A 50-year-old man presented with 9-years history of progressive ataxia. His father had undiagnosed ataxia. Examination showed ataxia and pyramidal signs. Brain MRI: olivoponto cerebellar atrophy; spine MRI: global spinal cord atrophy (Figure). Genetic test confirmed spinocerebellar ataxia type-1 (SCA1).

SCA1 is characterized by cerebellar ataxia with variable degrees of ophthalmoplegia, pyramidal signs, and peripheral neuropathy. Spinal cord atrophy was described in other SCA subtypes, but not in SCA1. Prominent pyramidal signs and spinal cord atrophy in SCA1 may be explained by long tracts involvement, as in hereditary spastic paraplegia. Spinal cord atrophy must be considered in neuroimaging features related to SCA1.

**References**

