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Case Study www.wjahr.com

MRI DIAGNOSIS OF A CASE OF SACRAL MYELOCELE- AN UNCOMMON ENTITY IN CHIARI MALFORMATION TYPE II

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ABSTRACT

Although myelocele is known association with chiari malformation II, isolated myelocele with absent dermal or meningeal components is very rare. Preoperative accurate diagnosis by using MRI plays a pivot role in surgical management.

KEYWORDS: Myelocele, chiari malformation II, MRI.

INTRODUCTION

CASE REPORT

Clinical History

Three months old male baby presented with swelling in lower back and progressively increasing head size since birth. Antenatal history is significant for multiple congenital anomalies in family and detection of hydrocephalus in third trimester ultrasound scan. H/o NICU admission is 2nd postnatal day for seizures. Examination of lower back revealed soft to firm well defined tear drop swelling with no evidence of defect in overlying skin. (Image-1)

Patient is evaluated with Neurosonogram, USG of occipital region and lower back,

- Neurosonogram-Dilated ventricles (Image-2) and USG of occipital region revealed descending cerebellar herniation (Image-3) and lower back – flushed out neural placode with the skin at upper sacral region.(6a and 6b)

  Patient is further evaluated with plain MRI-Brain and MRI Spine.
- MRI Brain showed small posterior cranial fossa with concave clivus and beak like tectum, descent of cerebellum and straw like fourth ventricle with dilated third and lateral ventricles-suggestive of obstructive of hydrocephalus. (Image-4)
- MRI Spine revealed absence of convergence of posterior elements at S2-S3 level- spina bifida with low lying cord and neural placode flushing out with skin surface posteriorly. No fat or CSF density signal intensity areas between neural placode and skin. Thoracolumbar spine showed flow artifacts, mild cord compression and cord thinning. (Image-5a and 5b)

DISCUSSION

- The Chiari II malformation is a complex developmental deformity characterized by an elongated small cerebellum and brainstem with caudal displacement of the medulla, parts of the cerebellum and pons through an enlarged foramen magnum into the cervical spinal canal.[4] A meningomyelocele is nearly a constant accompanying feature. However it is uncommon to present with isolated myelocele compared to isolated meningomyelocele and meningocele.[5]
- Almost all neonatal patients with Chari II have myelomeningocele, it has been suggested that the underlying etiology - In utero CSF leak due to open spinal dysraphism. Majority of the patients with Chiari malformation type-II is diagnosed during routine antenatal ultrasound - shows dilated lateral ventricles and third ventricles with pointed appearance of occipital horns, abnormal size and shape of cerebellum. Shows ventriculomegaly with pointed appearance of occipital horns and abnormal size and shape of cerebellum. Meticulous ultrasound examination reveals defects in spine and adjacent soft tissue changes.[3]
- Externally myelomeningocele presents with defect in posterior aspect of body in midline with leakage of CSF and increased risk of infections. MRI is the assessment of choice because of its better investigative performance, exceptional soft tissue characterization and importance in presurgical

planning and does not involve ionizing radiation, has no biological risk.[2]

MR findings in Chiari-II:

- Brain manifestations- Telencephalon changes are frontal horn beaking, corpus callosum dysgenesis and third ventricle diverticulum. Enlarged third ventricle, large massa intermedia and elevation of hypothalamus as diencephalon changes and non visualization of aqueduct, bulbous tectum and beaked tectum are seen in mesencephalon. Rhombencephalon changes such as absent cerebellar folia or superior vermis or inferior vermis and cervicomedullary deformities, cerebellar vermial pegs and pyramidal hypogenesis. Mesodermal changes in the form of clival scalloping, gyral interdigitations, wide incisura and gyral indentations are obserbved.[4]
- Spinal cord manifestations such as wide anterior space, syringohydromyelia and spina bifida(meningomyelocele, meningocele and myelocele).[4]
- MRI offers several advantages in the evaluation of children with suspected spinal dysraphism and help in accurate diagnosis and type of spina bifida.[2] Although the postnatal clinical implications remain unclear, differentiating myelomeningocele from myelocele has important implications from a neurosurgical standpoint because skin closure may be technically more difficult for a myelocele and graft closure is often necessary.[1]
- Fetal MRI allows ready differentiation between myelomeningocele and myelocele. Ultrasound is also useful adjunct to confirm the diagnosis.[1] Though historically the postnatal clinical signs and symptoms have been described as nearly identical for the two entities, a higher incidence of scoliosis and high-risk bladder dysfunction was found in association with prenatally diagnosed myelocele compared with myelomeningocele. [1]

