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Susac Syndrome - A Case Report

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Abstract

Susac syndrome is a rare autoimmune endotheliopathy characterized by a classical triad of encephalopathy, partial or complete small retinal arteries occlusion, and sensory neural hearing loss. At the on-onset, commonly not all classical triad features are present, and they do not necessarily develop in all patients. Often patients present initially with one of the main features followed by others later in their lives [1]. In many patients, headaches (including migraine-like headaches) precede the development of other symptoms of Susac syndrome. Young women are primarily affected, but it has occurred in individuals ranging from 9 to 72 in age. Although, considered a rare disorder, Susac syndrome is being recognized more often worldwide and may be more common than initially thought. However, it often goes unrecognized or misdiagnosed as 'atypical' Multiple Sclerosis or Acute Disseminated Encephalomyelitis (ADEM).

We present a Susac syndrome case of a 64-year-old woman presenting with headache and visual disturbance.

Introduction

Susac syndrome (SuS) is a rare immune-mediated occlusive microvascular disease, and its main clinical features include acute multiple encephalopathies, Branch Retinal Artery Occlusion (BRAO), and sensorineural hearing loss. Its pathophysiology involves the endothelial cells injury leading to their swelling. These damaged and swollen cells play a vital role in the partial or complete occlusion of the tiny vessels in the brain, retina, and inner ear [2]. This blockage results in decreased blood flow through the vessels - causing either temporary dysfunction or permanent damage to these three organs.

Case description

A 64-year-old female patient initially presented with headache and visual disturbance. A CT scan of the head was done which turned out to be normal. A provisional diagnosis of migraine was made, and an outpatient MRI was advised as a follow-up.

On MRI Head: A 20 X 10 mm lesion of high signal within the right side corpus callosum body was found **(Figure A)**. A little swelling was observed. There was no major associated edema but there was restricted diffusion. A contrast-enhanced study was recommended to rule outacute plaque and neoplasia as they could also have these appearances.

MRI Head with contrast showed that the focus of signal abnormality in the right corpus callosum body was hyperintense on T2-weighted, FLAIR images, and hypointense on T1-weighted images. It was a high signal on the Diffusion-weighted imaging (DWI) **(Figure E)**sequence but not a low signal on the Apparent Diffusion Coefficient (ADC) map, therefore not truly restricting but T2 shine through, hence did not demonstrate appreciable enhancement. The lesion would fit with a typical rounded T2 hyperintense "snow-ball lesion".

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Overall findings were highly suggestive of corpus callosal and white matter involvement with Susac syndrome, given there was a clinical history of the same.

Radiological differential including Multiple Sclerosis (orientation of the corpus callosum lesion not typical) and low-grade glioma (given non-enhancement) were ruled out.

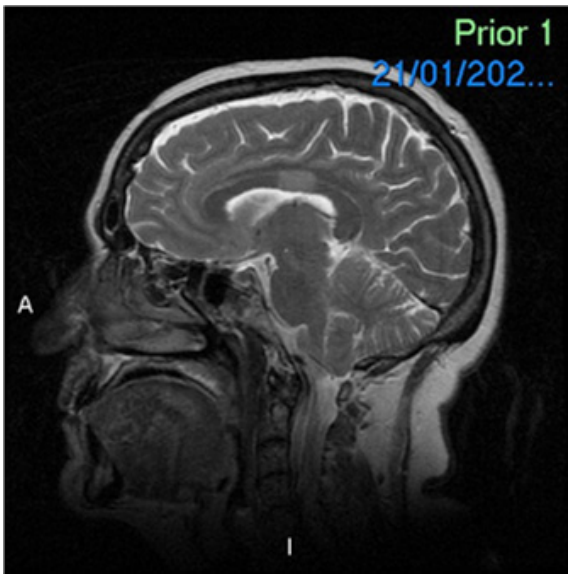

Figure A: T2 weighted (Sagittal section).

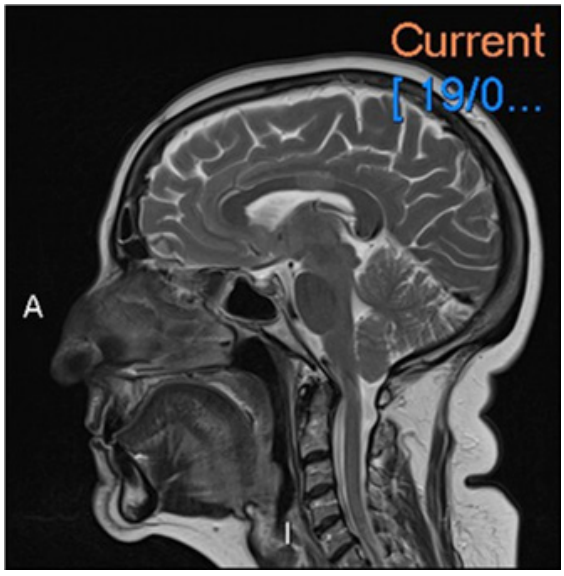

Figure B: T2 weighted (Sagittal section) Follow Up).

Taking into account of the typical findings on the imaging correlating with clinical features, a diagnosis of Susac syndrome was made. It is a rare diagnosis and further management was done accordingly.

On the follow up MRI **(Figure B)** there was a significant decrease in intensity of the signal in T2 weighted images suggesting improvement of the lesion.

