Case Report

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Huge Lumbosacral Lipomeningomyelocele with Good Post-Natal Surgical Outcome

Purvi Desai[1], Kairavi desai[2], Dinesh zinzala[3], Kalpana kathrotiya[4], Binodini Chauhan[5]

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Authors Affiliation: [1]Professor, Head of Department in Radiology, [2,3]Fellow in Fetal Medicine, [4]Consultant in Fetal Medicine, [5]Fetal Medicine Consultant, Government Medical College and New Civil Hospital Surat, Gujarat 395001, India.

Correspondence Author: Dinesh Zinzala, Fellow in Fetal Medicine, Department of Radiology, Government Medical College and New Civil Hospital, Majuragate, Surat, Gujarat 395001.

E-mail: zinzala100@gmail.com

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Abstract

Management of lipomyelomeningocele is one of the most discussed and controversial topic in recent years. Till date, there is no consensus on most appropriate mode of management for lipomyelomeningocele, particularly in asymptomatic patients. It represents a complex disorder that may present with neurological deficit secondary to the inherent tethered cord. Here we present an antenataly diagnosed case of huge lumbosacral lipomeningomyelocele with good post-natal outcome.

Keywords: Lumbosacral lipomeningomyelocele; Asymptomatic patients; Ultrasonography; Sacrococcygeal teratoma.

Introduction

Incidence of lipomyelomeningocele is 1 in 4000 live births with slight female preponderance. Lipomyelomeningocele may be defined as a defect in the spine through which the lipomatous substance arising from subcutaneous tissue is inserted into spinal canal. Lipomeningomyelocele may present with neurological deterioration secondary to inherent tethered cord.

Ultrasonography is initially useful in the diagnosis of lipomeningomyelocele during antenatal period but MRI done antenataly or postnataly is definitive for diagnosis and preoperative planning. More than 90% cases present with obvious soft tissue swelling over the spine in lumbosacral region and most develop neurological symptoms within the first few months to years of life.[2] There is progressive neurological, urological and orthopaedic deficit in patients with lipomyelomeningocele if left untreated.[3]

Case Report

A case of a young 2nd gravida with 36 weeks pregnancy visited the department of fetal medicine, New Civil Hospital for second opinion for sacrococcygeal teratoma. She had one healthy male child. There was no significant medical history or history of consanguinity. Previous scan done at private centre had diagnosed huge sacrococcygeal teratoma at 35 weeks. NT scan and anomaly scans were not done. Ultrasound findings at our centre were: Normal Intra cranial anatomy and posterior fossa. Examination of spine revealed a thick walled huge cystic lesion of 68x52 mm covered with skin

