RESIDENT & FELLOW SECTION

Teaching NeuroImages: Radiologic features of septo-optic dysplasia plus syndrome

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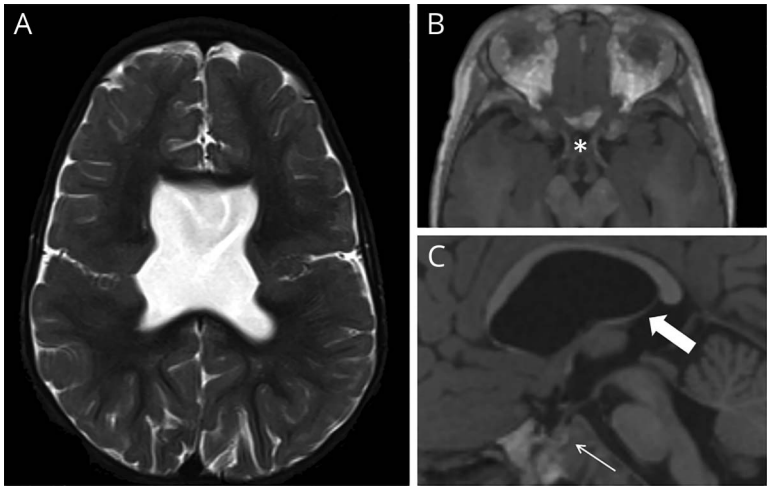
Figure MRI brain



(A and B) T2/T1-weighted axial views. (C) T1-weighted sagittal views.Multiple intracranial abnormalities are identified: (A) absent septum pellucidum and bilateral closed-lips schizencephaly; (B) achiasmia (asterisk); (C) pituitary hypoplasia (thin arrow), horizontal course of the fornix (thick arrow), and hypoplastic genu and splenium of corpus callosum.

A 17-month-old boy showing features of global developmental delay and visual impairment, despite physiologic head circumference growth (between the 50th and 85th centile), was admitted for investigations. On examination, he appeared alert, with pale optic discs, horizontal jerk nystagmus, spasticity, and small genitalia.

MRI brain (figure) excluded hydrocephalus, demonstrating instead hallmarks of neuronal migration disorder.[1] Besides schizencephaly, multiple midline abnormalities were noticed, such as optic nerve hypoplasia and achiasmia (deficient chiasmal decussation), pituitary hypoplasia, and absence of septum pellucidum.[1,2] Hormonal tests demonstrated panhypopituitarism; hence care was transferred to the septo-optic dysplasia (SOD) multidisciplinary group for management of SOD plus syndrome.[1]

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Author contributions

M. Ganau: study concept and design. M. Ganau and Dr. Talenti: analysis and interpretation. Dr. D'Arco: acquisition of data, critical revision of the manuscript, study supervision.

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