CONCLUSION

A rare association of isolated Myelocele in Chiari II malformation is accurately diagnosed by preoperative MRI with ultrasound as adjuvant by demonstrating spina bifida and low lying spinal cord and flushed out neural placode with skin. Myelocele repair is more technically challenging and it requires graft closure, hence accurate preoperative diagnosis is very necessary.

IMAGES

Image– 1:Description- Well defined tear drop like swelling pointed inferiorly visualized in sacral region.

Image- 2
Description - Neurosonogram (through anterior frontanalle) image showing dilated ventricles.

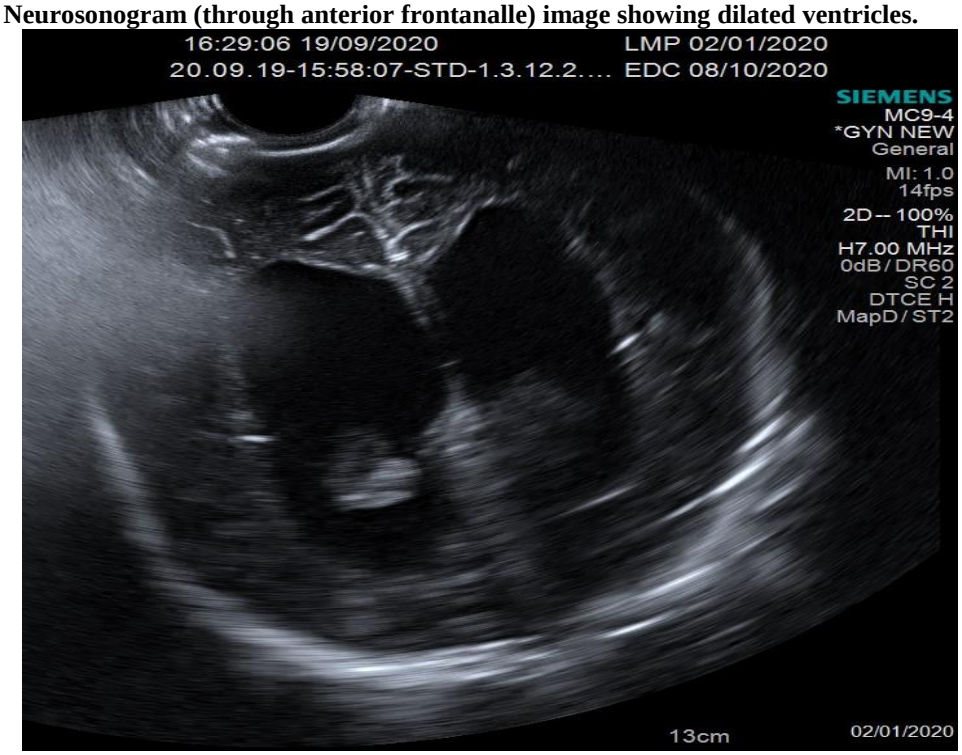


Image-3
Description- USG occipital region showing inferiorly herniating cerebellum into cervical canal.

