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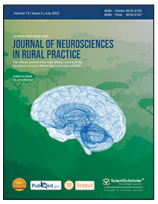

Case Report

Chiari III malformation with defect in Liliequist membrane on MR imaging

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ABSTRACT

The Liliequist membrane is a radiologically neglected structure, with routine evaluation only carried out in pre-operative and post-operative cases of third ventriculostomy. We report two cases of Chiari III malformation in two unrelated females with similar findings on magnetic resonance imaging study including occipital and low cervical encephalocele, hydrocephalus, and segmentation anomalies in cervical spine. Along with these findings, we report a flow void on T2-weighted images observed in both cases across the site of Liliequist membrane between interpeduncular and chiasmatic cistern. Our findings of CSF flow across the Liliequist membrane may represent spontaneous third ventriculostomy or another congenital defect in the myriad of anomalies seen in cases of Chiari III malformation.

Keywords: Chiari III, Liliequist membrane, Third ventriculostomy, Hindbrain malformation, Congenital

INTRODUCTION

Chiari III malformation is an extremely rare hindbrain deformity, characterized by occipital or low cervical encephalocele and osseous defect along with features of Chiari II malformation. Compared to other Chiari malformations, Chiari III malformation has a worse prognosis and relatively higher post-operative mortality.[1]

Liliequist membrane is a thin arachnoid membrane in subarachnoid space separating the chiasmatic and interpeduncular cistern. At present, its only established relevance is in cases of neurosurgical procedures wherein the Liliequist membrane is the site of endoscopic third ventriculostomy for management of hydrocephalus. Liliequist membrane is rarely evaluated on magnetic resonance imaging (MRI) in routine setting. Being thin (<1 mm in size), it is only appreciated on 3D CISS sequence. However, in cases of endoscopic third ventriculostomy, a flow void across the membrane is appreciated on spin echo T2-weighted sequences.[2]

Although osseous and brain deformities have been reported in cases of Chiari III malformation, spontaneous third ventriculostomy or congenital defect in Liliequist membrane has not been described previously.

Here, we report two cases having Chiari III malformation with segmentation anomalies in cervical spine and showing flow void across the site of Liliequist membrane.

CASE SERIES

Case 1 – An 18-month-old girl presented with irregular, gradually increasing occipital swelling since birth ultrasonologically diagnosed as meningoencephalocele. Another swelling was noted in right frontal region, which developed two months after head trauma due to fall, which was diagnosed as leptomeningeal cyst on computed tomography. Patient had never undergone any neurosurgical intervention. The patient was taken up for MRI for evaluation of the congenital defect. The MRI was carried out under general anesthesia.

MRI revealed defect in occipital bone with occipital meningoencephalocele measuring 4.5 × 2.5 cm along with crowding of foramen magnum, herniation of medulla into spinal canal, cervicomedullary kinking, tectal beaking, and gross hydrocephalus. In supratentorial area, the right frontal lobe porencephalic cyst herniating extracranially through defect in dura mater and frontal bone was identified suggestive of leptomeningeal cyst [Figure 1a].

Osseous deformities in skull included J-shaped sella, dorsal scalloping of clivus, and multiple segmentation anomalies

