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Images in…

Occipital meningoencephalocele in a preterm neonate

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DESCRIPTION

A preterm (33 weeks) male neonate was born to a 34-year-old mother by caesarean section. The mother had a history of chronic hypertension, type 1 diabetes mellitus, stroke and asthma. Her medications included enoxaparin, insulin, nifedipine, methyldopa, inhaled albuterol and inhaled corticosteroids. She received regular prenatal care. The obstetric ultrasound done at 21 weeks of gestation revealed an occipital encephalocele, measuring ~3.3×2.0×2.5 cm in size and protruding through a 0.95 cm cranial defect in the occipital area. The baby had a spontaneous cry at birth with Apgar score of 9 and 9 at 1 and 5 min, respectively. His birth weight was 1650 g (10th–25th percentile). On physical examination, he was found to have a large (~5×4×5 cm in size), soft, skin-covered mass protruding from the occipital area (figure 1). MRI of the head done on day 3 of life revealed a large defect in the occipital bone with extra-axial fluid, meninges, brain matter and ventricles entering the defect (figure 2). A diagnosis of occipital meningoencephalocele was made. Reparative surgery was performed by the neurosurgery team at the age of 35 days. Postoperatively, he had uneventful recovery and was discharged home on full oral feeds on postoperative day 4. Encephalocele is a rare type of neural tube defect with an incidence of 1–4 cases per 10 000 live births.[1] Meningoencephalocele develops after the failure of normal midline fusion of cranial neural tube leading to a congenital bony defect through which brain and meninges herniate. Approximately 75% of the encephaloceles are located in the occipital region.[1] They usually contain a degenerative cerebral cortex, which is often excised during reparative surgery. If the sac contains normal brain tissue, techniques such as 'expansion cranioplasty' and 'ventricular volume reduction' have been described to preserve the herniated brain.[2 3] However, in most of the cases, herniated brain tissue is excised.

Figure 1 Large, soft, skin-covered mass protruding from the occipital area.

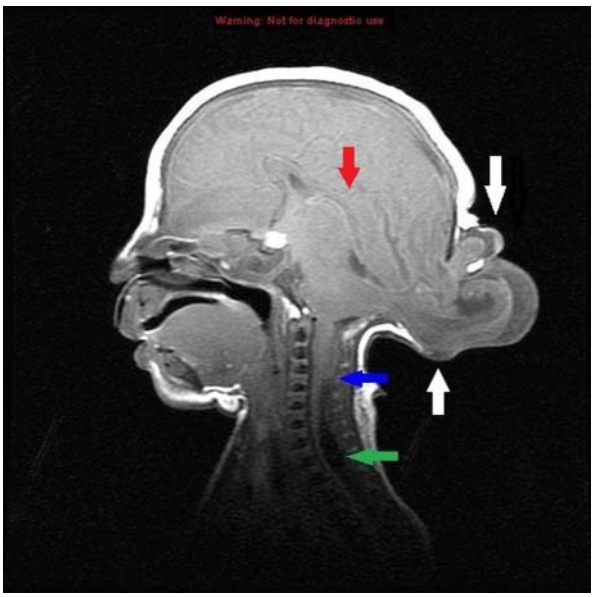

Figure 2 MRI of the head (sagittal T1-weighted midline image) showing a large occipital meningoencephalocele (white arrows) with herniation of the occipital lobe, occipital horn of the lateral ventricle and small dysmorphic cerebellar tissue into the sac. Other findings include dysgenetic corpus callosum with absent splenium (red arrow), small posterior fossa with dysmorphic cerebellar hemispheres and non-visualised fourth ventricle, herniation of the medulla into the cervical region with cervicomedullary kink (blue arrow) and a small syrinx in the cervicothoracic region (green arrow).

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Learning points

- Occipital meningoencephalocele is a very rare type of neural tube defect.
- Preoperative MRI provides useful information to assess prognosis and plan the surgical management of occipital meningoencephalocele.
- Despite the surgical management, prognosis remains poor with an extremely high risk of mortality and morbidities including mental and/or physical impairment.

Competing interest None.

Patient consent Obtained.

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