ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 09 | September - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

TRIPEDUS DEFORMITY WITH SPINAL DYSRAPHISM A RARE SCENARIO.

Neurosurgery

Dr. Akash Chetpet* Post Graduate, Department of Neurosurgery, Guntur Medical College Guntur Andhra Pradesh *Corresponding Author

Dr. B. Hanuma srinivas Consultant Neurosurgeon, Brinda Hospital, Guntur, Andhra Pradesh.

Dr. Seshadri shekar Associate Professor, Department of Neurosurgey, Siddhartha Medical College, Vijayawada, Andhra Pradesh.

Dr. KVV Satyanarayana Murthy Professor and HOD, Department of Neurosurgery, Guntur Medical College, Guntur, Andhra Pradesh.

ABSTRACT

Accessory limb (Tripedus) with spinal dysraphism is a rare anomaly with only 70 cases described all over the world till date. Of which only 5 cases have been reported in India. This is the 6th case of Accessory limb with spinal dysraphism in India which was treated successfully. Though such deformities do not carry immediate threat to life, significant distress among the parents concerning the deformity of their child will become a primary issue, to be handled by the surgeon. Adequately planned surgical ablation of the limb and repair of the associated spinal dysraphism demonstrated to help to convent the grotesque deformity into a humanoid appearance.

KEYWORDS

spinal dysraphism, tripedus deformity,Lipomyelomeningocele.

INTRODUCTION

Accessory limb (Tripedus) with spinal dysraphism is a rare anomaly with only 70 cases described all over the world till date. Of which only 5 cases have been reported in India. We present to you the 6th case of Accessory limb with spinal dysraphism which was treated successfully. Accessory limb in spinal dysraphism occurs due to mesodermal defect involving the paraxial mesoderm or due to failure of regression of limb bud of conjoined twin.

CASE REPORT

A just born full term female child, weighing 3100gm born out of a consanguineous marriage delivered through LSCS was presented to us with an accessory appendage attached to midline at lumbar region. TIFA scan was not done in the antenatal period. Clinical examination of the infant revealed a well-developed accessory limb of 15cm in length arising from back of lumbar region. Grossly, it resembled a lower limb with hip, knee, and a foot with polydactyl, at the base on the medial aspect of the accessory limb a rudimentary phallus and dark pigmented skin resembling scrotal skin was present (Figure 1). Radiographically, x-ray showed bony components of accessory lower limb and MRI whole spine showed lumbar lipomyelomeningocele with a defect of 15mm noted in the posterior elements of L1,L2,L3 and an accessory limb attached at lumbar region (Figure 2). There were no spontaneous movements of the accessory limb noted but sensation of pain is intact in the accessory limb. Baby had normal looking external genitalia, Foot drop was present in the right lower limb. Rest of the limbs had normal movements.

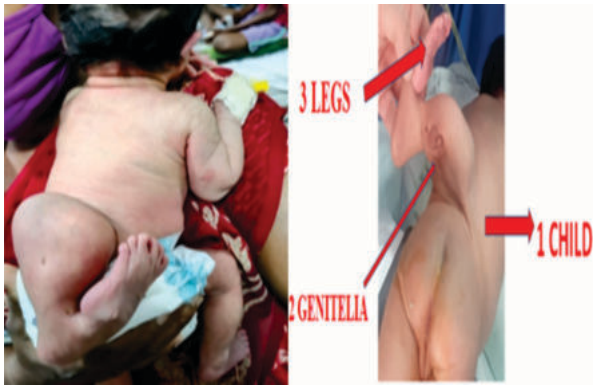

Figure 1: Photograph Showing Tripedus Deformity And Ambiguous Genitalia.