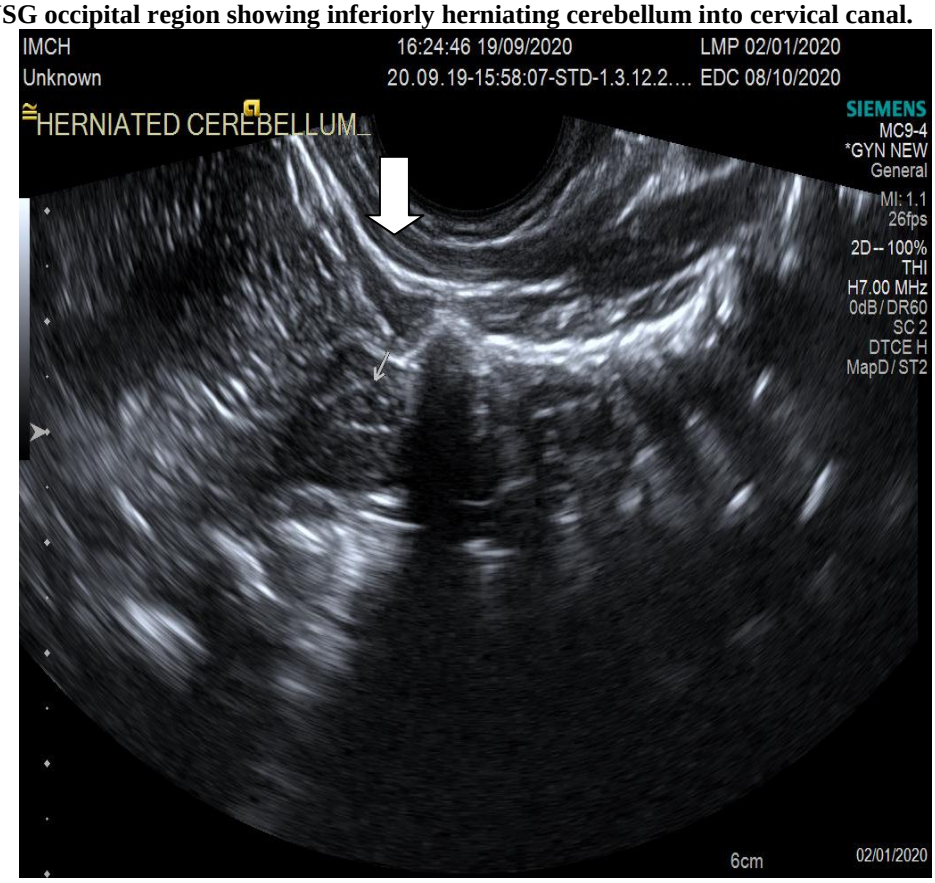


Image-4

Description- MR Brain screening section showing small posterior cranial fossa with concave clivus and beak like tectum. Descent of cerebellum and straw like fourth ventricle is observed with dilated third and lateral ventricles- suggestive of obstructive hydrocephalus.

