Extensive subarachnoid venous angiomatosis with hydrocephalus in phacomatosis pigmentovascularis

An 8-month-old boy with cutaneous vascular malformations and dermal melanocytosis (Mongolian spots, figure, A) on the face and trunk was diagnosed with phacomatosis pigmentovascularis type 2. He had normal neurodevelopment, but progressive macrocephaly (figure, B). Linear brain ultrasonography showed extensive venous angiomatosis in the prominent subarachnoid space (figure, C and D). MRI revealed cortical sulcal widening, prominent leptomeningeal vessels in an enlarged subarachnoid space (figure, E and F), and communicating hydrocephalus (figure, F). Neurologic involvement in phacomatosis pigmentovascularis is uncommon except in Sturge-Weber and Klippel-Trenaunay syndromes. Communicating hydrocephalus due to subarachnoid angiomatosis may be underdiagnosed in phacomatosis pigmentovascularis, and should be considered in cases of progressive macrocephaly.

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