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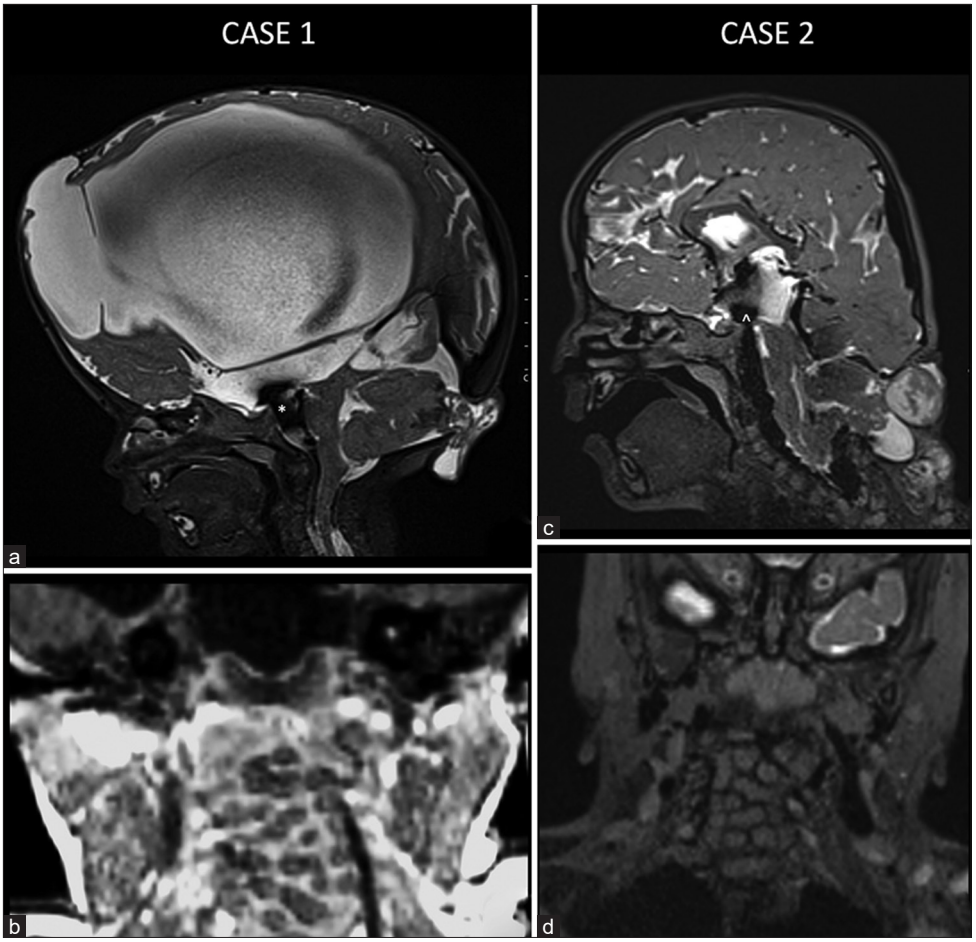

Figure 1: (a) Case 1 – Sagittal T2-weighted image – shows defect in occipital bone with occipital meningoencephalocele, and crowding of foramen magnum, gross hydrocephalus, and frontal leptomeningeal cyst. J shaped Sella and scalloping of clivus is also seen. Prominent flow void is noted in region of Liliequist membrane between interpeduncular cistern and third ventricle (*). (b) Case 1 – Coronal T1-weighted image revealing multiple segmentation anomalies in cervical spine. (c) Case 2 – Sagittal T2-weighted image revealing small posterior fossa with inferior herniation of brainstem and cerebellum along with high cervical encephalocele. Prominent CSF flow void is noted at the site of Liliequist membrane (^). (d) Case 2 – Coronal T2WI showing multiple incarcerated hemivertebrae in cervical spine.

in cervical and dorsal vertebrae including incarcerated hemivertebrae and partial block vertebrae [Figure 1b].

On heavily T2-weighted 3D sequences (T2 SPACE), flow void was identified at site of Liliequist membrane between chiasmatic and interpeduncular cistern, just anterior to basilar artery flow void [Figure 1a].

Case 2 – A 30-month-old girl presented with occipital swelling since birth, gradually increasing in size along with altered sensorium. Similar to the first case, MRI was carried out under general anesthesia.

MRI revealed high cervical encephalocele with herniation of dysplastic cerebellar vermis, cerebellar hemispheres, and meninges in suboccipital region. Small posterior fossa was noted with inferior displacement of brainstem and cerebellar hemispheres into spinal canal. Mild dilation of bilateral lateral and third ventricle was also noted [Figure 1c].

Osseous abnormalities included J-shaped sella and multiple segmentation anomalies in cervical spine [Figure 1d].

Similar to the Case 1, flow void on T2WI was identified between interpeduncular cistern and chiasmatic cistern, representing CSF flow through the site of Liliequist membrane [Figure 1c].

DISCUSSION

Chiari Type III malformation was first described by Hans Chiari in 1891. Accounting for < 1% of cases, it is the rarest hindbrain anomaly.[3]

Herniation of cerebellum in occipital or high cervical encephalocele is present in all cases of Chiari III malformation. Other than encephalocele, findings of Chiari II malformation are present in cases of Chiari III malformation including small posterior fossa and inferior herniation of cerebellum and brainstem.[1,4]

Osseous malformations include occipital bone defects, dorsal scalloping of clivus and posterior petrous pyramids, posterior cervical agenesis, and segmentation anomalies of cervical spine.[5]

Erol FS *et al.* in 2011 reported a case of Chiari III malformation with Klippel Feil syndrome in a 4-year-old boy.[6]

In our cases, one case had occipital encephalocele while other had high cervical encephalocele. Both encephaloceles contained dysplastic cerebellum along with meninges. Caudal herniation of brainstem was noted in both cases along with small posterior fossa.

