

# Multiple dysraphic anomalies with double thoracic meningocele and lumbosacral myelomeningocele, concurrent Chiari malformation Type I, diastematomyelia, lipomyelomeningocele and hydrocephalus: a case report and literature review.

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## Introduction

The coexistence of multiple dysraphic anomalies is extremely uncommon. In particular, double meningocele and myelomeningocele pathologies. Here, we report a case of a three-month-old infant presenting with multiple coexistent dysraphic lesions that includes thoracic meningocele and lumbosacral myelomeningocele, concurrent Chiari malformation type I, diastematomyelia and lipomyelomeningocele in the lumbosacral region and hydrocephalus. In addition, pertinent literature is also discussed.

## Case Report

### Clinical History

A three-month-old infant was brought to the Neurosurgical Clinic, Tribhuvan University Teaching Hospital, Kathmandu, Nepal, by her mother, presenting with two soft cystic swellings on the back since birth. The lesions were progressively getting enlarged. The mother also noticed that her child did not move her left leg and there was only minimal movement on her right leg. The infant would not cry on passing urine or stool and was always dribbling urine. On probing the family and gestation history. It was found that the mother was a 23 year-old housewife from a village in Shindhupalchowk. The infant was delivered at home at 40 weeks gestation with complications. She had not sought any ante/postnatal check-ups. There was no past history of any serous medical illnesses and no history of drug abuse or use of medicines and infections during pregnancy. She did not take any vitamins or iron supplements during her gestation. This was her second child of non-consanguineous parents; and the first child had no known congenital anomalies.

### Physical Examination (Fig 1)

Physical examination revealed two soft cystic swellings on the back, smaller one at the lower thoracic level and the larger one on the lumbosacral region. The rostral lesion on the lower

thoracic region was approximately 2 x 3 x 2cm while the caudal lesion in the lumbar region was 6x8x5cm in size. The peripheral skin overlying the lumbar myelomeningocele was thick, hyperpigmented with purplish discoloration and the central portion of the sac was thin and glistening and had evidence of squamous epithelization while the thoracic lesion was covered with thick and rugged skin outgrowths. The area surrounding the lumbosacral lump was soft and bulging with fatty subcutaneous padding with skin dumpling at lower end but without dermal sinus, hairy tuft or skin appendages. There was no cerebrospinal fluid (CSF) leakage noted from either of the lesions. There were no clinical manifestations of neurofibromatosis. Initially, the head circumference was 42 cm (98<sup>th</sup> percentile); and the anterior fontanelle was not bulging. Neurological examination revealed motor power of 0/5 in the left lower extremity while the right leg had 2/5 motor power. There was excoriation of skin in the perianal region due to continuous passage of stool and urine.



**Fig 1:** Photograph is of a three-month-old infant with multiple coexistent dysraphic pathologies including double thoracic meningocele and lumbosacral myelomeningocele, lipomyelomeningocele, diastematomyelia and hydrocephalus.

### Laboratory Evaluation

Hematological studies revealed hemoglobin of 10.7g%, white

blood cells of 14900, and neutrophils of 46% lymphocytes of 50%, monocytes of 3% and eosinophils of 1% The blood sugar was 4.9 mmol/L and urea of 3.0 mmol/L.

### Radiological Evaluation

Sagittal view of T2-weighted MR image of the whole spine including brain (Fig. 2A) demonstrated double thoracic meningocele and lumbosacral myelomeningocele. Gross overt hydrocephalus and cerebellar tonsillar herniation were also noted; suggesting Chiari malformation type I. Axial cuts revealed diastematomyelia with two hemicords separated by a large bony spur. (Fig. 2B).

