Case Report

Split Cord Malformation Type 2 with Double Dorsal Lipoma: A Sequela or a Chance

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ABSTRACT

An 11-month-old baby girl, with normal perinatal history, presented with a congenital lumbosacral swelling and a sacral dimple. Imaging revealed a split cord malformation (SCM) type 2 at L1–L3 with a lipomeningocoele extending intradurally and tethering both the hemicords, the conus was noted to be at L4. She underwent excision of the lipoma and detethering of the hemicords. Intraoperatively, the split cord was seen from L1 to L3 with an aberrant median vessel passing between the hemicords. Two lipomas were seen separately attached to each of the hemicords, the lipomas were dissected off the hemicords, and the hemicords were neurulated. The case helps revisit the unified theory proposed by Pang for SCM as well as the theory of premature disjunction in the pathogenesis of lipomeningocoele. Formation of the endomesenchymal tract splits the developing cord into two, whereas the premature detachment of neuroectoderm from the cutaneous ectoderm can lead to lipomeningocoeles. The present case is only the fourth case ever reported of an SCM type 2 with double lipoma. In the case report, we discuss the embryological basis of this condition and surgical nuances of management.

KEYWORDS: Double lipoma, premature disjunction, split cord malformation, unified theory

INTRODUCTION

Our current understanding of the embryological basis of split cord malformations (SCMs) is based on the unified theory proposed by Pang et al.[1,2] Meanwhile, the pathogenesis of lipomeningocoele has been well explained by the premature disjunction of the neural ectoderm from the cutaneous ectoderm. These malformations take place during the stage of primary neurulation. Even though a concatenation of malformations can be speculated from the embryological hypothesis, a potpourri of malformations arising during the same epoch are also found. Co-occurrence of lipomeningocoele with SCMs is a well-known entity.[3] But double lipomeningocoele arising from the split cords is rare, which contradicts the earlier notion.

CASE REPORT

An 11-month-old baby girl, born out of non-consanguineous parentage with normal antenatal and perinatal history presented to our outpatient clinic with a gradually progressive painless swelling over the lower back since birth. Her development assessment for motor and psychosocial development was appropriate for age. Physical examination revealed an ill-defined solitary non-tender swelling measuring about 3 cm × 3 cm with an overlying skin dimple over the lumbosacral region. The child was preserved neurologically appropriate for age. High-resolution computed tomography of the lumbar spine showed an absence of spinous process from L1 to S5 levels. No bony spur or septum was seen...
within the spinal canal. No segmentation anomalies were seen in the lower dorsal and lumbar vertebrae. Magnetic resonance imaging revealed the division of the spinal cord into two from L1 to L3 vertebral body levels. These hemicords were noted to rejoin and form the conus, which was located at the upper border of L4. A subcutaneous lipoma was seen extending intradurally at L1–L3; the lipoma was seen tethering both the hemicords [Figure 1]. Screening ruled out any associated cervical or intracranial pathology.

**Procedure**

Under general anesthesia, with child positioned prone, an incision was made from L1 to L5 [Video 1]. Lipoma was seen extending from the subcutaneous level into the epidural space through a defect in the spinous process of L3. The posterior elements of L1–L5 were noted to be deficient. The dissection of the paraspinal muscles was carried out laterally in the subperiosteally to identify the intact lamina on both sides. The lamina from L1 to L5 was removed and the underlying normal dura above and below the dural entry of the lipoma was identified. Durotomy was performed above and below the lipoma to identify the spinal cord and the nerve roots. The lipoma was detached from the dural edges. The borders of the lipoma were then traced down to the spinal cord. The spinal cord was seen dividing into two hemicords at L1 level and rejoining to form single cord near the conus medullaris, which was located at L3 level. An aberrant median vessel was noted passing through the rostral end of the split. Adhesion of the lipoma to both the hemicords was identified; after meticulously dissecting away the attachments between the nerve roots, the lipoma was detached for the hemicords using fine microsurgical dissection. The hemicords were then neurulated [Figure 2]. The thecal cavity was inspected for any further tethering elements. Filum terminale was cut. Dura closed watertight and wound closed in multiple layers over a subfascial drain. The infant had an otherwise uneventful recovery with no neurologic deficits. Postoperative ultrasound of the urinary bladder revealed no post-void residual urine or hydroureronephrosis.