Segmentation anomalies in cervical spine were identified in both cases with no craniovertebral junction abnormality.

Hydrocephalus is reported in 88% cases of Chiari III malformation.[7] We reported hydrocephalus in both cases, with gross hydrocephalus noted in one case. Prominent flow void of T2WI was noted in both cases between interpeduncular and chiasmatic cistern representing defect in Liliequist membrane. Congenital defect in Liliequist membrane has not been reported by the previous studies either in context of Chiari malformations or other congenital malformations.

Liliequist membrane is a radiologically neglected structure. Located under the floor of third ventricle, it is a thin arachnoid membrane, usually measuring < 1 mm. It divides the chiasmatic cistern from interpeduncular cistern and blocks CSF flow between them. Anatomically, it can be divided into sellar, mesencephalic, and diencephalic segments. Inferiorly and anteriorly, it attaches to dorsum sellae while superior posterior attachment of the membrane is either premamillary or retromamillary. Lateral attachments of the membrane are to the arachnoid sheath adjacent to oculomotor nerves.[2,8]

Neurosurgical studies have described physiological perforations in Liliequist membrane.[9] Cases of spontaneous third ventriculostomy have been reported in patients of chronic hydrocephalus.[10] Cavalheiro *et al*. in 2021 reported spontaneous third ventriculostomy in three cases of myelomeningocele and were able to demonstrate flow void across the site of Liliequist membrane on MRI in two cases.[11]

Our findings may represent spontaneous third ventriculostomy as a consequence of chronic hydrocephalus and altered CSF dynamics or may represent congenital defect due to abnormal mesenchymal development.

MRI in both the cases was performed under general anesthesia. Due to time constraints, CSF flow study was not performed in these cases. Further studies should be undertaken to evaluate the status of Liliequist membrane in cases of hindbrain malformations and chronic hydrocephalus.

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.

REFERENCES

1. Ivashchuk G, Loukas M, Blount JP, Tubbs RS, Oakes WJ. Chiari III malformation: A comprehensive review of this enigmatic anomaly. Childs Nerv Syst 2015;31:2035-40.
2. Fushimi Y, Miki Y, Ueba T, Kanagaki M, Takahashi T, Yamamoto A, *et al*. Liliequist membrane: Three-dimensional constructive interference in steady state MR imaging. Radiology 2003;229:360-5.
3. Cesmebasi A, Loukas M, Hogan E, Kralovic S, Tubbs RS, Cohen-Gadol AA. The Chiari malformations: A review with emphasis on anatomical traits. Clin Anat 2015;28:184-94.
4. Hiremath SB, Fitsiori A, Boto J, Torres C, Zakhari N, Dietemann JL, *et al*. The perplexity surrounding chiari malformations Are we any wiser now? AJNR Am J Neuroradiol 2020;41:1975-81.
5. Garg K, Tandon V, Mahapatra AK. Chiari III malformation with proatlas abnormality. Pediatr Neurosurg 2011;47:295-8.
6. Erol FS, Ucler N, Yakar H. The association of Chiari Type III malformation and Klippel-Feil syndrome with mirror movement: A case report. Turk Neurosurg 2011;21:655-8.
7. Young RM, Shafa JS, Myseros JS. The chiari 3 malformation and a systemic review of the literature. Pediatr Neurosurg 2015;50:235-42.
8. Dias DA, Castro FL, Yared JH, Lucas A Jr., Filho LA, Ferreira NF. Liliequist membrane: Radiological evaluation, clinical and therapeutic implications. Radiol Bras 2014;47:182-5.
9. Volovici V, Varvari I, Dirven CM, Dammers R. The membrane of Liliequist-a safe haven in the middle of the brain. A narrative review. Acta Neurochir (Wien) 2020;162:2235-44.
10. Abdulsalam HA, Nissiri F, Das S. Letter: Spontaneous third ventriculostomy in a patient following traumatic brain injury. Oper Neurosurg (Hagerstown) 2018;14:E26-7.
11. Cavalheiro S, da Costa MD, Dastoli PA, Mendonça JN, da Silveira Ximenes RL, Sarmento SG, *et al*. Spontaneous third ventriculostomy in patients undergoing fetal surgery for myelomeningocele correction. Childs Nerv Syst 2021;37:3429-36.

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