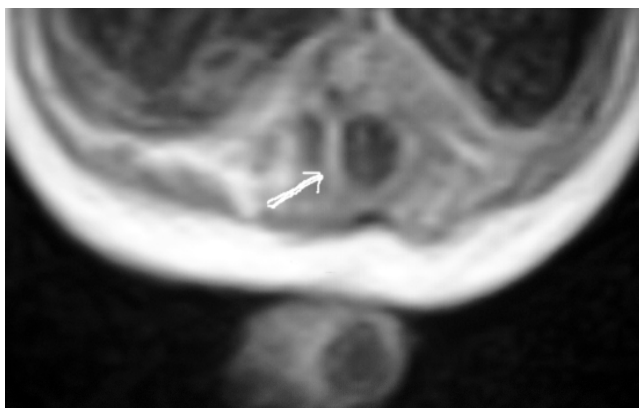
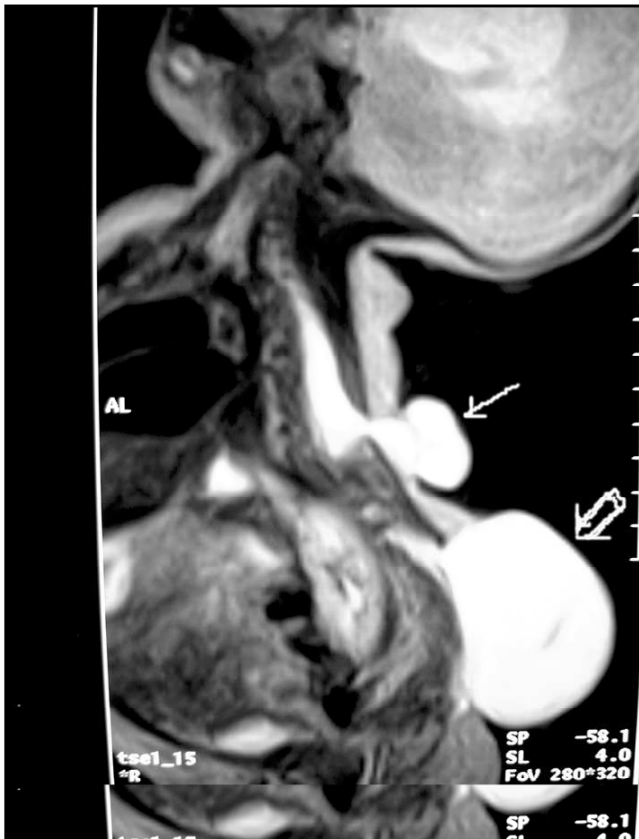


Fig 2: MR Images of the whole spine including brain (A) Sagittal. T2W1 demonstrating thoracic meningocele (thin arrow) and lumbosacral myelomeningocele (hollow arrow). Note the over hydrocephalus and Chiari malformation type I. (B) Axial cuts revealing two hemicords separated by a large bony spur (thin arrow).

### Surgical Approach

The patient underwent surgical exploration under general anesthesia in a heated room with standard latex-allergy protocol. The patient was given intravenous ceftriaxone 250 mg preoperatively. She was put on the prone position on the cloth rolls with padding and with abdomen hanging free. The whole of the back was prepped thoroughly with Betadine and draped. Both the lesions were explored at the same setting, using circumferential elliptical incisions in a longitudinal fashion at the base of the sac and extension at the rostral and caudal end in midline. Adequate normal skin was preserved at the peripheral and caudal end in midline. Adequate normal skin was preserved at the peripheral margin of the sac for closure without tension. The excess of the skin was completely dissected off the placode and excised to prevent retained skin elements, which may predispose to formation of dermoids and epidermoids in the long run.

Some of the subcutaneous fat was removed. (Fig 3 A) and the circumferential dissection was achieved at the margin of the placode, starting from the lateral aspect and leaving the cephalic and caudate ends for the last.

A large bony spur was noted placed slightly oblique, splitting the spinal cord in two hemicords (Fig 3B). The left hemicord and its neural tissues formed the placode and were contained in the meningeal sac while the right hemicord passed rostrally underneath the bony spur and conjoined into one cord at upper lumbar region. There were significant adhesions. There was also fatty tissue covering the nerve roots and spanning the lateral gutter of the spinal canal, covering the nerve roots, extending caudally and to the subcutaneous layer. The remaining fatty tissues were removed meticulously and the adhesions were carefully dissected.

