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SUSAC'S SYNDROME: A CLINICAL AND RADIOLOGICAL CHALLENGE

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We describe a patient with Susac's syndrome presenting with the triad of encephalopathy, branch retinal artery occlusion and hearing loss. MRI has some characteristic features and can be helpful in establishing the diagnosis, particularly in an early stage before the classic triad is complete. Besides MRI often allows to differentiate this rare disorder from more common neurologic diseases like multiple sclerosis.

Key-word: Brain, diseases.

Case report

A 34-year-old woman presented initially with acute vertigo. MRI of the brain at this stage was normal. Two months later, still suffering from persistent instability, she developed headache and a visual field scotoma in the right eye. Fluoangiography demonstrated an occlusion of a retinal branch artery. In spite of treatment with steroids she presented two months later with bilateral deafness, tinnitus, increasing headache and confusion. At that time the diagnosis of Susac's syndrome (SS) became clear. Extensive laboratory studies, including CSF electrophoresis for oligoclonal bands, were negative. Pd and T2 weighted MRI demonstrated linear and nodular hyperintense lesions in the central part of the corpus callosum (CC) (Fig. 1A, B), and hyperintensities in the crus posterius of the left internal capsula (Fig. 1C). On subsequent studies small T1 hypointense lesions became visible in the CC (Fig. 1D). Gadolinium administration visualised leptomeningeal enhancement on T1 and FLAIR imaging (Fig. 2A, B). Treatment with steroids and cyclophosphamide controlled the disease activity only partially and had to be continued for two years.

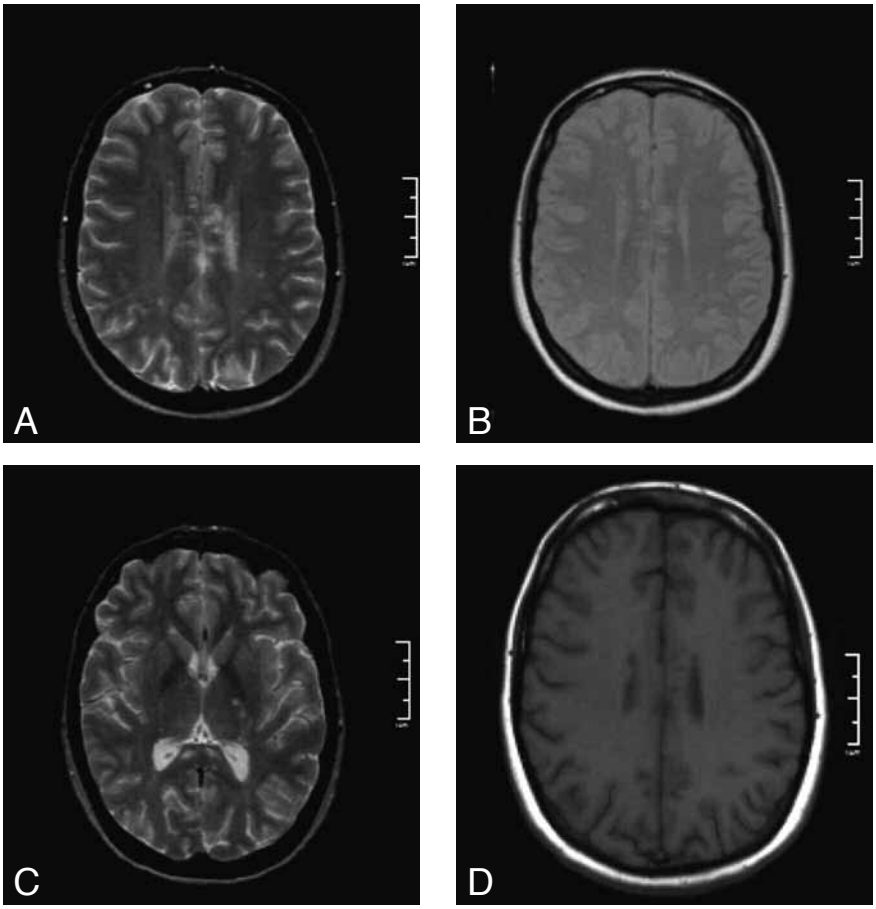

Fig. 1. — A and B. Axial Pd and T2 imaging showing small linear ('spokes') and large nodular ('snowballs') hyperintense lesions in the central part of the corpus callosum. C. Axial T2 with multiple nodular hyperintensities in the crus posterius of the internal capsula ("string of pearls"). D. Axial T1 demonstrating small hypointensities in the splenium of the corpus callosum (callosal holes).