**Discussion**

Our current understanding of the embryological basis of SCMs is based on the unified theory proposed by Pang et al.[1,2] SCM type 1 refers two hemicords within separate dural sacs with an intervening rigid osseocartilaginous spur, whereas in SCM type 2 the two hemicords, with an intervening nonrigid fibrous median septum, reside in a single dural sac.[1,4,5] The unified

![Figure 1: Preoperative lumbosacral imaging. (A) Sagittal section of MR imaging showing first dorsal lipoma seen attached to the larger right hemicord. (B) Sagittal section of MR imaging showing second dorsal lipoma seen attached to the smaller paramedian left hemicord. (C) Axial section of lumbar MR imaging showing split cord malformation without any bony septum and a dorsal lipoma. (D) 3D reconstruction of the CT lumbosacral spine showing spina bifida involving the L1 to S5 vertebra. No bony spur seen](image)
theory proposes that during gastrulation an adhesion between developing endoderm and ectoderm, rostral to the original neurenteric canal, results in an abnormal fistula connecting the amniotic cavity and the yolk sac called the accessory neurenteric canal. \[1,5-9\] Mesenchymal cells condense around this abnormal accessory midline neurenteric fistula resulting in the endomesenchymal tract (EMT), which then forms the key developmental error in the formation of the SCMs. \[1,5,7,8\] EMT bisects the neural placode and the underlying notochord, thereby inducing the neurulation of the two hemineural plates separately into two independent hemicords. \[1,7,8\] Timing of the development of the EMT in relation to the appearance of meninx primitiva, which contributes to the formation of the dura, adjacent arachnoid, and vertebra, determines the type of SCM. \[8\] If the EMT has condensed after postovulatory Day 30, it will incorporate cells of the meninx primitiva as it develops and will result in dural coverings separately around each hemicord. It also induces osteogenesis of the mesenchyme and gives rise to the osseocartilaginous septum so characteristic of SCM type 1. But if the condensation had occurred before postovulatory Day 21 when the meninx primitiva has not yet developed, it remains as a fibrous septum with the meninx primitiva forming a single dural tube enclosing both dural tubes, the type 2 SCM. \[5,8,10\] The condensation of mesenchyme between postovulatory Days 21 and 30 as well as chaotic mesenchymal condensations can result in composite/mixed SCM as seen in many case reports. \[1,5,8,11-13\] Multiple EMTs (2%) can lead to SCM of different types at different levels. \[3,7\]

The presence of midline lipomas is also attributed to this “pluripotentiality” of mesenchymal cells, which may differentiate into bone, cartilage, vasculature, or fat along the course of the EMT. \[1,10,14\] Lipomas seen in classical SCMs are easily separable from the pial surface of the spinal cord. \[15-17\] In the case presented, herein, however, the lipomas attached to the hemicords were seen extending into the subpial plane. This would be possible only if the lipoma was laid down, while the process of neurulation was still incomplete. Thus, the case reiterates the hypothesis that this might be due to premature disjunction of the neuroectoderm from the cutaneous ectoderm (primary neurulation which occurs between postovulatory day 22 and day 28), resulting in mesenchymal cells entering central canal and coming in contact with the open neural tube, which induces the formation of a intramedullary lipoma on the hemicords in which disjunction occurred. \[5,7,10,15,17-21\]
The mesenchyme is induced by the dorsal surface of the closing neural tube to form fat, whereas on the ventral surface meninx primitiva is induced to form meninges. This also explains the presence of a dural defect around the point dural entry of the lipoma which is the junction between meninges and fat at the neural ridge.[14,17] The ectoderm then heals over the lipoma resulting in an intact skin covered lipoma.

There is a clear overlap in the timelines of the development of spilt cord malformation and lipomeningocele as described above. This fact is reaffirmed by the pattern of migration of cells of meninx primitiva in SCM type 1 as proposed by Pang’s unified theory.[1,2] Hence, the probability of SCM type 2 to be associated with the development of lipomas and lipomeningocoeles should be much more than SCM type 1 and thereby the incidence of double lipomas should be more in SCM type 2 as compared to SCM type 1. But literature reveals that the incidence of the double lipomas along with SCM type 2 is as rare as with SCM type 1 [Table 1].[7,8,22,23] Different unknown genetic pathways for the development of EMT and premature disjunction might be responsible for the phenomenon. It needs further genetic testing such as ARHGAP29 and RADIL genes and embryological studies and hypothesis testing to prove whether the development of double lipomas in association with SCMs are a sequel more than a chance.[24]

**CONCLUSION**

There seems to be a certain overlap in the timeline of embryological errors that result in SCM type 2 and lipomeningocoeles. Although it seems that the mesenchyme of the EMT in SCM type 2 stands a higher chance of developing into a double dorsal lipoma, reports of the phenomenon are still infrequent. The case presented herein is only the fourth case ever reported of an SCM type 2 with a double lipoma [Table 1]. This might be due to the fact that the hemicords formed induce neurulation independently of each other and the occurrence of lipoma over both hemicords could be by chance than a specific sequence.

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**Conflicts of interest**

There are no conflicts of interest.

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