Case Report

Split cord malformation type I distal to segmental myelomeningocele

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ABSTRACT

إن تواجد فلق النخاع الشوكي والقيلة النخاعية السحائية في حالة واحدة من العيوب الخلقية المعروفة. عاده ما يكون مكان فلق النخاع الشوكي أعلى من مكان القيلة ولكن تواجد فلق النخاع الشوكي أسفل القيلة السحائية نادر جداً. في هذا التقرير نستعرض حاله فلق النخاع الشوكي أسفل قيلة نخاعية سحائية مع وجود حركة بسيطة في القدمين. لم يفقد الطفل بعد العملية أي من الحركات التي كانت موجوده قبلها. تظهر هذه الحالة أهمية الإلمام بتزامن حالات فلق النخاع الشوكي مع القيلة النخاعية السحائية لتجنب الأضرار العصبية الممكنة نتيجة عملية إصلاح مثل هذه العيوب الخلقية النادرة.

The coexistence of myelomeningocele (MMC) and split cord malformation (SCM) is a well-known phenomenon. The SCM is usually above or at the level of the MMC. Split cord malformation distal to the MMC is considered to be the rarest form of such a combination. We report a case of SCM (type I) distal to the MMC diagnosed pre-operatively. Repair of the MMC and the SCM were carried out in the same setting.

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Myelomeningocele (MMC) is the most common and most severe congenital anomaly of the CNS that is compatible with life, occurring between the 25th and 27th post ovulatory days.[1] Myelomeningocele can be associated with other dysraphic malformations, with split cord malformation (SCM) being the most common association. The incidence of type I SCM in patients with MMC who had MRI performed ranges from 5-40%.[2] The most common location of SCM is one or 2 levels above the MMC, and is thus hidden from the surgical field during closure of the MMC. Usually, the MMC is repaired in the initial operation, and the SCM is dealt with in a later setting either following the symptoms of tethered cord later in childhood, or after routine MRI. This case highlights the importance of recognizing the rare combination of MMC and distal SCM to avoid inadvertent injury to the distal conus, and avoid the possibility of missing the SCM, which may require an additional operation in the future.

Case Report. A 28-year-old Yemeni female presented to the obstetric service in labor. Obstetric ultrasound showed the fetus to have severe hydrocephalus. A mid-lumbar MMC and a Chiari II malformation were also evident. The baby was delivered via cesarean section. Pre-operative examination of the placode showed a suspicious structure distal to the placode, and the diagnosis of segmental MMC with distal type I SCM was entertained (**Figure 1**). Pre-operative motor assessment showed a slight bilateral foot plantar flexion following plantar stimulation. Rectal tone and anal puckering were present. Intra-operatively it was quite evident that the malformation was indeed a proximal segmental open neural tube defect (ONTD) with a caudal type I SCM containing a cartilaginous midline septum (**Figure 2**). The hemicords reunite just distal to the septum to form a relatively normal looking conus. Distal to the open placode, the right hemicord was neurulated, but the left hemicord remained non-neurulated for a short distance. The cartilaginous septum was resected and the placode was

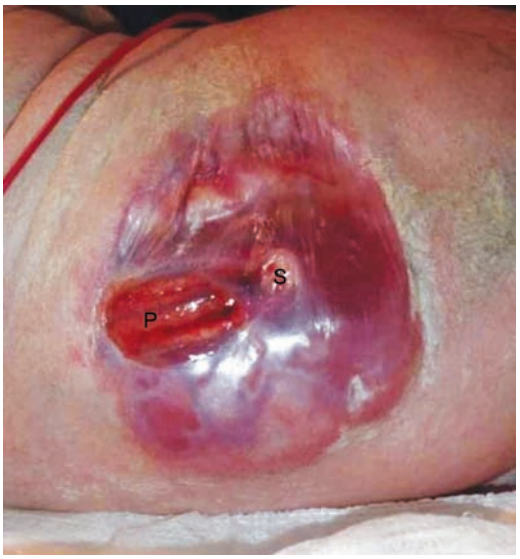

Figure 1 - Preoperative photograph showing the placode (P) and distal spur (S) through the transparent sac.

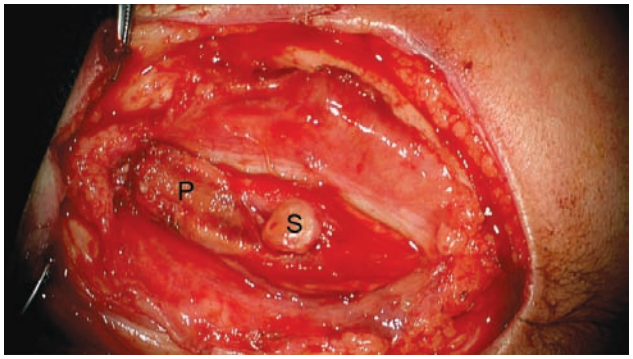

Figure 2 - Intra-operative photograph depicting the placode (P), and the cartilaginous spur (S).