Nerve roots, placode and spinal cord were dissected cautiously with preservation of all functional nerve roots. A few anomalous, rudimentary dorsal nerve roots were sacrificed and the adipose tissue was further excised meticulously without damaging the neural structures. Emphasis was given to untether the spinal cord completely by incising the thick filum terminale to prevent tethering of cord.

Magnification with use of optic loupes and powerful headlight was used for better visualization and illumination to minimize the chances of neural injury during dissection. Bipolar was used for coagulation and the use of monopolar electrocautery was completely avoided.

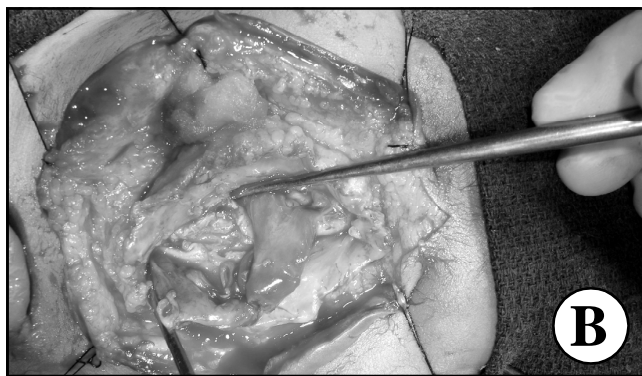
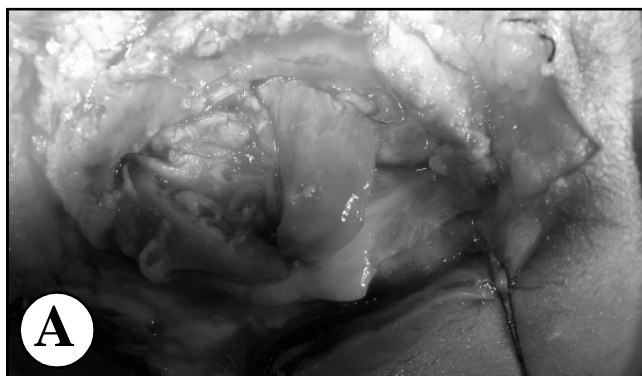
Furthermore, the bony spur was then removed by nibbling with double-action rongeur, 45° Kerrison punch and pituitary forceps. After complete removal of the bony spur, the raw bed was waxed thoroughly to avoid bleeding. The anterior dural flaps were approximated using 4-0 vicryl incorporating the two hemicords into one.

As the standard practice, multi-layer strategy was applied for closure. As far as possible an attempt was made to reconstitute left neural tube, using 5-0 absorbable microsutures. The reflection of dura was circumferentially harvested by continuing the incision circumferentially around sac. The neural elements were dissected off the lateral wall of the sac. Some redundant sac was excised. The posterior

dura was then closed continuously in a watertight, double-sleeving fashion without tension. The paraspinous muscular turnover flap was also used to reinforce the defect. Its integrity was further checked by intraoperative Valsalva maneuver at the end of the dural closure and then upon reinforcement with paraspinous muscular flaps to determine if there was any CSF leakage through the dural and fascial closures.

Simultaneously, the lower thoracic sac was also opened in the similar fashion as mentioned above. The meningeal sac was dissected circumferentially. The sac was then opened. There were no neural elements in the sac. The dura was then closed in watertight, double-sleeving fashion and reinforced by paraspinous muscular flaps.

After thorough irrigation and hemostasis, the two incisions were finally closed in layers; subcutaneous layer with 3-0 vicryl interrupted and skin with 4-0 prolene in interrupted mattress fashion. The incision was painted with Neosporin ointment and covered with Jelonet gauge and opsite to prevent from stool contamination.



