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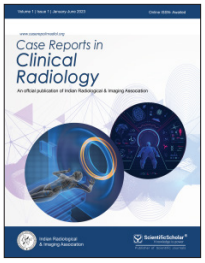

Case Report

Anomalous osseous limb: A sneak peek into rare association with lipomyelomeningocele – A case report

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ABSTRACT

Anomalous osseous limb is a rare entity and its association with spinal dysraphism is all the more rarer. We report one such case of lipomyelomeningocele in 1-month-old female, with associated anomalous osseous structure/limb in relation to iliac bones. Various investigators in the past, though, have published association of anomalous osseous limb with lipomyelomeningocele, the uniqueness of our report lies in documenting such an anomaly at this early age, with paucity of literature in this young age, to the best of our knowledge.

Keywords: Anomalous osseous limb, Lipomyelomeningocele, Spinal dysraphism

INTRODUCTION

Spinal dysraphisms are congenital developmental anomalies, classified as open or closed. Lipomyelomeningocele is a closed neural tube defect (NTD), due to defective primary neurulation, where the mesenchymal tissue enters into neural placode, differentiating into fat. Although the exact mechanism of occurrence of anomalous non-neural structures with spinal dysraphisms is not well understood, various tissues such as muscle, fat, nerve, and bone can be observed rarely with closed spinal dysraphisms, more likely lipomyelomeningoceles.[1-4]

CASE REPORT

A 1-month-old female presented with skin covered swelling in the lumbosacral region since birth, with dimpling on the skin overlying the swelling [Figure 1]. Her mother gave history of absent antenatal folate intake. The baby was born at 40 weeks of gestation by spontaneous normal vaginal delivery. Neonate was referred for ultrasound (US) of spine to rule out spinal dysraphism. Mother's consent was obtained for further imaging. US spine revealed diverging posterior elements in lumbosacral region with a large subcutaneous lipoma extending into the spinal canal. The CSF space in this region appeared expanded with herniation of CSF and neural elements outside the spinal canal into subcutaneous fat. An anomalous midline bony structure was seen in the sacral subcutaneous plane [Figure 2a]. Furthermore, a thin hypoechoic tract was seen extending from subcutaneous lipomatous mass up to this anomalous bone. Plain radiograph of LS spine AP and lateral views showed lumbosacral soft-tissue mass, with accessory bone in the sacral subcutaneous plane [Figure 2b]. CT scan correlation was done for better corroboration of anomalous bone on US and radiographs [Figure 2c]. MRI

lumbosacral spine with whole spine screening was done for pre-surgical planning, on 3T GE discovery MRI scanner. The sequences acquired were axial T1 FSE (with and without fat suppression) and T2 FSE (with and without fat suppression); sagittal T1 FSE non-fat-suppressed and T2 FSE. MRI brain was also performed for any associated intracranial malformations using T1W, T2W, FLAIR, DWI, and SWAN. MRI spine demonstrated lumbosacral spina bifida spanning from L4 to S5 vertebrae, covered by skin and subcutaneous tissue.

A large subcutaneous lipomatous mass was noted in the back at the level of defect, which was herniating through the bony defect into spinal canal. Spinal cord was low lying and exiting through the defect at L5–S1 level, along with CSF, posteroinferiorly into the subcutaneous fat, to reach until S5. The neural placode lipoma interface was lying outside the spinal canal, at the level of lower border of S5 [Figure 3a and b]. An anomalous osseous structure of length 3.2 cm, differentiated into cortex and medulla, was seen arising from the right iliac wing posteriorly and dividing into the right and left limbs extending posterosuperiorly within the subcutaneous fat [Figure 3c and d]. In addition, a thin fibrous tract was seen extending from the dimpled skin surface, tracking anteriorly up to the left limb of this osseous structure [Figure 3c]. MRI brain revealed no associated parenchymal abnormality.

