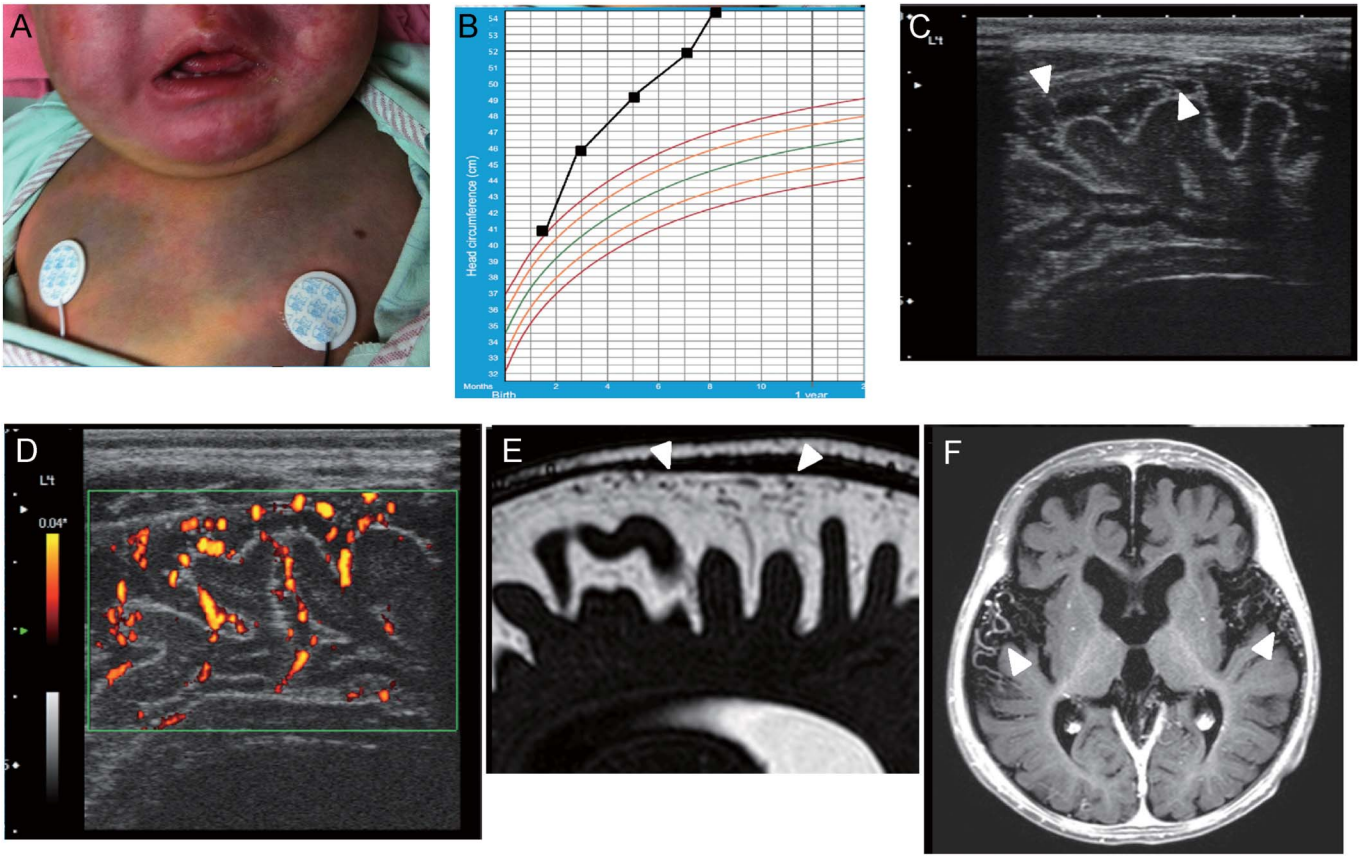
Extensive subarachnoid venous angiomatosis with hydrocephalus in phacomatosis pigmentovascularis

Figure Cutaneous findings, head circumference changes, and neuroimages



Nevus flammeus and pigmentary abnormalities (A). Progressive macrocephaly (B). Prominent vasculatures in the subarachnoid space (white arrowheads) under linear (C) and power Doppler sonography (D) and 3D steady-state acquisition sequence MRI (E). Clustered leptomeningeal vessels in the enlarged subarachnoid space and nonobstructive ventriculomegaly on enhanced T1-weighted image (F).

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.[1,2] 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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