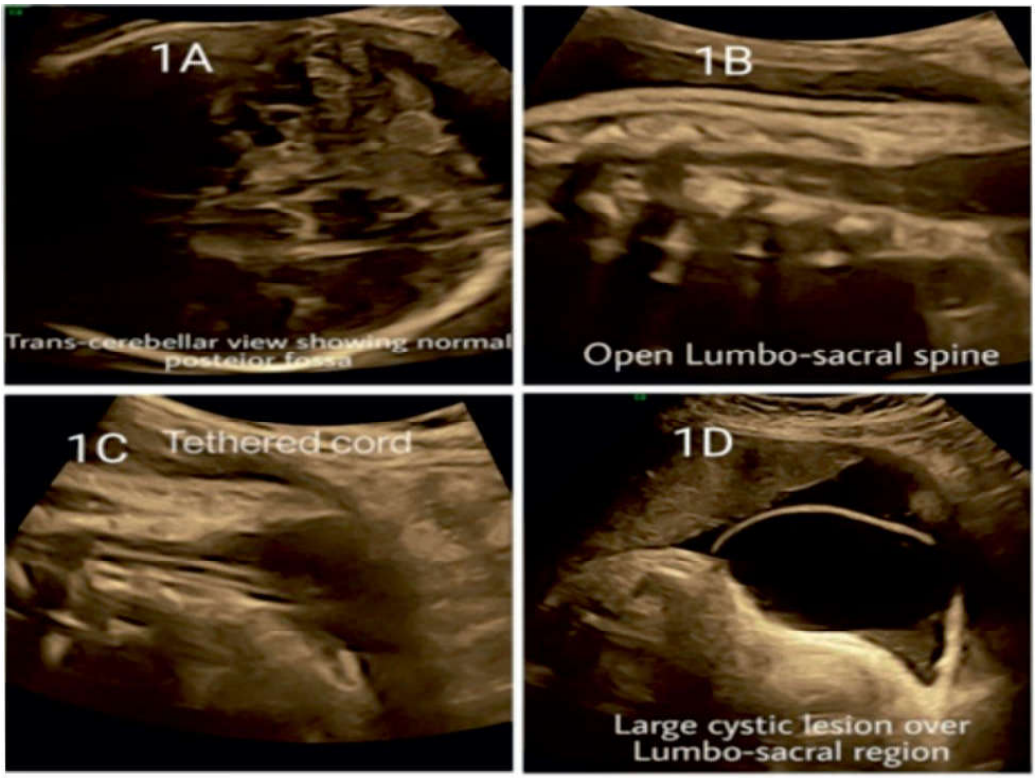

Fig. 1: ultrasound finding at 36 weeks.

1A normal posterior fossa in trans-cerebellar view; 1B open lumbo sacral spine in saggital view; 1C Neural elements extending in the lesion-tethered cord; 1D large cystic lesion at lumbosacral region.

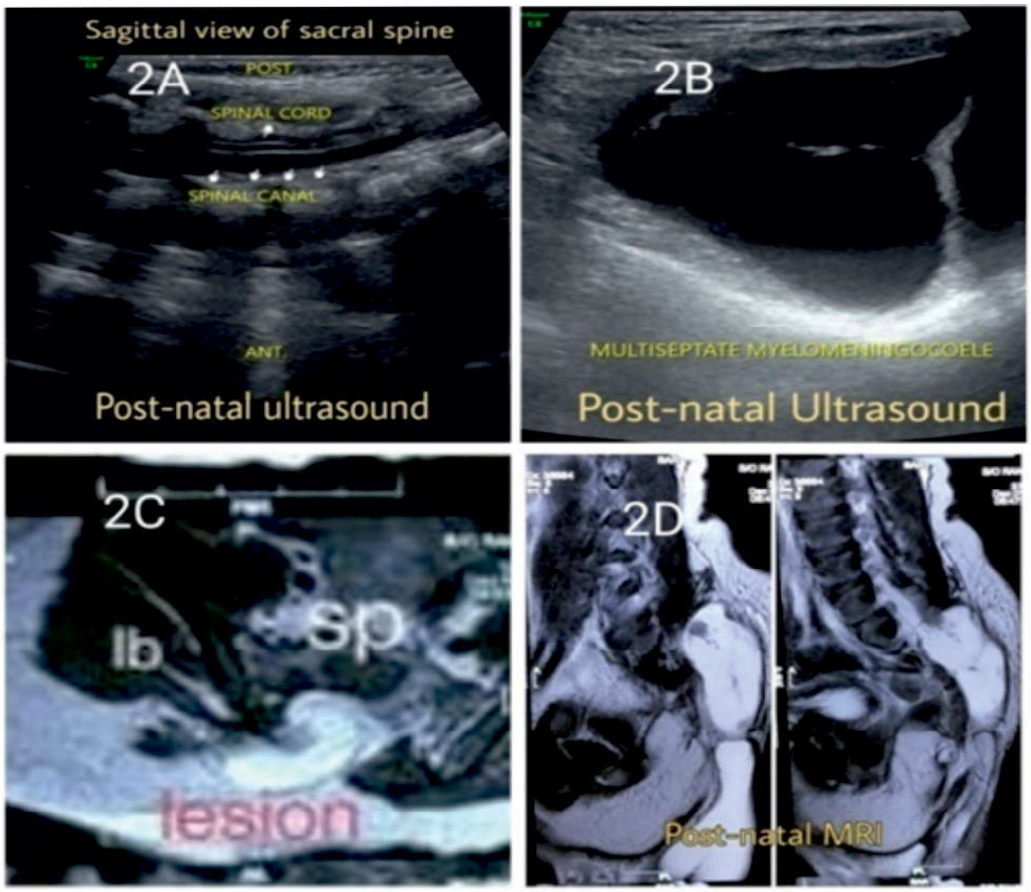

Fig. 2: Showing post natal ultrasound and MRI findings.

