

Schizencephaly on fetal MRI

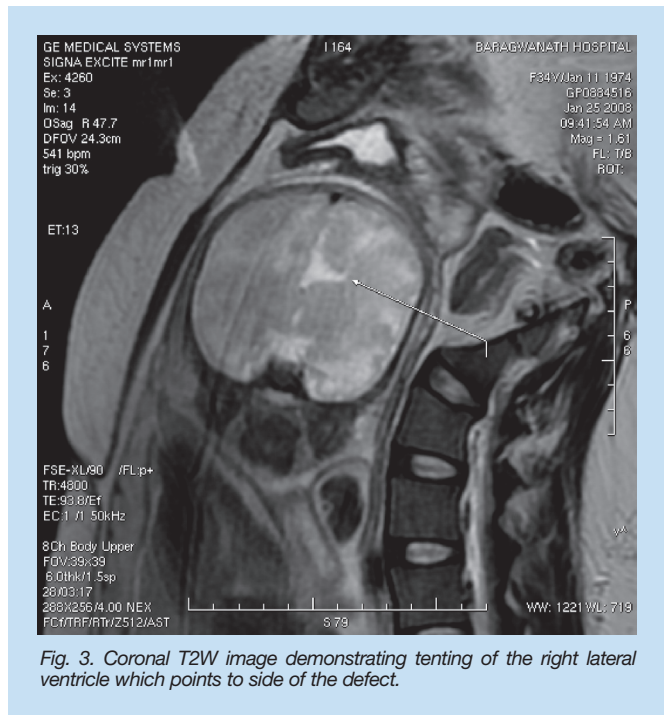
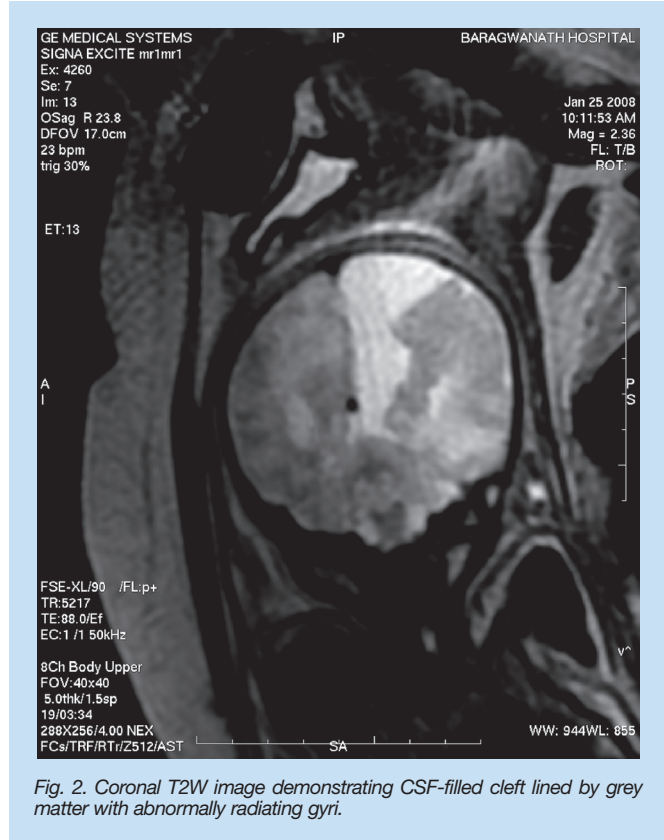
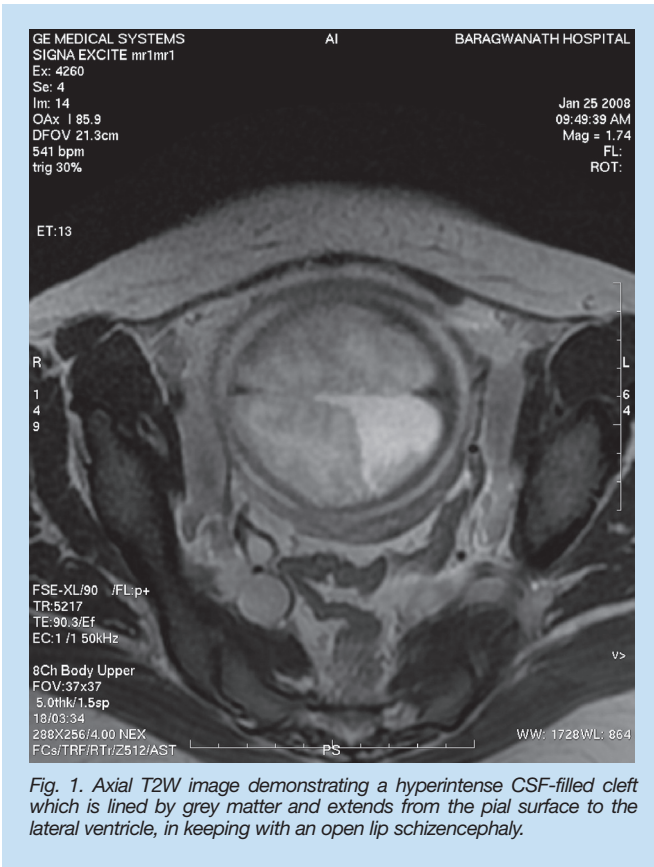
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Abstract

