THE SPLIT NOTOCHORD SYNDROME WITH DORSAL ENTERIC FISTULA, MENINGOMYELOCELE AND IMPERFORATE ANUS*

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SUMMARY: Dindar H, Kanmaz T, Çakmak M, Savaş Ç, Yücesan S. (Department of Pediatric Surgery, Ankara University Faculty of Medicine, Ankara, Turkey). The split notochord syndrome with dorsal enteric fistula, meningomyelocele and imperforate anus. Turk J Pediatr 1999; 41: 147-150.

A male infant was referred to our department because of lumbosacral meningomyelocele, dorsal enteric fistula and imperforate anus. The mother had received a parenteral drug containing estradiol benzoate and progesterone for inducing abortion in the first trimester. She also used an anal pomade containing triamcinolone and lidocaine-HCl during the pregnancy for hemorrhoids.

Sigmoid end colostomy was performed after meningomyelocele repair. On abdominal exploration a wandering spleen was detected but no other anomalies. Two months later, an abdominoperineal pullthrough was performed, and the patient was discharged well after three weeks.

Our case is the sixth that had split notochord syndrome associated with dorsal enteric fistula and imperforate anus. Additionally, penoscrotal transposition and wandering spleen were present in this case. To our knowledge, these associated anomalies have been extremely rare. *Key words: split notochord syndrome, dorsal enteric fistula, anal atresia.*

Split notochord syndrome (SNS) is rare, There are vertebral, central nervous system, and visceral anomalies in various combinations. In most of the reported cases, the cervicothoracic spine is involved. Split notochord syndrome (SNS) of the lumbosacral area is less common. Associated meningomyelocele with a dorsal enteric fistula is interesting in this whole group. The following is a description of the clinical, radiographical and surgical findings of such a case.

Case Report

A male newborn was referred to the Department of Pediatric Surgery at the University of Ankara on the day of birth because of gross anomalies overlying the spine. The gestational age of the newborn had been estimated as 41 weeks and the birth weight 3100 g. The 35-year-old mother received a parenteral drug containing estradiol benzoate and progesterone for inducing abortion in the first

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trimester. She also used an anal pomade containing triamcinolone and lidocaine-HCl during the pregnancy for hemorrhoids. She had no x-ray examinations. Previously, the mother had had one normal child, who was 15 years of age. There was consanguinity between the parents, but no family history of similar anomalies.

Physical examination revealed a large head with wide sutures and large, full fontanels. A meningomyelocele was present in the thoracolumbar region. Caudal to the meningomyelocele, intestinal mucosa protruded and meconium was excreted from the enteric opening (Fig. 1). There was a palpable defect in the lumbar spine and sacrum. Penoscrotal transposition with a small phallus was present. Testes were palpable within the bifid scrotum. The anus was imperforate. There was active movement of the lower extremities.

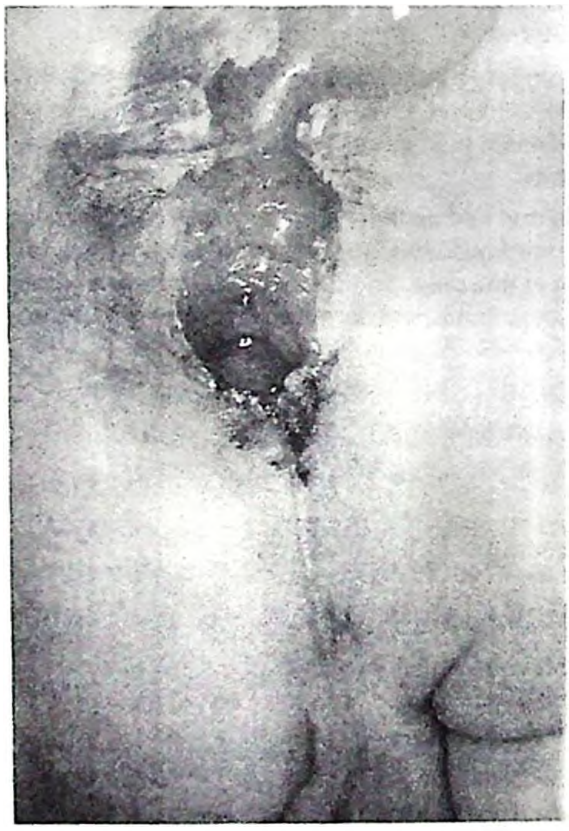

Fig. 1: Intestinal mucosa is protruded and meconium excreted from the enteric opening caudal to the meningomyelocele in the thoracolumbar region.

An x-ray of the spine showed a complete split of the lower lumbar spine and absence of sacrum (Fig. 2). Magnetic resonance images of the spine demonstrated two separate division of the spinal chord at the L2-L4 levels. A 5x3.5x1.5 cm meningomyelocele was seen posterior to the L2 vertebral body. Computed tomographic images showed posterior spina bifida at all levels, thoracolumbar rotoscoliosis and sacral dysgenesis.