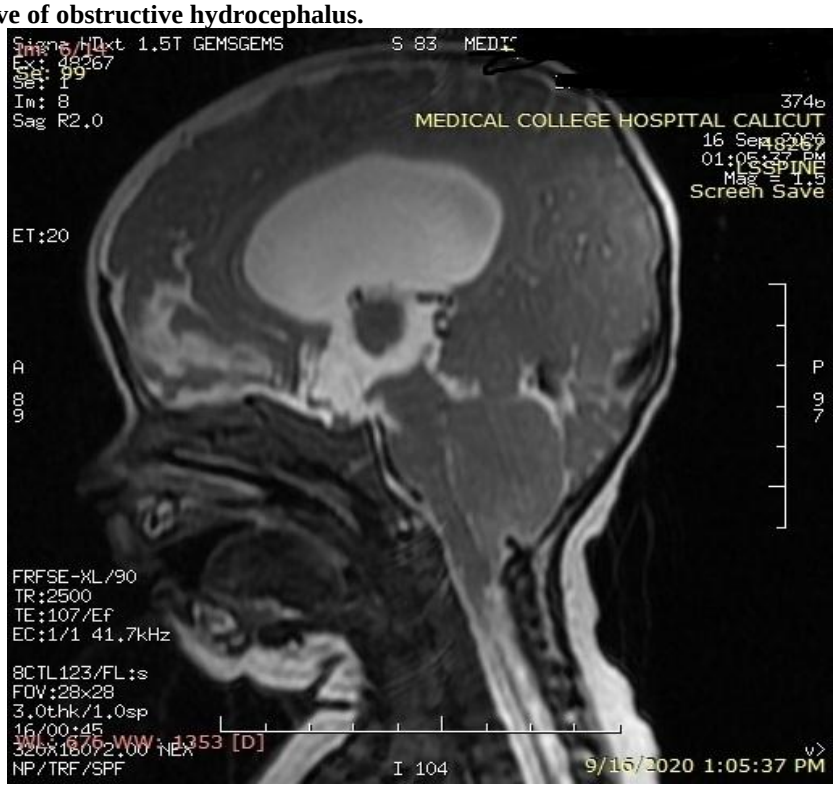


Image -5a and 5b

Description - MR -LS Spine showing absent convergence of posterior elements at S2-S3 level – spina bifida and low lying spinal cord with evidence of neural placode flushed out with skin posteriorly. No evidence of fat or CSF signal intensity observed between flushed neural placode and skin. Thoracolumbar spine shows flow artifacts, mild cord compression and cord thinning.