*Fig 3: Intraoperative photographs (A) showing the lumbosacral myelomeningocele sac covered with adipose tissue, suggesting lipomyelomeningocele with transitional type of spinal lipoma (B) showing diastematomyelia with the bony spur separating the cord into two halves (direction shown by two Penfield dissector).*

#### **Intraoperative Findings (Fig 3A, 3B)**

The lumbosacral lesion was a large cystic distension with split cord and fatty tissue. The large bony spur noted placed slightly oblique splitting the spinal cord in two hemicords of nearly equal caliber, having individual dorsal and ventral nerve roots. The left hemicord and its neural tissues formed the placode and contained in the meningeal sac while the right hemicord passed rostrally underneath the bony spur. There were significant adhesions. There was also fatty tissue

covering the nerve roots and spanning the lateral gutter of the spinal canal, covering the nerve roots, extending caudally and to the subcutaneous layer.

The thoracic lesion was consistent with a meningocele. There was a bony defect of about 2 x 3sq cm in size at the lower thoracic spine with CSF-filled cystic sac but without any neural tissues such as spinal cord and nerve roots present in the cele.

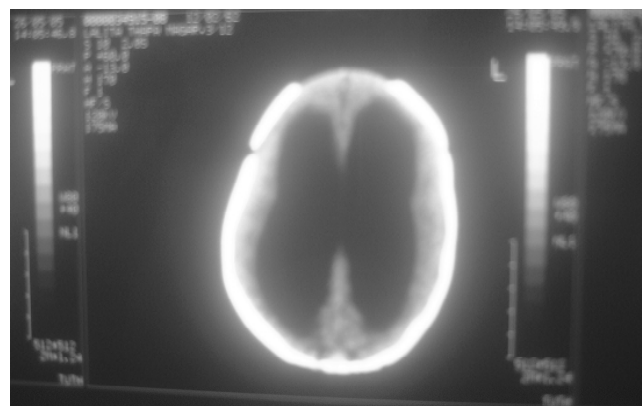
#### **Postoperative Course**

After surgery, the patient was transferred to intensive care unit and then to step-down unit. On postoperative day (POD) one, routine hematological studies revealed hemoglobin of less than 8 %. The patient was tachycardic and pale. Hence, the patient was transfused with 50ml of whole blood.

On POD four, the patient deteriorated with high-grade fever, diarrhea and abdominal distension. Abdominal ultrasound was normal and liver function test was within normal limits. However, urinalysis revealed white cell counts of 8 to 10 and plenty of bacteria were present, suggesting urinary tract infection. Urine culture grew *Acinetobacter calcoaceticus*, which was sensitive to cefotaxime. The infection was completely resolved with use of intravenous antibiotic.

On POD nine, the patient became more irritable. Anterior fontanelle was noted to be very tense and bulging; and head circumference increased to 44 cm (over 98<sup>th</sup> percentile). The patient also developed upward gaze palsy. Immediate CT-scan of head was done which revealed gross triventricular hydrocephalus (Fig 4). Promptly, the patient underwent ventriculoperitoneal shunting (via right frontal approach), using Chhabra medium-pressure shunt under general anesthesia (Fig 1B).

The patient was discharged on POD12 after removal of stitches. There was no drastic change in the neurological status on discharge.



*Fig 4: CT-scan of head eliciting triventricular hydrocephalus.*

#### **Follow-up Study (Fig 5)**

The patient was routinely followed up in the Out-Patient Clinic. On first follow-up after one month of discharge, the patient was alert and playful with well-healed incisional scar. The anterior fontanelle was soft the shunt refilled promptly on compression, suggesting well-functioning shunt. There was no obvious improvement in the paraparesis status and urinary and rectal incontinence situation.



Fig 5: one month after surgery with mother.