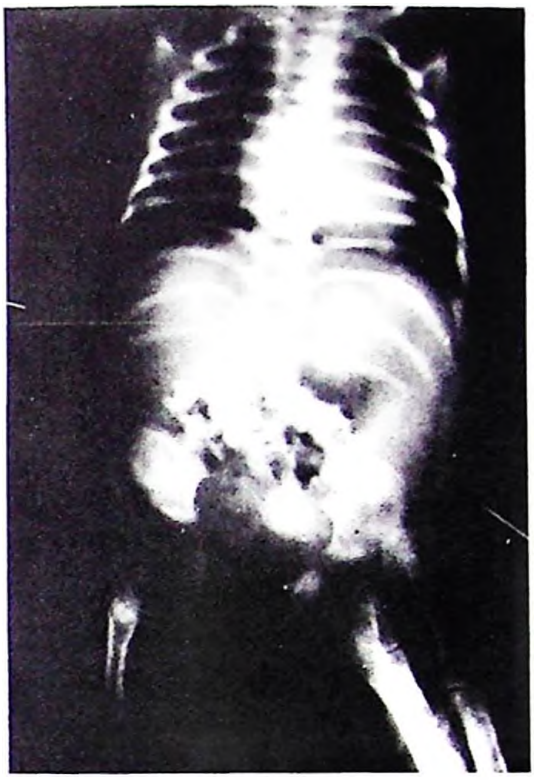

Fig. 2: An x-ray of the spine showed a complete split of the lower lumbar spine and absence of sacrum.

The meningomyelocele was repaired on the first day of life. It was found to communicate with the spinodural canal by means of a small narrow sac. After excision of the neural lesion, the protruded bowel opening was dissected, ligated and placed into the abdomen. After closing the dura, the defect was reinforced with paraspinal fascia and the skin edges approximated over the defect. A laparotomy was then performed, and a wandering spleen (movable spleen because of a lengthened pedicle) was detected on surgical abdominal exploration. The colon ended at the sigmoid level and the rectum was not found. The fistula was between the sigmoid end and neural canal. Sigmoid end colostomy was performed and no other anomalies were found in the abdomen. The postoperative period was uneventful. He was discharged well after one week. Two months later, an abdominoperineal pullthrough was performed for anal atresia. The patient was discharged well after three weeks.

Discussion

Split notochord syndrome (SNS) is an extremely rare anomaly that may be associated with a wide spectrum of developmental anomalies involving skin of the back, spine, central nervous system, and gut. The majority of cases of SNS have involved the spine above the level of the diaphragm. Bentley and Smith[1] were the first to note that an abnormal splitting of the notochord could cause a wide variety of malformations of not only the vertebral bodies and neural tissue but also enteric viscera.

Split notochord syndrome (SNS) of the lumbosacral spine in association with dorsal enteric fistula is a rare phenomenon. To date, only 15 human cases have been reported in the literature[2]. Hoffman et al.[2] described such a case and reviewed the previous cases: of the 15 cases, eight were associated with meningomyelocele, five had an imperforate anus, two were associated with bladder extrophy, and one had a rudimentary extremity extending from the spinal cleft. Another case of SNS was associated with dorsal sinus lipoma (dorsal mesodermal sinus), colonic duplication, annular pancreas, and meconium peritonitis[3].

Several etiologic theories have been proposed, including the persistence of the neuroenteric canal, the occurrence of an ectopic or accessory neuroenteric canal, a division or local redundancy of the notochord, an endodermal-ectodermal adhesion, neural tube rupture caused by oversecretion of fluid, and failure or aberrance of dorsal aortic distribution to the region of the neural folds resulting in prevention of timely neural tube closure[4]. It was shown that a large malformation spectra, especially abdominal wall defects, occur following intraamniotic administration of glucocorticoids in the chick embryo[5]. The mother of this case used an anal pomade containing triamcinolone. This drug might have been absorbed from the rectal veins and in might have affected the fetus. The mother also used a parenteral drug containing estradiol and progesterone for inducing an abortion. This drug might also have affected the patient. Despite these theories, the split notochord syndrome remains complex and poorly understood.

Our case is the sixth one that had SNS associated with dorsal enteric fistula and imperforate anus. Additionally, penoscrotal transposition and wandering spleen were present in this case. To our knowledge, these associated anomalies have not been reported previously. Gupta et al.[6], after reviewing the literature, found very few cases involving the lumbosacral area with very few survivals. The presented patient also had lumbosacral cleft, but was nevertheless alive and healthy. We recommend repair of both meningomyelocele and dorsal enteric fistula as fast as possible to prevent the contamination of the meningocele sac by enteric bacteria.

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