# Multiple sclerosis or Susac syndrome – can you tell the difference?

Josefine Britze[1] (main author), Marianne Wegener[2], Rikke Marie Jensen[3], Jette Lautrup Frederiksen[1]

[1] *The Multiple Sclerosis and Optic Nerve Clinic, Dept. of Neurology, Rigshospitalet-Glostrup, Copenhagen University Hospital, Denmark*
[2] *Dept. of Ophthalmology, Rigshospitalet-Glostrup, Copenhagen University Hospital, Denmark*
[3] *The Multiple Sclerosis Clinic, Dept. of Neurology, Rigshospitalet-Glostrup, Copenhagen University Hospital, Denmark*

**Background**

Susac syndrome is a recently defined, presumed autoimmune microvasculopathy, which may not be as rare as previously assumed. It is difficult to diagnose, because the symptoms of the classic triad (sensorineural hearing loss, branch retinal arterial occlusion (BRAO) and encephalopathy) seldom appear at the same time and some never develop the full triad. It mostly affects young women and many are therefore misdiagnosed with multiple sclerosis (MS).

**Methods**

We present a 42 year old patient with Susac syndrome, who was misdiagnosed with primary progressive MS (PPMS) for 12 years. At the age of 29, the patient experienced sudden sensorineural hearing impairment. A year later she developed dizziness, diminished balance, severe fatigue, weakness in the fingers, intermittent paresthesia, bladder dysfunction, and was diagnosed with PPMS. MRI showed several white matter lesions, some in the corpus callosum, while the cerebrospinal fluid analysis was normal. 8 years later, her general practitioner incidentally noticed deficits in the patient's peripheral vision, unnoticed by the patient. 4 years hereafter, the patient complained of visual impairment and we found quadrantanopsias. We suspected Susac syndrome and carried out a fluorescein angiogram, which was normal. However, optical coherence tomography (OCT) detected a selective loss of inner retinal layers consistent with BRAO, thus confirming Susac syndrome.

**Discussion**

It is essential that Susac patients are diagnosed correctly, quickly and receive immediate immunosuppressive treatment. BRAO is often mistaken for optic neuritis. Fluorescein angiography can detect BRAO in the acute stage, but not later. OCT may help differentiate between BRAO and optic neuritis, many years after it took place.

**Conclusion**

Susac syndrome is a challenging diagnosis, which is probably underdiagnosed and may be misinterpreted as MS. It is therefore essential to increase physician awareness of this disease entity, especially among those who diagnose MS patients. OCT may help determine the correct diagnosis.