2A Tethered spinal cord

2B multiseptate myelomeningocele at lumbosacral region

2C and 2D MRI images showing spinal defect with lipomatous tissue covered with skin.

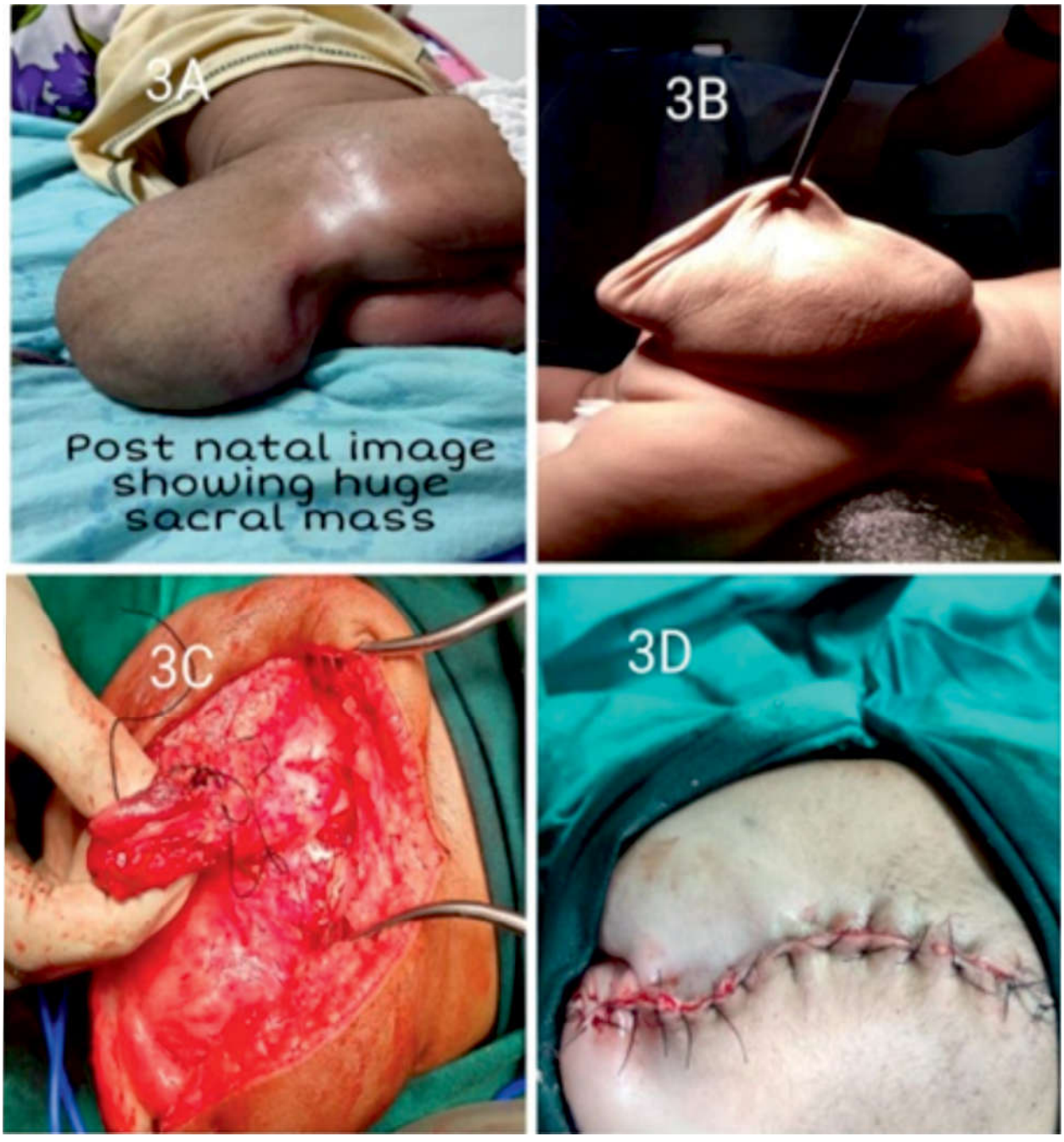

Fig. 3: Showing intra-operative finding.