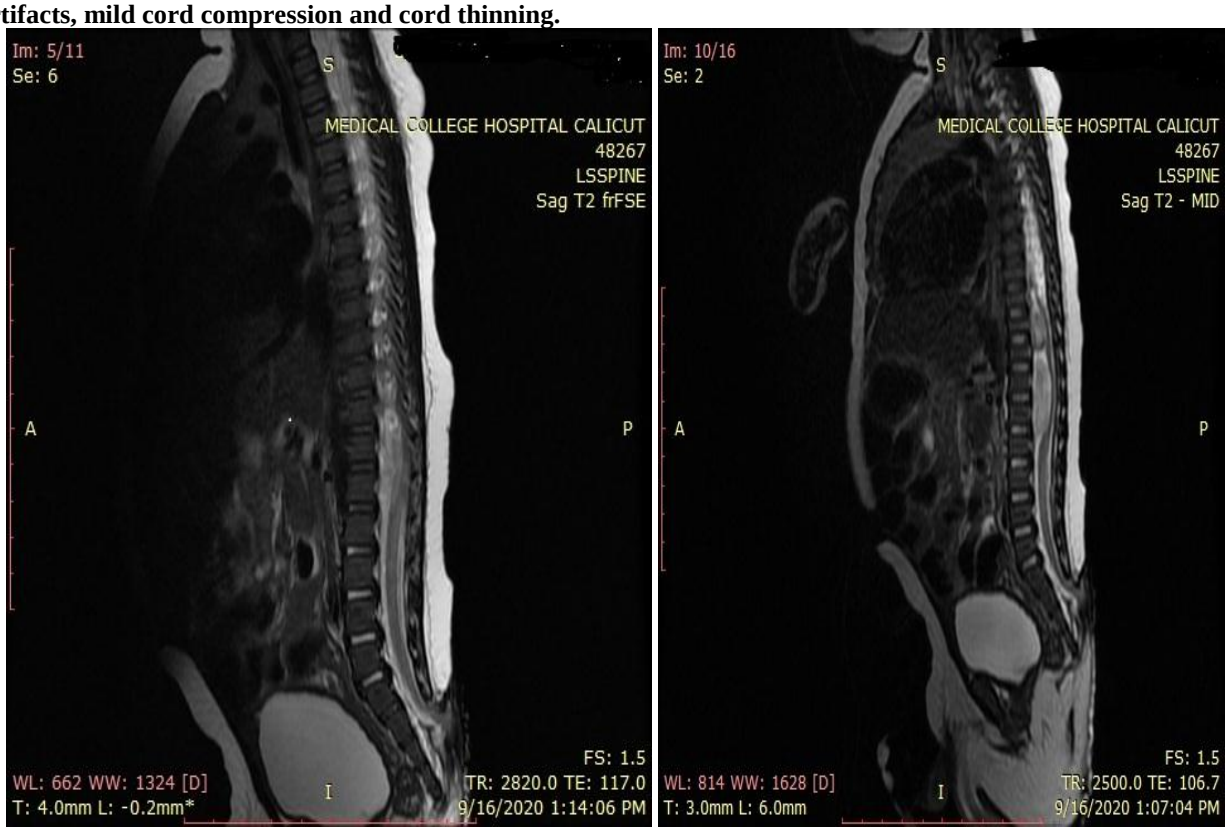
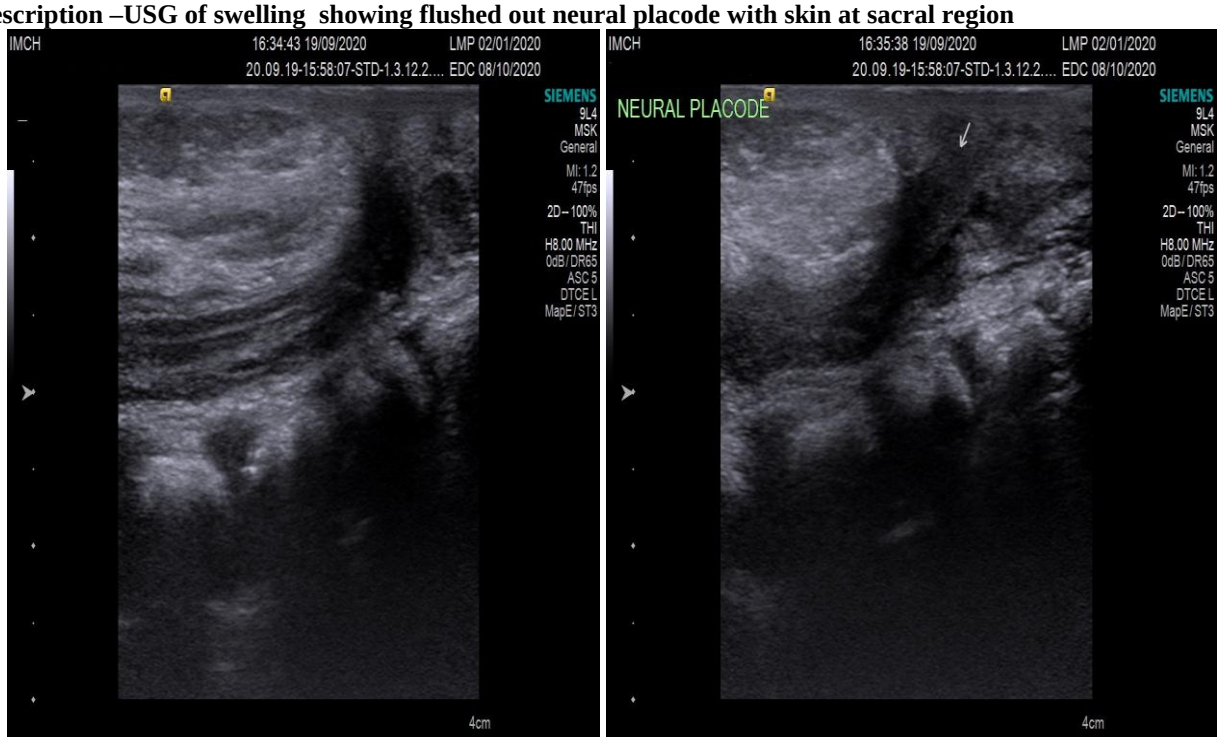


Image-6a and 6b
Description –USG of swelling showing flushed out neural placode with skin at sacral region



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