Characteristic imaging findings in encephalocraniocutaneous lipomatosis

Figure 1 Brain MRI

Brain MRI at age 21 years shows left cerebral hemiatrophy (A) and arachnoid cysts (B) in the left frontal and temporal regions.

Our patient was worked up for mild developmental delay associated with focal scalp alopecia at age 6, and was diagnosed with encephalocraniocutaneous lipomatosis (also known as Haberland or Fishman syndrome), a rare neurocutaneous syndrome characterized by skin lesions (most commonly focal alopecia with or without underlying lipoma), ocular lesions, and CNS anomalies.¹ CNS anomalies are usually seen ipsilateral to the skin lesions, including cerebral hemiatrophy (figure 1A), intracranial and spinal lipomas, arachnoid cysts (figure 1B), brain malformation, and vascular defects.¹ Leptomeningeal lipomatosis (figure 2A) is an uncommon finding, but can be characteristic.² Vessel anomalies include leptomeningeal angiomas, variations in arterial caliber (figure 2B), and excessive vessels.

Figure 2 CT angiogram of the brain

CT angiogram at age 21 years shows leptomeningeal lipomatosis along cortical veins (A) and tangle of excess vessels in the left parietal region (arrow) with dolichoectasia of a M2 branch of the middle cerebral artery (arrowhead) (B).

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