

Lipomyelo Meningocele - A rare congenital anomaly in a female child

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Abstract

A 4-month old female child reported to Paediatric Out Patient Department at Nepalgunj Medical College, Teaching Hospital, Nepalgunj, Nepal with a history of progressive slow growth over the lumbosacral region since birth. There was no neurological deficit, without bladder involvement but she had only single growth over the lumbosacral region with depression in center. This growth was diagnosed as lipomyelomeningocele which is very rare congenital anomaly. Hence this case was studied and reported.

Keywords: Lumbosacral; Myelomeningocele; Lipomyelomeningocele; Meningocele; Neural tube defect.

Introduction

Lipomyelo meningocele is rare congenital growth seen in newborn. It is slow growing progressive tumour intermingled with nervous tissue which gets stretched when the lower end of the cord moves upwards in relation to spine. Usually it is not associated with neurological signs. Hence symptoms appear in lower limbs and urinary bladder very progressively and slowly. So excision of Lipoma has to be done within 6 months of age in order to prevent progressive neurological symptoms. Surgery is very difficult when once nervous tissue gets entangled with Lipomatous tissue. Skin over the swelling gets dimpled in region of Lumbo sacral area. This tumour can be diagnosed on radiological examination. The treatment is same as that of neurosurgical approach.¹

Case Report

A 4-month female child came to paediatric out patient department of Nepalgunj Medical College Teaching Hospital at Nepalgunj with complaints of progressively increasing growth in lumbo sacral region since birth. The growth gradually increased in size and attained the present size associated with watery discharge but not foul smelling. There were no other complaints nor any other growth in the body.

Fig. 1

Fig. 2

Family History

No family history of similar growth. No other abnormal pregnancy in the family

Obstetrical History

There was no antenatal check up during pregnancy. Child was delivered at home normally after full term. Prenatal and postnatal periods were uneventful.

On examination

Child was poorly nourished with pallor. No evidence of clubbing, jaundice, cyanosis and generalized lymph adenopathy was found. Afebrile with heart rate of 110/m and respiratory rate of 32/min. The size of head, its shape and circumference were within limit without any facial dysmorphism. There were no abnormalities of eye, ear, nasal and oral cavities.

Weight of the child = 3 kg

Length of the child = 63.8 cm

Circumference of head = 36.5 cm

Chest circumference = 34.4 cm

On systemic examination

CVS and RS – NAD P/A – Soft, No Organo megalia CNS – No Neurological deficit. No involvement of bowel and bladder. Lower limbs were normal.

Local examination

Over the Lumbosacral region, a single swelling measuring 4" * 5" was seen. Skin over the swelling was not discoloured but normal healthy skin, pedunculated having midline cleavage extending from the center to the lower end with sinus to the left. Consistency was soft with trans illumination test negative.

Investigation

Routine Haematological examination normal except she was anaemic.

X-ray of lumbosacral SPINE in A-P and lateral view revealed soft tissue mass seen at the sacro coccygeal region. Lower sacral and coccygeal spines are deviated to right side. The child was advised for further investigation.

Ultrasound report revealed a growth at coccygeal area which is large mass but no fluid present. Neuro fibroma ?

MRI showed lipomyelomeningocele said to be a diagnosed by an expert Radiologist. Child was referred to Neuro surgeon for further management.

Discussion

1. **Lipo myelomeningocele:** They are subcutaneous lipoma diagnosed at birth seen over the lower spine in lumbo sacral region. They always terminate in Conus medullaris which is displaced caudally. They are neurologically normal at birth but slowly loss of normal neurological functions sets in within few weeks to few months. Hence early removal of lipoma is advisable made to (to prevent neurological deficit) maintain normal neurological function.⁴

2. **Myelomeningocele:** Incidence is 1 in 1000 live birth. Exact cause is unknown. It is neural tube defect with genetic predisposition. After on affected child, risk of re occurrence rises to 3-4%. Use of folic reduces the incidence of neural tube defect by 50% with dose of 0.4 mgm of folic acid.³ It is growth in the form of protrusion at lumbosacral region containing CSF, NERVE ROOTS, MENINGS WITH DYSPLASTIC SPINAL CORD. There is definite line of demarcation between skin and membranous sac which is protruding. There may be no neurological deficit but there may be association with Arnold Chiari – malformation with hydrocephalous. There may be also motor and sensory neurological deficit with involvement of bowel and bladder. Surgery is indicated when there is involvement of bowel, bladder and both lower limbs.² There may be associated extensive Kyphosis, severe hydrocephalous with paralysis of lower limbs. Hence radical line of treatment is very much contraindicated.⁶

3. **Meningocele:** It is also a growth over the back covered by the skin with no line of demarcation between two areas of skin and meninges.² Meningocele is the result of herniation of meninges through defect in post vertebral arches characterized by mid line mass in low back, fluctuant and transilluminant. Spinal cord is normal and in normal position, sometimes it is associated with diastematomyelia, tethered spinal cord, hydrocephalous and lipoma. If associated with hydrocephalous, CT scan is must. Increase in size of lesion may result in dysfunction of bladder and constipation. May have genital anomalies like recto vaginal fistula and vaginal septa.³ The use of plain X-ray useful to show sacral defect. Ultrasonography, MRI outline (determines) the extent of meningocele. Later surgical excision should be done immediately if there is CSF leak or covered by very thin skin to prevent meningitis.³ If the growth is covered by full thickness skin with normal neurological functioning surgical excision may be delayed.³

4. **Sacro coccygeal Teratoma:** They are the teratomas with a frequency of one in 37000, seen 3 times more in females than males in the sacral region. Hence the name **Sacro Coccygeal Teratoma**. They are formed by proliferation of pluripotent cells which contain cell arising from all 3 germinal layers. Hence it has tissues arising from ectoderm, mesoderm and endoderm. There are persistence of remnants of primitive streak.⁵

In the present study

- child is female aged 4 months old.
- with a mass over the lower back at lumbo sacral region.
- skin over the mass is healthy with depression. It is not fluctuant and transilluminant. No other swelling over the back.
- child had no neurological deficit. No involvement of bladder and brain but size of growth is increasing progressively which is not associated with hydrocephalous.

Since the growth is progressively increasing in size covered by healthy skin with grooved derpression without involving the lower limb, bladder and brain. The diagnosis goes in favour of **lipomyelomeningocele**, since

- growth progressive and slow
- no line of demarcation of skin between skin and growth - healthy skin but there is depression with a groove.
- power, tone, bulk of the muscles of lower limbs normal.
- no incontinence of bowel and bladder.
- no neurological deficit.
- other systems like CVS and RS were normal.

Conclusion

In the present study lipomeningocele is a congenital growth formed due to neural tube defect which is preventable by administration of 0.4 mg of Folic acid during pregnancy. Hence timely administration of Folic acid and proper antenatal checkup with Ultrasound screening is mandatory during pregnancy. Awareness of the usage of Folic acid and Ultrasound screening with proper antenatal checkup especially in rural population (especially in rural and hilly areas of Nepal) will bring down the incidence of above congenital defects.

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