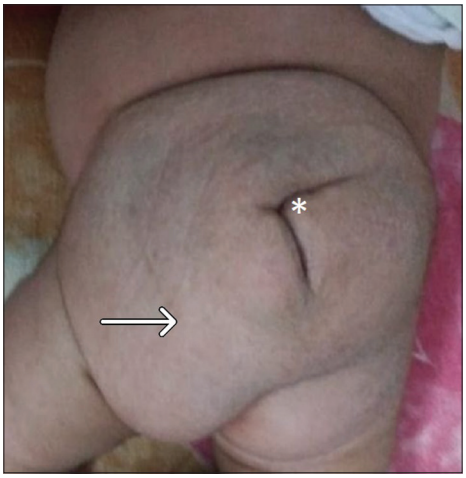

Figure 1: A large well defined skin covered mass seen in lower back region (arrow) with a skin pit (*).

DISCUSSION

Closed spinal dysraphism with subcutaneous mass (lipoma with dural defect) includes lipomyelomeningocele and lipomyelocele depending on the location of cord lipoma interface.[1] The association of anomalous osseous limb with spinal dysraphism is rare.[1-6] Such association was first described by Krishna *et al*. in 1989.[4] Gardner and Egar hypothesis proposes lipomyelomeningoceles to be a secondary NTD, where rupture of neural tube beneath an intact ectoderm leads to accumulation of proteinaceous fluid under skin, serving as rich source of Schwann cells, which further dedifferentiates into non-neural elements such as fat, cartilage, bone, nerves, muscle, or finally into an accessory limb.[2,3] Anomalous osseous limb in our case was partially developed, with no cartilage or muscle or digits formation, may be explained by this theory. Another explanation for accessory limb development is parasitic twinning. According to this theory, failure of development of an embryo in multiple gestations in intrauterine life

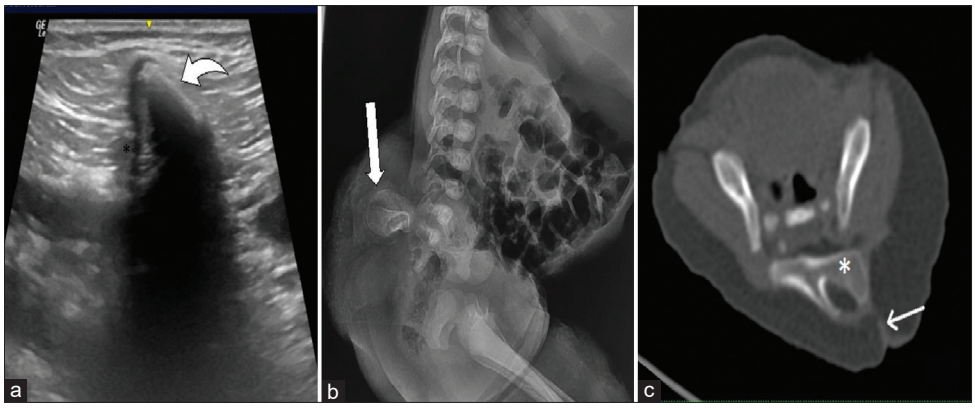

Figure 2: (a) An echogenic structure with dense posterior shadowing seen in the subcutaneous plane distal to the posterior midline sacral defect (curved arrow). (b) Lateral radiograph of lumbosacral spine showing a well-corticated bone in the lower sacral region posterior to the spine (thick arrow). (c) Axial CT section of the pelvis showing an extra bone posterior to the sacrum and iliac bones in the midline which is dividing into right and left limbs (*) with a hyperdense tract running anteriorly from skin surface and joining the anomalous bone (thin arrow).