Discussion

Susac syndrome is a clinically challenging disease because cases with the typical clinical triad at the initial stage of the disease are rare. It usually has a self-limiting, wavy and monophasic course, lasting from months to years, with varying functional outcomes and residual disabilities. Sometimes it is recurrent

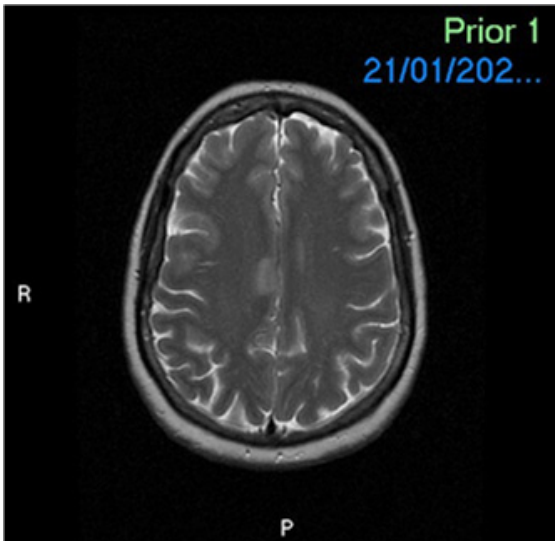

Figure C: T2 weighted (Axial).

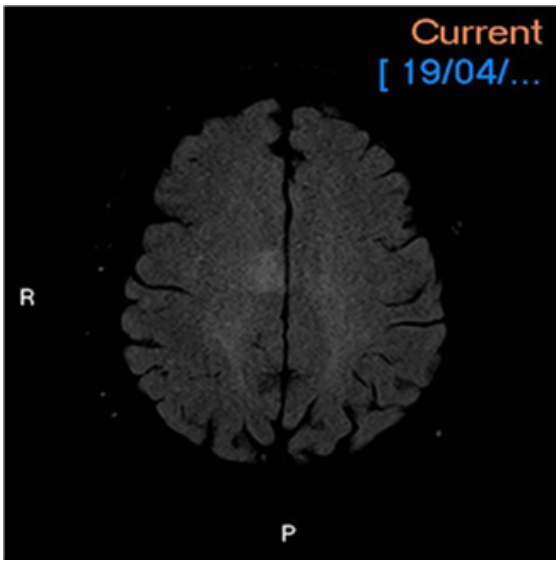

Figure D: Hyperintense on FLAIR. 2 weighted (Axial).

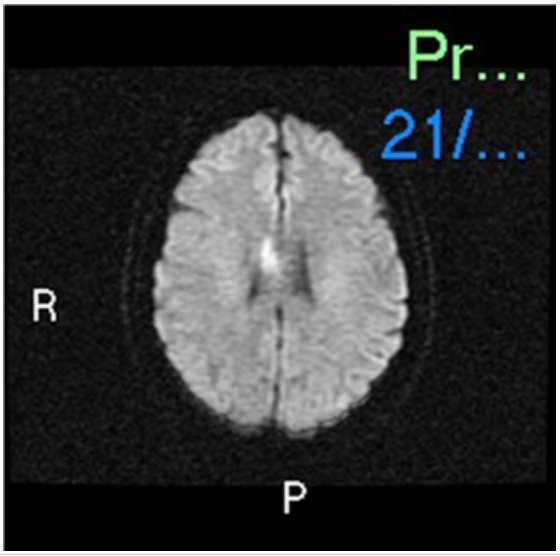

Figure E: High signal on *Diffusion-weighted.*

and even progressive. That's why it is vital to diagnose it early to receive timely treatment for a good prognosis [3]. At present, no standard treatment is there for SuS. Immunosuppression and immunoregulation drugs are being recommended. Steroid drugs, anticoagulant therapy, antiplatelet therapy, and calcium antagonists are also used as adjuvant therapy [4].

The aetiology of the disease is presumed to be autoimmune. It is frequently misdiagnosed as multiple sclerosis, migraine, lupus erythematosus, encephalitis, Ménière disease, thromboembolic stroke, and even schizophrenia. It is needless to say that it is being underreported too as the clinical symptoms mimic the above diseases as well [5]. The objective of this case report is to make doctors aware of the typical findings on the imaging that could help us diagnose this disease at the right time so that the right management can be done, further decreasing morbidity and mortality.

Radiology plays a paramount role in identifying this disease. MRI remains the investigation of choice and is undoubtedly most useful to establish a definite diagnosis and ruling out differentials as well.

Like in our case, NCCT was done first when a patient presented with visual disturbance and headache and the study was found to be normal. Further, MRI could point us towards the diagnosis and was finally established with the help of contrast studies.

References

1. Ke Hu, Yang Yang, Jinzhi Liu, Xiaochong Chen, Chunyu Li, et al. Susac syndrome with the typical clinical triad: A case report and literature review. Journal of Neuroimmunology. 2022; 367: 577822.
2. Adelpoor M, Farahvash MS, Fard MA, Nikdel M, Kiarudi MY. Susac's syndrome in a 27-Year-Old female. Middle East Afr J Ophthalmol. 2011; 18: 320-322
3. Zhao Q, Du XL, Yang Y, Zhou YL, Zhang M. A case report on Susac syndrome. Chin Med J (Engl). 2020; 133: 1754-1756.
4. Grygiel-Górniak B, Puszczewicz M, Czaplicka E. Susac syndrome--clinical insight and strategies of therapy. Eur Rev Med Pharmacol Sci. 2015; 19: 1729-1735.
5. Tai Hung Do, Christiane Fisch, François Evoy. Susac Syndrome: Report of Four Cases and Review of the Literature American Journal of Neuroradiology. 2004; 25: 382-388.