Superficial siderosis (SS) of the central nervous system (SS) is a rare disorder that results from chronic hemosiderin deposition in the subpial layers of the brain and the spinal cord. Although recurrent bleeding in the subarachnoid space is the most likely explanation, a definite history of subarachnoid hemorrhage (SAH) is often lacking. Among the clinical presentations described in the literature include sensorineural deafness, dementia, anosmia, pyramidal tract signs and cerebellar ataxia. However, due to its rarity, SS remains one of the least considered differential diagnosis in patients with sporadic ataxia syndrome. We describe a case of progressive gait imbalance that was initially misdiagnosed for several years until a brain MRI study showed evidence of diffuse hemosiderin deposition suggestive of SS of CNS.