A 4-year-old boy presented with a 1-week history of left eye proptosis. Examination showed multiple café-au-lait spots and impaired left eye vision. MRI revealed left optic nerve glioma with perineurial arachnoid gliomatosis (PAG) (figure, A–C). The growth patterns of optic nerve gliomas are classified into intraneural and perineural forms. The intraneural form is characterized by fusiform enlargement of the nerve, whereas PAG shows astrocytic proliferation in the subarachnoid space surrounding the relatively preserved optic nerve. Rarely, both forms can coexist, as in this case. PAG should be differentiated from optic nerve sheath meningioma, which is more common in patients with neurofibromatosis 2.
Teaching NeuroImages: Optic nerve glioma with perineural arachnoid gliomatosis in a patient with neurofibromatosis-1
K. Srinivasan and Bejoy Thomas
Neurology 2015;84:e97
DOI 10.1212/WNL.0000000000001424

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