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Neuro*Images*

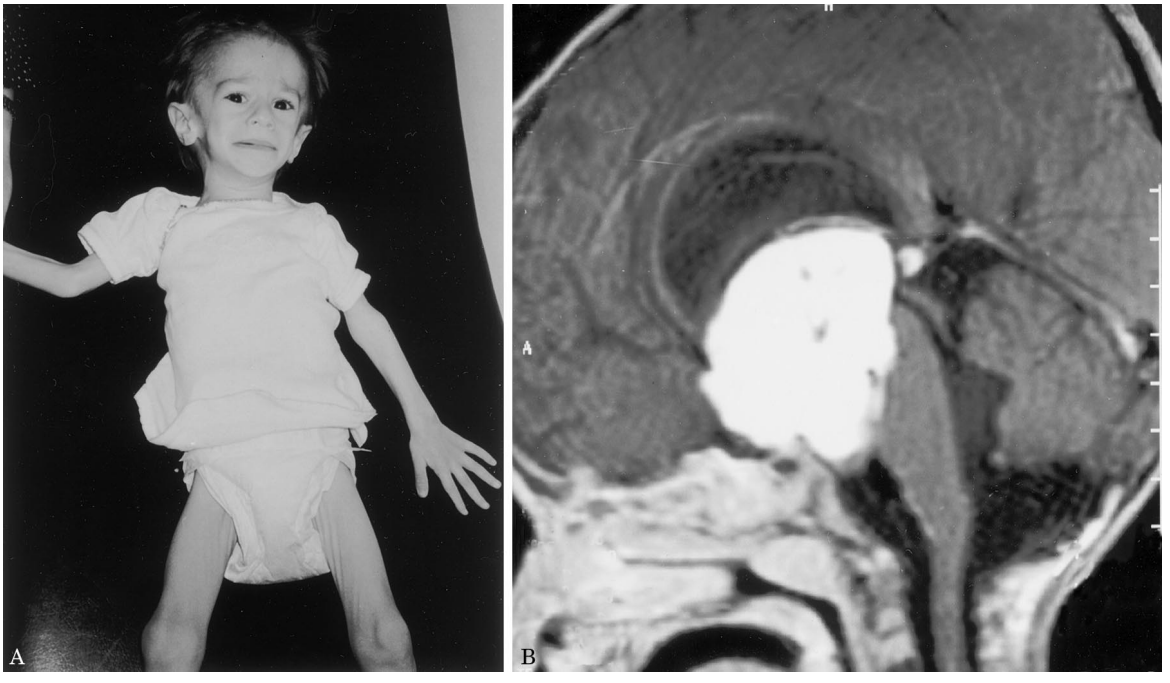

Figure. (A) The index patient. Note the severe emaciation of the whole body and the characteristic "pseudohydrocephalic" appearance. (B) MRI of the brain. T1-weighted sagittal images (repetition time/echo time: 570/15) after gadolinium enhancement demonstrate the presence of a large tumor involving the hypothalamic region, distorting the chiasm and brainstem, and extending into the third ventricle. Neuropathologically, the tumor proved to be a hypothalamic astrocytoma with pilomyxoid features.

Russell's diencephalic syndrome

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A 21-month-old boy was initially admitted to our hospital because of failure to thrive, which began at the age of 6 months. Physical examination revealed emaciation (weight < third percentile), normal body length, normal head circumference with a characteristic "pseudohydrocephalic" face (figure, A), as well as a mild pyramidal tract dysfunction, in an otherwise alert child with limited speech but with appropriate cognitive abilities for his age. An MRI of the brain was performed, which revealed a large, partly solid, partly cystic mass in the suprasellar region, distorting the chiasm and brainstem and extending well up through the hypothalamus and into the third ventricle (see the figure, B); the above-mentioned tumor neuropathologically proved to be an astrocytoma with pilomyxoid features.

Russell's diencephalic syndrome, which is manifested by progressive emaciation and failure to thrive in an apparently alert infant, usually is due to a low-grade hypothalamic astrocytoma.[1,2] The normal head circumference despite the severe loss of adipose tissue of the face sometimes gives the impression of a characteristic pseudohydrocephalic appearance.

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Russell's diencephalic syndrome

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