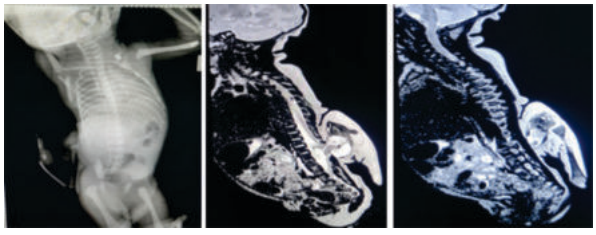

Figure 2: X ray. and MRI Images Showing Herniation Of Meninges Into Accessory Limb.

Procedure

Baby was operated on day 25th day. Positioned in prone. An elliptical incision taken around the accessory appendage. Deeper dissection undertaken,. Sac noted to be communicating with spinal canal at L2 and L3. L1 Laminectomy done. Dura delineated around the accessory limb and limb excised, dissection and repair of lipomyelomeningocele was done. Neural placode was carefully detached. A new dural tube was reconstructed (Figure 3). Skin was approximated primarily.

Figure 3: Intraoperative Pictures Showing Dural Sac Entering Accessory Limb, Disconnection Of Neural Placode, Resection Of Accessory Limb.

DISCUSSION

In India, the incidence of spinal dysraphism is 1.9 per 1000 births. [1] According to literature these anomalies like accessory limb with spinal dysraphism are 4 times more common in females than in males. Our patient was a female child. Literature review reports that the accessory lower limb is associated with lipomyelomeiningocele in 50% of cases with meningocele in 36% of cases and with myelomeningocele in 14% of cases[2]. According to hypotheses of Gardner and Egar[1] lipomyelomeningoceles are secondary neural-tube defects that occur due to rupture of the neural tube under intact ectoderm[3]. The first case of accessory limb removal was reported by Jones and Larkin in 1889[4]. During embryogenesis the leakage of the proteinaceous neural tube fluid acts as an abundant source of Schwann cells, which de-differentiate into a lipomatous mass that may Sometimes produce

cartilage, bone, muscles, and nerves .Occasionally, these components may grow in an organized manner and develop into an accessory limb. Stimulus for this organized growth remains in the genitical make-up of the de differentiated cells. Two signaling centers are responsible. Apical ectodermal ridge (AER) and a second signaling center, the zone of polarizing activity (ZPA) in the posterior mesenchyme, Which secretes fibroblast growth factor and another critical secretory factor called Sonic hedgehog (Shh) by AER and ZPA respectively, abnormal concentrations of Shh or inhibition of cessation or downregulation of Shh thought to be the causes of abnormal limb growth at the site of spinal dysraphism.

CONCLUSION

Spinal dysraphism is not a rare disorder in India with a prevalence of 1.9/1000 live births. But spinal dysraphism presenting with a fully formed accessory limb (with ambiguous genitalia) is a rare scenario. With only 5 cases reported all over India till now. This was a closed type of dysraphism. Emergency surgical repair is not warranted in closed dysraphism. However, unnatural rural Customary beliefs tends to linger around the babies unusual deformity ,and the burden of societies rebuttal of their innocent child would tend the parents to prefer excision of accessory limb which has to be handled by the surgeon. Surgical excision of accessory limb and repair of spinal dysraphism will yield good results in such deformities.

REFERENCES

1. Bhide, P., Sagoo, G. S., Moorthie, S., Burton, H., & Kar, A. (2013). Systematic review of birth prevalence of neural tube defects in India. Birth Defects Research Part A: Clinical and Molecular Teratology, 97(7), 437-443.
2. Bayri, Y., Tanrıkulu, B., Ekşi, M. Ş., & Dağçınar, A. (2014). Accessory lower limb associated with spina bifida: case report. Child's Nervous System, 30, 2123-2126.
3. Gardner, W. J. (1980). Hypothesis: Overdistention of the neural tube may cause anomalies of non□neural organs. Teratology, 22(2), 229-238.
4. Jones, R., & Larkin, F. C. (1889). Removal of accessory limb and meningocele from the back of a child, and its anatomy. British medical journal, 2(1493), 310.