A 45-year-old man without relevant family history presented with prune belly syndrome (PBS). 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

Guillermo A. Navarro-Arenas, MD, Diego R. Orozco-Valera, MD, Erwin Chiquete, MD, PhD, José L. Ruiz-Sandoval, MD

From the Hospital Civil de Guadalajara “Fray Antonio Alcalde” (G.A.N.-A., D.R.O.-V., J.L.R.-S.); the Centro Universitario de Ciencias de la Salud (J.L.R.-S.), Universidad de Guadalajara; and the Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán (E.C.), Mexico City, Mexico.

Author contributions: Dr. Navarro-Arenas: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and final approval. Dr. Orozco-Valera: drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval. Dr. Chiquete: drafting/revising the manuscript, accepts responsibility for conduct of research and final approval, acquisition of data.

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Correspondence to Dr. Ruiz-Sandoval: jorulej-1nj@prodigy.net.mx

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