Progressive supranuclear palsy motor phenotype in a patient with pineocytoma

Figure MRI of the pineal lesion

(A) Axial T2-weighted image demonstrates an extra-axial right paramedian mass of the pineal region, with a “bubbly” aspect, with midbrain distortion and aqueduct compression. (B) Sagittal, contrast-enhanced, T1-weighted image shows no enhancement, in keeping with a nonaggressive behavior.

A 76-year-old man developed gait disorder with falls over the course of 2 years. On neurologic examination, he had apraxia of eyelid opening, vertical supranuclear gaze palsy, dysarthria, and dysphagia. There was facial hypomimia, moderate axial rigidity, symmetrical bradykinesia, and no tremor. Gait was characterized by freezing and postural instability. Cognitive assessment was normal. Brain MRI revealed a multiloculated lesion of the pineal region causing midbrain (tectal plate) distortion (figure). Histology was compatible with pineocytoma. Descriptions of clinical pictures resembling progressive supranuclear palsy caused by midbrain tumors are historical,¹ especially by a rare tumor in adulthood.

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