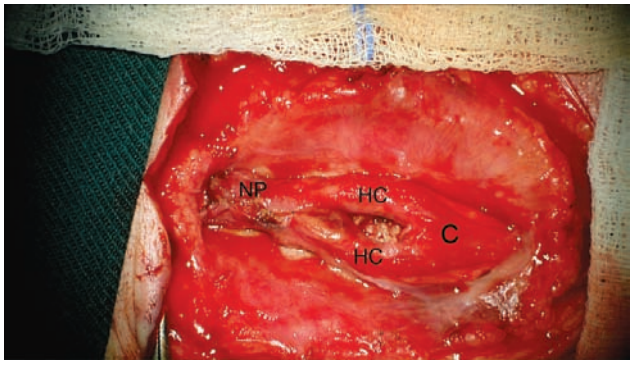

Figure 3 - Intra-operative photograph following neurulation of the neural placode (NP), union of both hemicords (HC), and the distal conus (C).

neurulated using 8/0 Ethilon sutures (**Figure 3**). The dura was closed primarily. A left unilateral skin relaxing incision was needed to approximate the skin over the defect. A ventriculo-peritoneal shunt was inserted in the third week following closure of the defect. The child was examined at 2 months of age and showed preservation of foot and toe flexion with the right side showing more robust movement compared with the left.

Discussion. Up to 40% of SCM cases may have an associated ONTD, affecting either both hemicords, or only one hemicord in the form of hemi-myelomeningocele.[2] According to the unified theory of Pang,[2] all SCMs arise from a common basic embryogenetic error that occurs during gastrulation (formation of trilaminar embryo), an event preceding even primary neurulation. The main event of gastrulation is the formation of the notochord. The primitive knot (Hensen's node) provides pre-notochordal cells, which eventually transform the notochord into a solid cord. The notochord become adherent to the midline, with the aid of adhesion molecules, probably fibronectin. A communication between the ectoderm and the endoderm takes place and a temporary neurenteric canal forms, which heals in 2-3 days. Failure of such a healing and the persistence of such a communication, which can be called the ecto-endomesenchymal tract, is probably the most important error in the embryogenesis of SCMs. At post ovulatory day 28, cells derived from the primitive meninx (the precursors of dura matter) become involved with the ecto-endomesenchymal tract and eventually dictate the formation of type I SCM with double dural sleeves, or type II SCM with a single dural sleeve.[2]

For more than 100 years, different theories on the pathogenesis of MMC have been proposed. The most widely accepted ones are the simple non-closure theory and the reopening theory.[3] It is generally agreed upon that the caudal neuropore closes at days 25-27 and is the last part of the neural tube that undergoes closure.[1] This clearly indicates that the pathogenesis of SCM predates the pathogenesis of MMC. The combination of both pathologies may suggest a common pathogenesis.[3]

In the largest reported series of SCM, Mahapatra[4] did mention the association with MMC, but he did not specify the relation between the level of the split and the placode in detail. Ansari and associates,[5] in a retrospective review of 330 cases of MMC found 33 cases of SCM, 17 were at the level of the placode, 6 proximal, and 10 cases were located distally; one of these cases was associated with hemi-myelomeningocele. Kumar et al[6] reported 16 cases of such a combination, 12 cases of

SCM were proximal and 4 cases were at the level of the MMC, none were found distal to the placode.[6] Erşahin[7] expanded his previous series of SCM from 74 to 131 cases and found 26 cases of MMC, 22 cases were above the placode, 3 distal to the placode, and the only one at the level of the placode was hemi-myelomeningocele. In 20 cases of combined SCM and MMC, Iskandar and associates[8] found the SCM to be located distally to the placode in 2 patients only. Higashida et al[9] reported one case of SCM caudal to the placode repaired in a delayed fashion following closure of the MMC.[9] From the above reports, only 16 cases of SCM distal to the MMC were clearly identified.

In the clinical setting, the SCM is usually identified following the primary repair, either following a routine MRI or later on when the clinical picture of tethered cord syndrome develops. Obtaining MRI examination prior to the MMC repair is not commonly practiced in our community either because of unawareness, lack of facilities, or for financial reasons. Beside these factors, the emergency nature of MMC closure makes obtaining MRI prior to the repair impractical. With such a high prevalence of congenital malformation associated with MMC, pre-repair MRI should be encouraged, whenever possible, to enable a more comprehensive management of such lesions, preferably before the emergence of new neurological deficits, especially when there is a discrepancy in leg function and or hypertrichosis. Although delayed repair of SCM has been shown to be safe, a second operation for SCM near a scarred, previously repaired MMC can be difficult and may cause further neurological deficits.

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Case Reports

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