Prune belly and schizencephaly

Figure T1-weighted MRI and a photograph of the characteristic abdomen

(A–C) Discrete global brain atrophy, ventricular system asymmetry, left hemisphere schizencephaly, and bilateral heterotopias. (D) Wrinkly folds of skin covering the abdomen due to thin abdominal wall muscles, a major feature of the prune belly syndrome.

A 45-year-old man without relevant family history presented with prune belly syndrome (PBS).¹ He had a 34-year history of right simple partial motor seizures with and without secondary generalization, with a good response to carbamazepine monotherapy; there was no cognitive or social impairment. MRI showed global brain atrophy, ventricular asymmetry, left hemisphere schizencephaly, and bihemispheric heterotopias (figure, A–C). Although PBS is considered a mesoderm layer defect characterized by total or partial absence of abdominal muscles (figure, D), urinary tract abnormalities, and cryptorchidism of unknown etiology, involvement of other embryologic tissues has also been reported, including the ectodermic layer, as in this case.^{1,2}

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