3A and 3B post natal image of huge lumbosacral mass

3C intraoperative picture

3D postoperative operative site

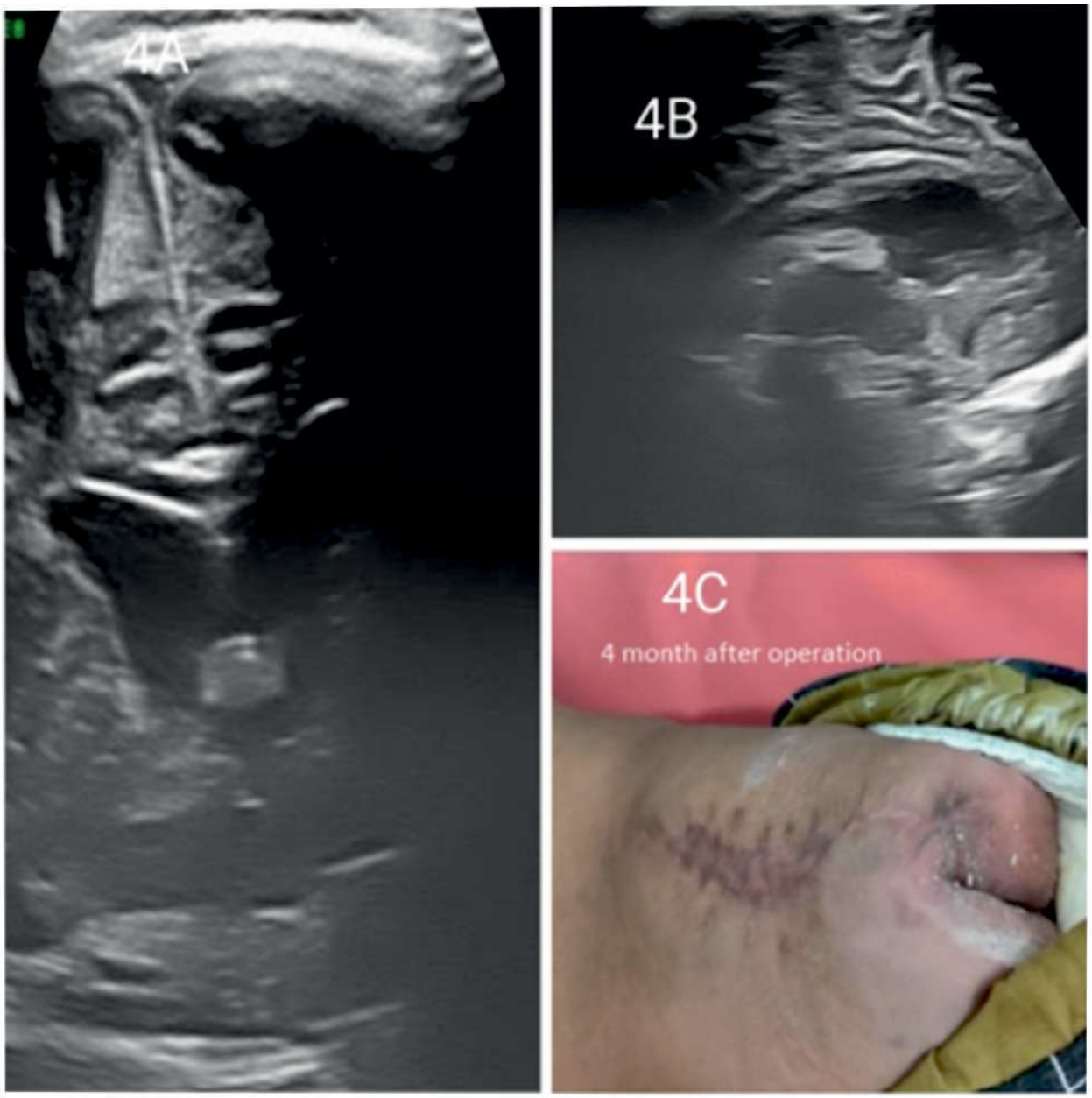

Fig. 4: Showing usg finding and operative site 4 months post surgery.

4A coronal view and 4B sagital view showing normal anterior complex of brain in Fontanalle scan
4C scar site 4 month post surgery

arising from the lumbo-sacral region. Entire lesion appeared external. Neural elements could be seen extending into the lesion suggesting tethered cord. Rest of fetal anatomy and fetal growth appeared normal. The findings were suggestive of "A single Live Intrauterine gestation corresponding to 36 weeks 6 days with huge Lumbo-sacral Lipomemningocoele/spina bifida occulta with Tethered Cord".

Baby delivered by caesarean section at term. There was huge mass, soft in consistency, covered with skin over lumbosacral region. Post natal ultrasound and MRI were done and were suggestive of spina bifida occulta (lipomeningomyelocele). Neurosurgeon was consulted and newborn was operated at 4 months post delivery. At present baby is 10 months old and there are no complications.

Discussion

Recent advances in both ultrasonography and MRI have been helpful in the diagnosis and treatment of spinal dysraphism, both prenatally and postnatally. Detail examination of fetal spine

in various planes is helpful for prenatal diagnosis of lipomyelomeningocele. All open spinal dysraphisms are associated with an abnormal appearance of the posterior fossa on obstetric ultrasonography.[4] The unique feature in our case was an absolutely normal posterior fossa despite such a huge lesion.[5] A late 3rd trimester ultrasound detected a well-demarcated subcutaneous mass in the lower lumbo-sacral area.

The spinal cord could be seen extending into the mass. Postnatally, MRI aided in confirmation of diagnosis and planning surgery of the huge lipomyelomeningocele. Neurological symptoms may be absent at birth in nearly half of the cases. As the infant ages and axial growth occurs, the infant may experience progressive loss of neurological function.