Discussion

SS is a rare microangiopathy of unknown etiology, causing microinfarcts in the brain, the retina and the inner ear. Those are responsible for the clinical triad of encephalopathy, visual scotomas, vertigo and hearing loss (1). Encephalopathy is characterized by headache, confusion, behavioral changes, memory loss, ataxia and pyramidal dysfunction (2). Once the triad completed, diagnosis becomes relatively easy, but all the elements are almost never present from onset (3). Auditory or visual disturbances are often the initial presentation. Young women between 20 and 40 years are most often afflicted, but SS is not limited to this age group nor to the female gender (4). Although SS has long been considered a monophasic and spontaneously remitting disease, recent reviews distinguish also episodic and chronic variants (1). Immoglobulins, immunosuppressants or rituximab are indicated if disease activity cannot be controlled with steroids alone (1). Although the diagnosis of SS remains essentially clinical, MRI is important for confirmation. Besides MRI characteristics allow differential diagnosis with

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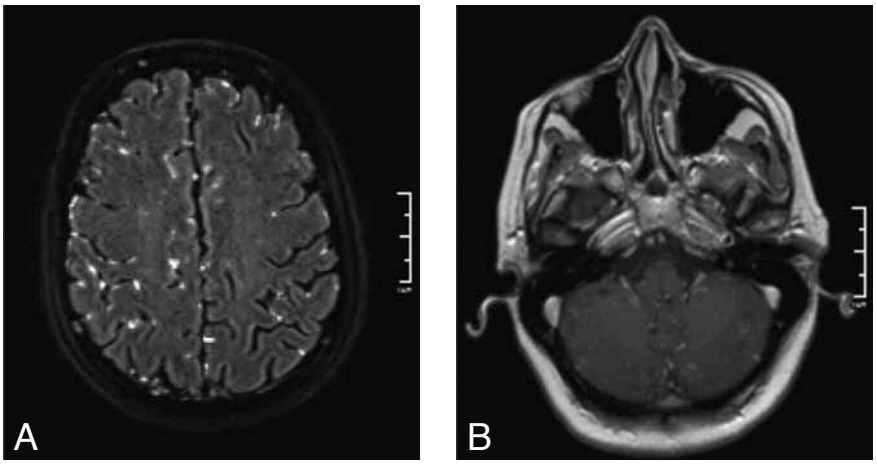

Fig. 2. — Axial FLAIR (A) and T1 after Gadolinium (B) showing leptomeningeal enhancement.

multiple sclerosis (MS) and acute disseminated encephalomyelitis (ADEM), which can be confused with SS (1).

The most striking MRI findings, especially in the encephalopathic form of SS, are microinfarcts presenting as small T2 and FLAIR hyperintensities in the central part of the CC, sparing the periphery and becoming T1 hypointense after the acute stage. Many of them are linear and radiating, so called "spokes", and are best seen on sagittal images (1, 4). "Snowballs" are larger centrally located CC lesions, evolving into "callosal holes". They can be accompanied by microinfarcts in the capsula interna, visible on DWI and sometimes on axial FLAIR as a "string of pearls". The combination of this central callosal lesions and the "string of pearls" is pathognomonic for SS (1). MS and ADEM on the contrary involve the undersurface of the CC and create ovoid or oblong plaques at the callososeptal interface with periventricular extensions into the deep white matter (2). Leptomeningeal enhancement is an important finding because it is often present in SS, but absent in MS and ADEM (1).

Conclusion

In patients with Susac's syndrome, MRI is often useful in establishing the correct diagnosis, even before the classic triad is completed. The radiologist should be aware of SS and include it in the differential diagnosis when T2 and FLAIR weighted imaging show hyperintense 'spokes' or 'snowballs' in the central part of the CC, when DWI and FLAIR demonstrate capsular microinfarcts like a 'string of pearls', and in case of meningeal enhancement.

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