

Susac's syndrome presented with leptomeningeal enhancement

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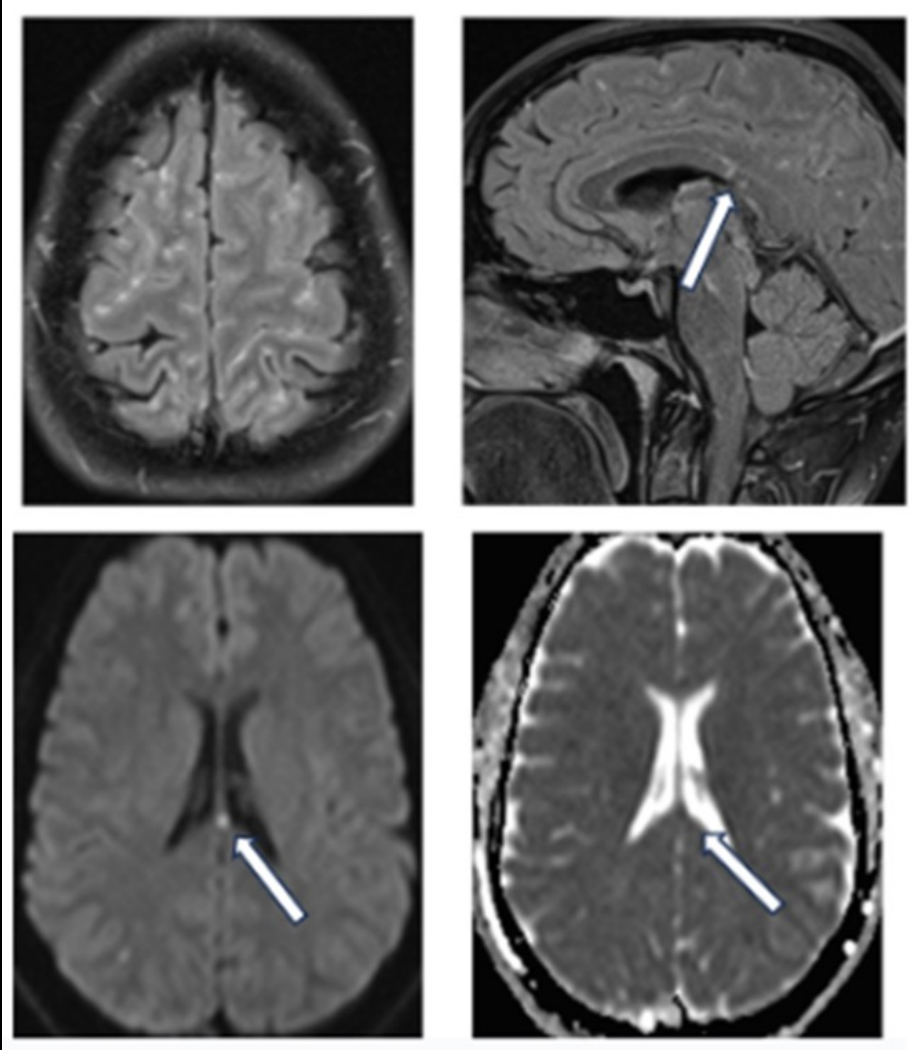


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Clinical Presentation

- A 39 year old female with symptoms including memory concerns, headaches, disequilibrium, floaters and visual disturbance as well as right sided hearing loss.

Imaging Discussion



MRI brain showed scattered focal areas of diffusion restriction involving white matter including corpus callosum. Corpus callosum lesions mainly involve the mid portion of corpus callosum and show diffusion restriction. There is diffuse sulcal enhancement on postcontrast FLAIR images.

Management and Outcome

- Fundus fluorescein angiography showed branch retinal artery occlusion on both sides.
- Audiometric data documented right sided sensorineural hearing loss.
- She was admitted to hospital for high dose IV steroid treatment. She has been clinically and radiographically stable in the setting of weaning Cellcept and on IVIG monotherapy.

Take Home Points

- Susac's syndrome is an immune-mediated endotheliopathy which affects arterioles mainly in the brain, inner ear and retina. Main differential consideration is multiple sclerosis which also most commonly affects young women as Susac's syndrome. "Snowball" lesions mainly involve the midportion of the corpus callosum on MRI as opposed to calloseseptal interface lesions in multiple sclerosis. Leptomeningeal enhancement can be seen in up to half of the cases and better seen on post contrast FLAIR images. Clinical presentation and characteristic imaging findings are helpful to distinguish Susac's syndrome from great mimicker multiple sclerosis.

References

- Roskal-Wałek J, Mackiewicz J, Wypchło Ł, Biskup M, Odrobina D. Susac's syndrome - the crucial role of imaging tests for proper diagnosis. *Ann Agric Environ Med*. 2022 Jun 24;29(2):190-200. doi: 10.26444/aaem/149954. Epub 2022 Jun 2. PMID: 35767750.