Often, a change in the pattern of bladder and bowel function is the presenting symptom of lipomyelomeningocele.[6] As axial growth continues, lower limb, and sacral motor and sensory dysfunction, such as radicular pain, leg spasticity, foot deformities, and gait abnormalities, can develop.[2] There is progressive neurological, urological and orthopaedic deficits in patients with lipomyelomeningocele if left untreated. Preoperative neuro-urological status is a prognostic factor in determining the postoperative outcome.[2] Poor pre-operative prognostic factors are hydrocephalus at birth, total paralysis of limb, other associated congenital defect and poor general condition.[7] In our case none of these were present

Surgical step for lipomeningomyelocele includes Step 1:-incision and laminectomy to identify the entry point of stalk, Step 2: detachment of lipoma from dura, Step 3: lipoma resection, Step 4: neurulation of neural placoda, Step 5: expansile duraplasty. Good post-natal surgical outcome is the presence of reflexes, spontaneuos leg movements and good urinary streaming. The newborn is now 10 months old and has a normal neurosonogram, the normal urine stream and the lower limb movements and relexes are also normal.

Conclusion

This is a rare case of a very late diagnosis of a huge lumbo-sacaral lipomeningocoele, successfully managed postnatally with no neurological, urological or orthopaedic deficiency till date (6 months post surgical). If it had been diagnosed in 1st or early 2nd trimester, Pt may not have continued pregnancy and even the counselling would have been difficult. Hence, the size of the lesion should not discourage the doctor and the pt in taking a bold step and continuing pregnancy.

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