## Discussion

Spinal dysraphism refers to a plethora of disorders arising from the incomplete formation of the dorsal midline structures during embryogenesis. Among such congenital anomalies of Central Nervous System (CNS), myelomeningocele is one of the most common midline defects, presenting with disturbance in the closure of neural groove and defects in its skeletal investment. Before 1980, the incidence of meningocele or myelomeningocele was reported to be 1 to 2 per 1000 live births; but however presently, there is a declining trend in many developed countries of the world. It is located in the distal thoracic, lumbar or sacral area in over 85% of the cases. Majority of such cases are associated with type II Chiari malformation as well as syringomyelia, hydrocephalus, diastematomyelia and cerebral malformations.

The true definition of meningocele is said to be as a congenital defect in vertebral arches with cystic distension of meninges without abnormality of neural tissue while myelomeningocele involves structural and functional abnormality of spinal cord or cauda equine as well. The presence of both meningocele and meningomyelocele in an individual is a rare clinical event. A thorough literature search revealed only nine published cases of double myelomeningocele so far. Among them, Rainov, et al (1995)<sup>1</sup> reported two cases with double myelomeningoceles associated with neurofibromatosis type I. Potter noted two cases with multiple open neural tube closure defects of the spine, one of which was associated with hydrocephalus and Chiari malformation<sup>2</sup>. Furthermore, Bertan and Wilson (1968)<sup>3</sup> published a case report of a double meningomyelocele at the thoracic and lumbosacral levels, associated with Chiari malformation and hydrocephalus presenting late nearly two weeks after birth with CNS infection and CSF leak. The patient subsequently succumbed to the shunt infection. Similarly, Bailey (1971)<sup>4</sup> also described one such case with Chiari malformation and hydrocephalus presenting late nearly two weeks after birth with CNS infection and CSF leak. The patient subsequently succumbed to the shunt infection. In 2000, Durmaz, et al,<sup>5</sup> reported a three-day-old boy with double myelomeningoceles at the thoracic and lumbar levels without any other congenital anomalies and hydrocephalus. In the more recent reports, Richards, et al, (2003)<sup>6</sup> described a newborn female who had two myelomeningoceles at thoracic and lumbar region without

hydrocephalus. Etus V, et al, (2005)<sup>7</sup> reported a case of a newborn male with double cervical and lumbar myelomeningoceles and associated with overt hydrocephalus. Apart from the repair of myelomeningoceles, the patient was also subjected to ventriculoperitoneal shunt at the same setting.

The pathogenesis of meningomyelocele is thought to stem from defective closure of the caudal neural tube between 26 and 28 days of gestation, a process known as neurulation<sup>8</sup>. Broadly, two theories have been postulated over the years. The more accepted nonclosure theory as described by Recklinghausen in 1886 proposes that neural tube defects represent a primary failure of neural tube closure<sup>9</sup>. On the other hand, the over distention theory introduced in 1769 by Morgagni<sup>10</sup> and later popularized by Gardner<sup>12-14</sup> postulate that the tube defects arise from hydromyelic overdistention of embryonic neural tube resulting in a rupture of the roof and floor of the previously closed neural tube without damage to the overlying cutaneous ectoderm or ventral endoderm.

The conventional concept “the zipper model” in respect to primary closure of the neural tube in humans describes the neural tube to start from a single initiation and to proceed continuously to the most rostral and caudal ends of the neural plate in a bi-directional manner.<sup>15</sup> It is thought to close in a linear fashion like zipper extending cranially and caudally from the point of initial closure.