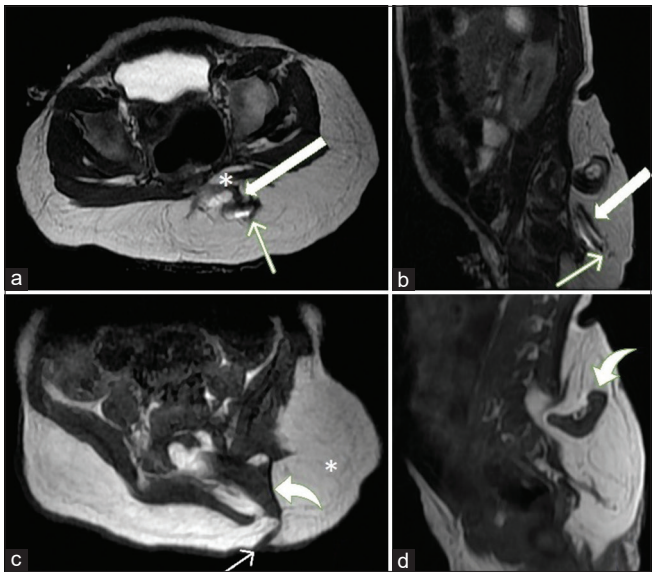

Figure 3: Axial T2W FRFSE (a) and sagittal T2W FRFSE (b) images Axial and Sagittal T2W FRFSE (3a and 3b respectively) shows neural elements along with CSF herniating through the spina bifida (*in Fig 3a) into subcutaneous plane (thick arrow) and is seen to form the cord lipoma interface in subcutaneous plane (thin arrow) suggestive of lipomyelomeningocele. (c and d) shows anomalous osseous limb (curved arrow) arising from right iliac wing posteriorly and dividing into right and left limb differentiated well into cortex and medulla growing posterosuperiorly in large subcutaneous lipoma (*in Fig 3c). A thin linear hypointense tract is seen in subcutaneous plane extending from skin surface up to the anomalous osseous bone (thin arrow).

leads to attachment of the rudimentary underdeveloped or partially developed parasitic body parts of this embryo to the intact well developed vertebral column of the normally developed embryo, as a survival mechanism.[7-9] Many other anomalies such as ectopic tissue, phallus, syndactyly, scrotal tissue, undescended testis, bladder exostrophy, syndactyly of the second and third toes, anorectal agenesis with rectovestibular fistula, renal agenesis, dermal sinus, hemangioma, and bilateral talipes deformities have also been reported in association with accessory limb.[7,8] Our patient showed associated dorsal dermal sinus. Various authors in the past have named this accessory osseous structure/limb differently such as intra-individual ipsilateral dipodia, heterotopic redundancies, aborted twinning, teratoma, tripedus and dysraphic appendage, rudimentary accessory limb, and anomalous bone associated with spinal dysraphism.[10,11] The anomalous osseous limb partially or fully developed can be differentiated from the teratomatous mass, by the presence of well corticated osseous structures with or without digits, while, in teratoma, there is presence of disorganized osseous/calcified structures with variable amount of soft tissue and fat. Another possibility of attached Rachipagus parasitic twin can be considered, which is another extreme end of the spectrum with formation of well-differentiated body parts, most commonly extremities and genitalia.[9-11]

CONCLUSION

Reviewing the literature and knowing the occurrence of non-neural components in association with closed spinal dysraphisms, more likely lipomyelomeningoceles, it is advisable to have a thorough sneak peek on imaging into the presence of such an accessory limb/osseous structure, as seen in our case, which proves to be highly pivotal in guiding appropriate surgical interventions.

TEACHING POINTS

1. Anomalous osseous limb is a rare association of closed spinal dysraphisms especially with lipomyelomeningocele.
2. Presence of well corticated bony structure, partially or fully developed osseous limb should be reported on imaging in cases of closed spinal dysraphisms for appropriate future management.

MCQs

Q1. Which of the following is not a closed spinal dysraphism with a subcutaneous mass?

A. Lipomyelocele
B. Lipomyelomeningocele
C. Meningocele
D. Myelomeningocele

Answer Key: d

Q2. Lipomyelomeningocele is associated with which of the following Arnold Chiari malformations?

A. Type 1
B. Type 2
C. Type 3
D. Type 0

Answer Key: a

Q3. Lipomyelomeningocele occurs due to defect in which stage of spinal development?

A. Gastrulation
B. Primary Neurulation
C. Secondary Neurulation
D. Retrogressive Differentiation

Answer Key: b

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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