Pregnancy and Susac Syndrome

Gomez-Figueroa Enrique[1] MD, Casallas-Vanegas Adriana[1] MD, Zabala-Angeles Indhira[1] MD, García-Estrada Christian[1] MD, López Martínez Ramón[1] MD, Neri Daniel[1] MD, Simón Miguel[1] MD, Corona Teresita[1] MD, Flores-Rivera José de Jesús[1]MD, Rivas-Alonso Verónica[1] MD.

[1]Neurology Department, Instituto Nacional de Neurología y Neurocirugía, México City, México.

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.

Objective

To analyze the reported cases of 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

7 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 analyisis 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%.

Conclusion

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.

Cases	1	2	3	4	5	6	7	8
Published by	Gordon1991[8]	McFayden1987[9]	Hua2014[10]	Ionnides2013[11]	Engelholm2013[12]	Antulov2014[13]	Feresiadou2014[14]	Our case
Origin	US	Canada	US	Australia	Germany	Croatia	Sweden	Mexico
Age	28	31	25	28	32	21	35	34
Gestational age at onset (weeks)	28	No specified	14	13	32	35	37	15
Previous medical history	None	None	None	Epilepsy from a perinatal ischaemic event	None	None	Apparent similar clinical picture at 12 yo, treated with steroids	None
System of onset	Eye	Eye	Auditory	Neurologic	Neurologic	Neurologic	Auditory	Auditory
Neurologic symptoms	Unilateral weakness, dysarthria and apathy	Ataxia and dysarthria	Amnestic syndrome, Gait disorder, Bilateral severe weakness	Bilateral severe weakness and dysarthria	Encephalopathic syndrome and Unilateral weakness	Bilateral severe weakness and progressive cognitive affection	None	Cognitive affection (frontal medial syndrome) and bilateral weakness
Ophthalmologic symptoms	Visual field deficit	Visual field deficit	Loss of visual acuity	Loss of visual acuity	Visual field deficit	None	Loss of visual acuity and visual field deficit	Loss of visual acuity
Auditory symptoms	Bilateral neurosensorial hearing loss	Bilateral tinnitus and neurosensorial hearing loss	Tinnutus	Right neurosensorial hearing loss	Neurosensorial hearing loss	Left neurosensorial hearing loss	Tinnitus and left neurosensorial hearing loss	Tinnitus and bilateral neurosensorial hearing loss
Additional or atypical affection	None	None	Cervical cord involvement	None	Livedo racemosa	None	None	None
Time until fully triad (months)	1	2	6	4	1.5	Not completed	Not completed	6
MRI Findings								
Deep grey matter	No	Not done	Yes	Yes	Yes	Yes	No	Yes
White matter	Callosal and periventricular	Not done	Callosal and periventricular lesions	Callosal	Callosal and periventricular lesions	Callosal and periventricular lesions	No	Callosal
Posterior fossa involvement	No	Not done	Yes	No, but also reported meningeal enhancement	Yes	Yes	No	No
Gadolinium enhancement	No reported	Not done	Yes	No reported	Yes	No reported	No	Yes
CSF findings								
Proteins mg/dl	No reported	252	95	2000	1800	1009	No performed	77
Cells (Mono)	No reported	0	6	9	No reported	No reported	No performed	0
Treatment								
Initial treatment	Heparin	None	IVMP x 5	IVMP x 3	IVMP x 5	IGIV x 5	IVMP x 5	IVMP x 5
Response	Partial		Partial	No response	Partial	Complete response	Partial	No response
2nd line treatment	None	None	Oral prednisone	PLEX and IVIg	Oral prednisone	None	None	IgIV
Chronic treatment	Warfarin	Oral prednisone	MMF	MMF	MMF + MTX	AZA	Oral prednisone	CCF
Response	Almost complete recovery	Partial remission	Partial remission	No response	Almost complete recovery	Almost complete recovery	Almost complete recovery	Partial remission
Prognosis								
Follow up (years)	0.2	4	1.5	3.5	1	0.2	3	0.5
Sequels	Visual deficit	Mild hearing loss	Cognitive deficit with visuospatial and word recall	No specified	Mild cognitive deficit	Subtle weakness	Mild left hearing loss	Cognitive deficit
Course of disease		Monocyclic	Monocyclic	Chronic continuous	Probably Monocyclic	Probably Monocyclic	Monocyclic	Probably Monocyclic
Final pregnancy state	Healthy product	Healthy product	healthy product	Therapeutic abortion at 15 weeks GA	Healthy product	Healthy product	Healthy product	Therapeutic abortion at 17 weeks GA