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Teaching NeuroImages: Syntelencephaly

Middle interhemispheric fusion

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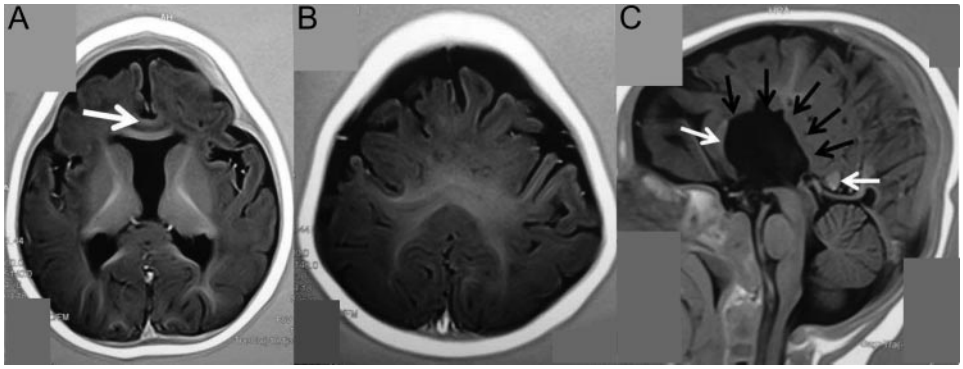
Figure Syntelencephaly: MRI appearance



Axial T1-weighted inversion recovery (IR) sequence shows absence of septum pellucidum with fusion of the cingulate gyrus (white arrow) (A) and continuous white matter across middle part of interhemispheric region (B). Sagittal T1-weighted IR sequence (C) with absence of midbody of corpus callosum (black arrows) and presence of genu and splenium (white arrows).

An 8-month-old infant presented to our hospital with global developmental delay. MRI brain findings revealed the diagnosis of syntelencephaly (figure, A–C).

Syntelencephaly, a distinct subset of holoprosencephaly,[1] is a rare brain malformation in which the hemispheric fusion does not occur at rostral forebrain but rather across posterior frontal region; hence the alternative term "middle interhemispheric fusion" or "dorsal lobar holoprosencephaly." The clinical presentations can vary, with mental defect or global developmental delay being common.[2] The septum pellucidum is absent in all cases. Unlike classic holoprosencephaly, typically the midbody of corpus callosum is deficient in this disorder.

AUTHOR CONTRIBUTIONS

Dr. Arora: study concept and design, drafting and revision of manuscript. Dr. Kumar Sahoo: drafting and revision of manuscript. Dr. Srivastava: study concept and design, drafting and revision of manuscript, guarantor of study.

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2. Coleman LT, McCubbin JP, Smith LJ, Reddihough DS, Gardner RJM. Syntelencephaly presenting with spastic diplegia. Neuropediatrics 2000;31:206–210.

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