However, this conventional concept of single-site neural tube closure failed to provide satisfactory explanation for all spectra of neural tube defect in human, particularly in cases with multiple coexistent dysraphic anomalies including double meningocele/myelomeningocele pathologies. Presently, neurulation in mammals (including humans) appears to extend from several initiation sites along its cranio-caudal axis<sup>15-16</sup>. This may be able to partly explain the presence of multiple dysraphic anomalies including double meningocele and myelomeningocele. Van Allen, et al, (1993)<sup>16</sup> proposed a concept with multi-site neural tube closure in humans, analogous to mice and other experimental animals. This model suggests that the closure of the neural tube occurs at five different initiation sites. Defects at different closure sites lead to variety of spinal dysraphism including double pathologies and multiple coexistent dysraphic lesions. In accordance to the Van Allen’s new concept of multi-site neural tube closure, the presence of double meningocele/myelomeningocele in our case may be explainable by defect at the caudal Closure I resulting in the thoracic meningocele and defect at Closure 5 resulting in formation of the lumbosacral meningomyelocele

## Literature review

In general, spinal lipomas are the most common occult dysraphic malformations and most frequently involve the lumbosacral spinal cord, conus medullaris and filum terminale. In 85% of the cases, there is a visible subcutaneous mass that is contiguous between bifid lamina. Lipomas arise within the area of diastrophism, occupying central canal of the involved spinal cord and occasionally extending cephalad within the central canal to a variable degree. These lesions

have been broadly categorized into three groups dorsal, terminal and transitional depending on the relationship with involved spinal cord segment<sup>19</sup>. The dorsal lipomas is exophytic dorsally in respect to the spinal cord and sits on the top of the dorsal surface of the involved segment while the terminal lipoma starts from the caudal end of the spinal cord and extends caudal to the tip of conus medullaris. On the other hand, the transitional type represents a combination of the other types and is exophytic both dorsally and caudally from the conus medullaris, analogous to the findings in our case. The current theory of embryogenesis proposes that lipomas arise from premature dysjunction during primary neurulation<sup>20,21</sup>. Lipomeningomyelocele is relatively complex, consisting of intra/extradural spinal lipoma, dorsal spinal dysraphism, spinal cord tethering and subcutaneous mass. The term diastematomyelia describes a primary embryological malformation involving division of a variable length of spinal cord into two hemicords of more or less equal caliber, each containing an individual set of dorsal and ventral nerve roots. The division may be partial or complete. Often a septum of bone, cartilage or fibrous band is present between the hemicords. Split cord malformation may arise in association with open neural tube defect or, more commonly as occult malformations, developing in isolation or in conjunction with other associated anomalies. It is seen in association with myelomeningocele in up to one third of cases<sup>22</sup>. In this case, the split cord malformation was noted at the lower lumbar region and there was a large bony spur separating the cord into two halves covered with lipomatous tissue extending up to the subcutaneous layer.

In respect to multiple coexistent dysraphic pathologies, Solanki reported an extremely rare case with quadruple dysraphic lesions, which included concurrent segmental meningocele, type-I split cord malformation associated with hemi vertebrae, lipomeningomyelocele in each hemicord and a terminal myelocystocele<sup>23</sup>. In this case report, the multiple dysraphic pathologies noted were thoracic meningocele, lumbosacral myelomeningocele, Chiari malformation type I, diastematomyelia with bony spur and lipomyelomeningocele at lumbosacral region, as well as hydrocephalus.

Existing embryologic hypotheses and explanations for presence of variety of dysraphic spectrum lack experimental evidence and studies in animal models. The mere presence of multiple dysraphic anomalies in an individual challenges the present accepted hypotheses and demonstrates our inadequacies in understanding of spinal cord embryogenesis. Basic knowledge of normal anatomy and thorough understanding of embryogenesis and pathogenesis provide a solid background for the accurate diagnosis and rational

treatment strategy in the dysraphic patients.

Over a period of ten years, we managed surgically 64 cases with spinal dysraphic defects with good results. Our recommendation in the technical considerations for surgical approaches of these challenging entities has been elaborately discussed in our previous publication<sup>24</sup>. In brief; surgical intervention should be directed at closing the defect and possibly decompressing the ventricles<sup>25</sup>.

The fundamental purpose of surgical repair should be to prevent further progression of the existing neurological deficits rather than to produce improvement.

In our case, there was no improvement or deterioration of neurological status with surgical intervention.

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