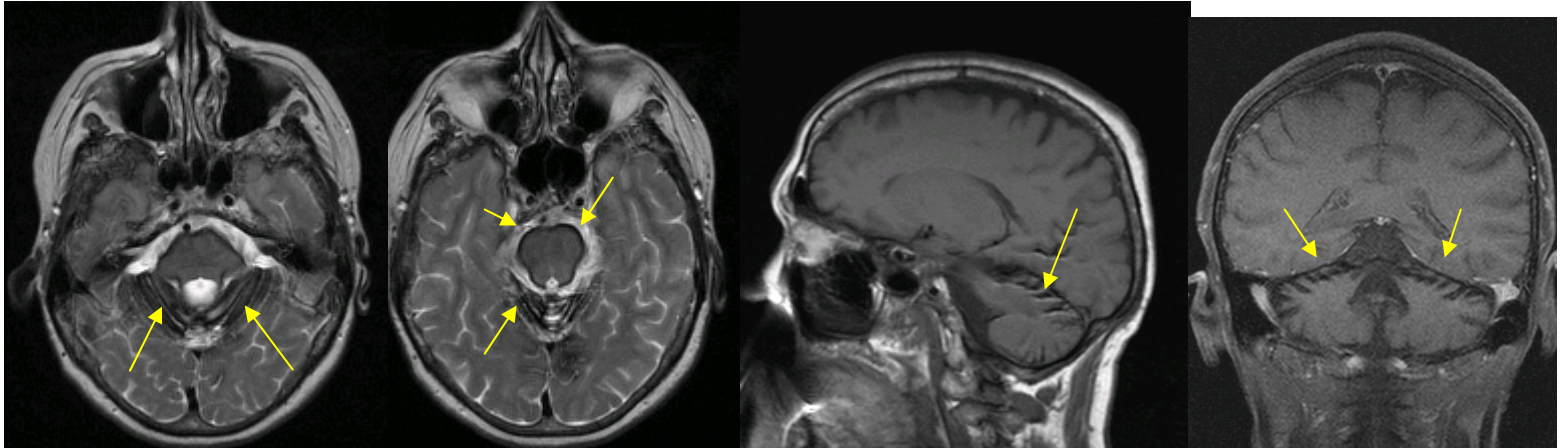


**INTERESTING CASE: SUPERFICIAL SIDEROSIS**



**Clinical Presentation:** This is a 55-year-old man who was referred to AIC for tinnitus and hearing loss. An MRI of the brain and internal auditory canals (IAC's) were performed.

**Imaging Findings:** There is unusual dark signal intensity over the leptomeninges, particularly in the posterior fossa over the cerebellar folia and around the cerebellar vermis (arrows). There is also a rim of dark hypointensity around the brainstem and other subarachnoid spaces. Similar findings were also noted in the interhemispheric fissure of the supratentorial region. There is cerebellar atrophy. The IAC's revealed no mass lesion.

**Diagnosis:** These findings are suggestive of **SUPERFICIAL SIDEROSIS**.

**Discussion:** Superficial Siderosis is a radiologic-pathologic condition characterized by a classic triad of sensorineural hearing loss, cerebellar ataxia, and myelopathy. It results from a prolonged chronic subarachnoid hemorrhage (SAH) with chronic hemosiderin (a blood breakdown product) deposition over the subpial layer of the subarachnoid CSF space in the brain and spine. Most cases are idiopathic, followed by traumatic origin and other CNS conditions resulting in prolonged SAH. Symptoms include unilateral or bilateral hearing loss in 81% of cases; cerebellar ataxia; myelopathy (such as extremity weakness or spasticity) in 53% of cases. Only 39% of patients, however, have all three of these symptoms.

**Treatment:** Superficial siderosis is a progressively deteriorating neurological condition that must be recognized early in order to be effectively treated. Once symptoms have appeared, the goal of therapy should be to prevent progression of the condition by removing the source of subarachnoid hemorrhage when it can be identified and corrected. In the rare instance that superficial siderosis is diagnosed by MRI in an asymptomatic person, a work-up should be pursued for the source of bleeding. Unfortunately, there is no therapy to reverse the damaging effects of hemosiderin deposition in the brain. The most commonly attempted therapy is iron chelation medications. Tin protoporphyrin, trientine and CSF shunting are other investigational therapies.

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**REFERENCE:** Michael Levy, Christine Turtzo and Rafael H Llinas. Superficial siderosis: a case report and review of the literature. Nature Clinical Practice Neurology (2007) 3, 54-58. Johns Hopkins University.