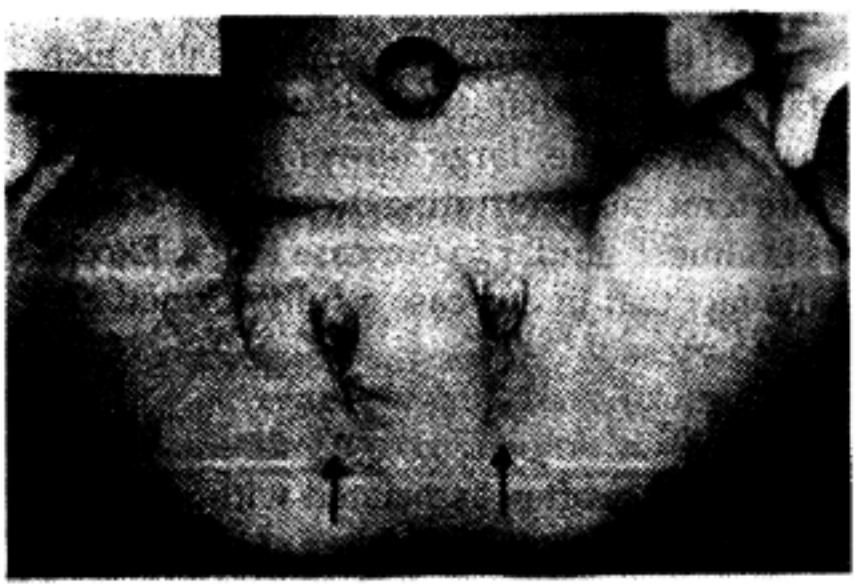

Figure 4: A girl with complete duplication of perineal organs which included urethra, vagina and clitoris. She also had double anal canals, one under each vulva.

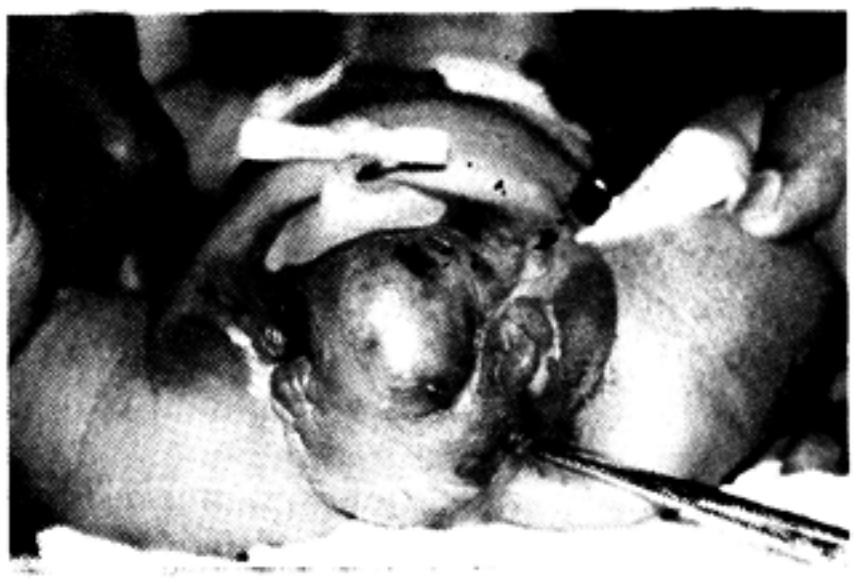

Figure 5: Male child with double exstrophy, double penis, anorectal agenesis on the right side and anocutaneous fistula on the left with omphalocele and interpositional skin fibro-fatty mass.

Figure 2(a): Split spine involving lower thoracic lumber and sacral vertebral column of the fetus in Fig.1

Figure 2(b): On lateral view of the same patient skeletal structures resembling limb skeleton can be seen within the teratomatous mass.

