

Pregnancy and Susac Syndrome

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Introduction:

Susac syndrome (SuS) or SICRET (small infarctions of cochlear, retinal and encephalic tissue) has persisted a rare disease. The largest systematic review described only 304 reported cases since the 1970s. Here we find and described 8 cases previously reported in the literature that have started with Susac syndrome during pregnancy.

Methods:

The MEDLINE, EMBASE and SCIELO databases, reference lists of retrieved articles and conference abstracts were searched. For inclusion, every case report or case series citing "Susac Syndrome", "pregnancy" "partum" or "postpartum" were included. Given the limited number of cases, only descriptive analysis was carried out.

Results:

8 cases of SuS during pregnancy were identified. The median age was 26.75 (21-32) years with a median gestational age of 24.8 (1-37) weeks. The debut affected system was auditory in 37.5%, 37.5% neurological and 25% debuted with visual involvement. The time necessary to complete the triad was 5.6 (1-15) months. MRI findings reported callosal involvement in 85% (6/7) with extension to deep gray matter in 71% and posterior fossa involvement in 42% and gadolinium enhancement in 42%. CSL analysis showed 872 mg/dl (77-2000) median protein levels and a median 7.5 (6-9) WBC count. Treatment was initiated with IV steroids in 75% with partial remission of symptoms and oral prednisone or IVIg or PLEX as a second line in 50%. Chronic immunomodulatory treatment includes steroids in 25% (2/8) with partial remission or immunosuppressant oral agents in 62.5% (5/8) with recovery in 40% and partial remission in 60%.

Conclusions

The characteristics of the patients who debuted with SuS in pregnancy do not seem to differ from cases without this association. All the cases found developed the disease during the first or second trimester without any case reported during the postpartum period. The course in general seems to be monocyclic in most patients with a good prognosis.