Schizencephaly is a rare CNS malformation consisting of a full-thickness CSF-filled parenchymal cleft lined by grey matter. Schizencephaly can be diagnosed on prenatal ultrasound but requires magnetic resonance imaging (MRI) for more specific diagnostic differentiation from other cerebrospinal fluid (CSF)-containing structures as well as for detection of associated features.

Introduction

Schizencephaly is a rare CNS malformation consisting of a full-thickness CSF-filled parenchymal cleft lined by grey matter (pathognomonic) extending usually from the subarachnoid space to the subependyma of the lateral ventricles. The cleft is usually situated in the region of sylvian



fissure and can be unilateral or bilateral, or can be closed lip (type 1) or open lip (type 2).¹ The cleft also has multiple abnormally radiating gyri and sulci that help to differentiate it from a perinatal infarct.

Schizencephaly can be diagnosed on prenatal ultrasound but requires MRI scanning for more specific diagnostic differentiation from other CSF-containing structures as well as for detection of associated features.²

MRI-associated features include:^{2,4}

- absent cavum septum pellucidum (80 - 90%)
- focally thin or absent corpus callosum
- polymicrogyria and heterotopias
- optic nerve hypoplasia
- roofing membrane covering the defect
- tenting of the ventricle, pointing to the defect.

Schizencephaly can be differentiated from porencephaly on MRI by the fact that porencephalic cysts are lined with white matter.²

Clinical details

We describe here a 26-year-old woman patient at >36 weeks' gestation, who presented to the antenatal clinic of Chris Hani Baragwanath Hospital. A fetal ultrasound was performed and a cystic lesion in the posterior fossa was suspected. A fetal MRI (fMRI) was requested to confirm the diagnosis and to assist in showing any other associated findings. The fMRI was done on a General Electric 1.5 tesla MR scanner using a phased array abdominal coil (due to the patient's advanced gestational

age). T1- and T2-weighted images, in 2 orthogonal planes only, were obtained, as the mother was uncomfortable. The fMRI findings were:

- a CSF-filled cleft lined by grey matter extending from the subarachnoid space from the right parietal lobe region communicating with the trigone and occipital horns of the right lateral ventricle
- an associated malformation of the right parieto-occipital lobe
- tenting of the right frontal horn
- the frontal lobes appeared normal
- the left cerebral hemisphere was normal
- the corpus callosum was present
- the cavum septum pellucidum was not visualised
- the Circle of Willis was normal in appearance
- the optic nerves appeared grossly normal.

Discussion and conclusion

Schizencephaly can be detected on prenatal ultrasound; however, for better differentiation of CSF-containing abnormalities of the fetal brain and detection of associated anomalies, fMRI is more sensitive and specific.³

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3. Oh KY, Kennedy AM, Frias Jr AE, Byrne JLB. Foetal schizencephaly: pre- and postnatal imaging with a review of the clinical manifestations. *Radiographics* 2005; 25: 647-657.
4. Sutton D. *Textbook of Radiology & Imaging*. Vol 2. 7th ed. Paris: Lavoisier Publishing, 2002: 1734.