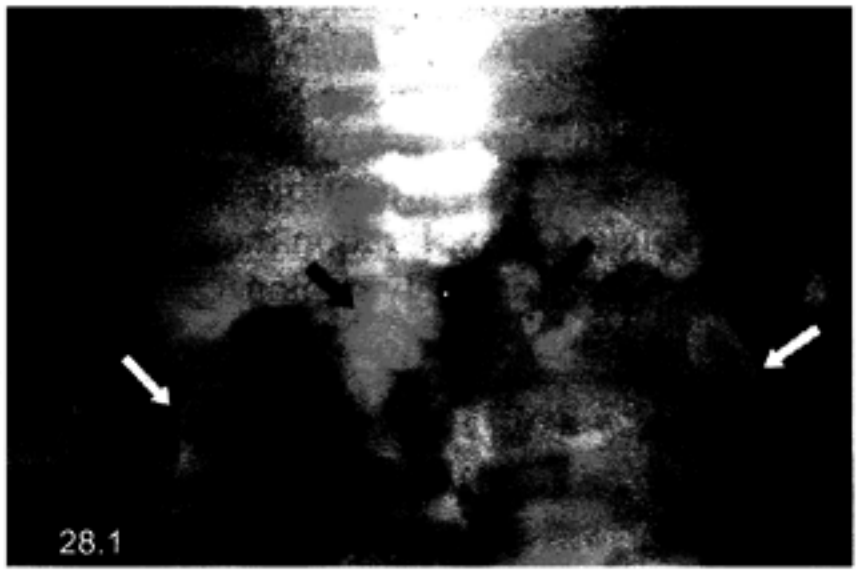

Figure 6: Skeletal X-Ray of he same child as in fig 5 with split spine involving fifth lumbar and entire sacrum (black arrows).

DISCUSSION

Split notochord syndrome is a rare, curious and serious malformation. It is often characterized by a wide defect in the spine and persistent endodermal and ectodermal communication the result of which is often gross spina bifida with associated fistula of the herniated enteric mucosal and genitourinary anomalies.[1,2] Muller et al. suggests that vertebral splitting occur in embryonic period as opposed to defects like spina bifida the period of which may extend to fetal stage.[3] If it is diagnosed antenatally with ultrasound and is associated with hydrominas and gross malformations and abnormalities like teratomatous mass and large meningomylocele, abortion with consent may be the right option,[4] as we did in our case one. In one case report split notochord syndrome was noted with dorsal enteric fistula, meningomylocele and imperforate anus. Our case 4 also had complete anorectal agenesis on right and an enteric fistula on the left duplicated colons.[5] A variant of split notochord syndrome was reported who had a skin covered mass on the back with a portion of intestine which on exploration proved to be a caecal duplication. This report is comparable to our case no. 2 where the defect was skin covered with enteric fistula but in our case there was also a fully developed accessory limb.[6] Two variants of split notochord syndrome have been reported where in one case anorectal agenesis with enteric fistula and a fully formed double spine was noted, this is comparable to Fig 2 of our case one.

In another case a lower limb like structure was present on a lumbosacral circular defect which was skin covered, during surgery the mass contained stomach, duodenum, jejunum, ileum and colon. These structures were covered with peritoneal membrane. Presence of limb in this case is similar to the limb noted in our case two Fig. 3.[7] Another case of split notochord reported from university of Miami, Florida where a girl was noted to have complete spinal canals which contained two spinal cords, it was associated with meningocele, large bowel fistula and a mature teratoma protruding through the defect. This is almost similar to our case Fig. 1. Except for the fact that in our case a fully formed eye was also present.[8]

In reporting these cases we would like to highlight that split notochord syndrome is a rare anomaly and is often associated with different congenital anomalies of other systems, like meningomylocele, enteric, genitourinary and teratoma lesions and accessory organs which point to the conjoined twinning and fetus-in-fetu associations. It is proposed that all spinal defects and lesions of the spine should be ex-

amined in detail to avoid lapses in diagnosis of split notochord syndrome.

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