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Central Retinal Artery Occlusion in a Healthy Pregnant Woman: A Suspected Case of Susac Syndrome

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An eighteen-year-old pregnant woman presented with history of acute loss of vision in the right eye for 4 days. Clinical examination revealed central retinal artery occlusion in the right eye. All investigations were normal except MRI brain. MRI report suggested the possibility of acute vasculitis or Susac syndrome; it should be considered in all cases of acute retinal artery occlusions occurring in apparently healthy young females without any precipitating factors. The syndrome may not manifest with full clinical triad.

Retinal artery occlusions (RAO) are uncommon in younger people and most of them will have some identifiable etiological disorders[1]. Carotid artery is a common source of endogenous emboli causing RAO. Exogenous emboli, thrombotic, vasospastic and vasculitic events could cause RAO. Susac syndrome is clinical triad of encephalopathy, retinal artery occlusions and hearing loss[2]. Many patients may not present with full blown clinical features in the beginning. Even though rare, unilateral central retinal artery occlusion has been reported as the sole presenting sign in cases of Susac syndrome[3]. Susac syndrome should be evaluated in all young patients presenting with retinal artery occlusions.

The aim of this report is to present a rare case of suspected Susac syndrome in pregnant young lady.

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THE CASE

An eighteen-year-old Bahraini female 24-weeks pregnant presented with sudden diminution of vision in her right eye for 4 days. She had no problems associated with her pregnancy and no other systemic illness. There was no history of any recent ocular trauma or preceding transient loss of vision. She had history of road traffic accident 2 years ago resulting in mandibular fracture on the left side, which was treated and healed. No history of cosmetic or surgical procedures was revealed. History of transient decrease in hearing was revealed.

Ophthalmologic examination showed a visual acuity of counting fingers close to face in the right eye and 20/20 in the left eye. Anterior segment examination revealed marked afferent papillary defect in the right eye. Fundus examination showed mild optic disc pallor with attenuated vessels and pale retina at the posterior pole with cherry red spot suggesting central retinal artery occlusion. Left eye fundus was normal.

Fundus photography and ocular coherent tomography (OCT) were performed showing early mild macular edema with late follow up OCT images showing atrophic changes at the macula and the posterior pole, see figure 1. Fundus fluorescein angiography (FFA) was not done (contraindicated as advised by the gynecologist). Systemic examination was within normal limits. Gynecological consultation showed normal 24-week gestation. ENT evaluation showed no abnormality with normal audiogram and tympanogram in both ears. Generally, there was no systemic abnormality.

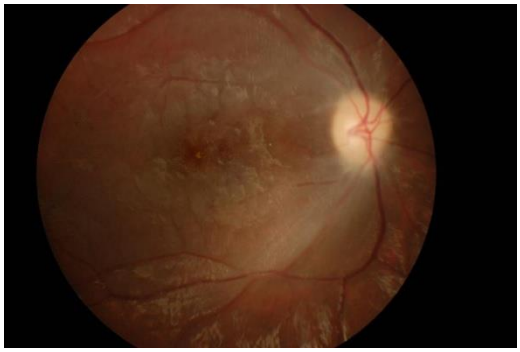

Figure 1: Central Retinal Artery Occlusion of the Right Eye

The following laboratory studies were normal: complete blood count, erythrocyte sedimentation rate, C reactive protein, platelet count, prothrombin time, activated prothrombin/activated partial thromboplastin time, bleeding time, lipid profile, plasma homocysteine, hyper hypercoagulability testing (Fibrinogen, antithrombin III, protein S and C), autoimmune markers antinuclear, anticardiolipin, antineutrophil cytoplasmic antibodies). ESR was 27 mm in the first hour. Fibrinogen was 4.91g/L (Normal 2-4 g/L), which was considered normal during pregnancy.

Carotid Doppler study and Cardiac Echography revealed no abnormality. MRV brain and MRI brain scan were performed. MRV did not show any abnormality. MRI brain scan showed multiple small patches of increased signal intensity in T2W1 and FLAIR images involving the deep periventricular white matter on both sides, more posteriorly and to less extent at both basal ganglia and corpus callosum suggesting encephalopathy, see figure 2 a, b. During the work-up period, the patient developed severe headache and was admitted with a working diagnosis of encephalopathy or vasculitis with demyelinating white matter disease and possibility of Susac syndrome. The patient was treated with intravenous methylprednisolone and subcutaneous enoxaparin. She became symptom free in 4 days except for the decreased vision and was discharged on tapering dose of prednisolone.

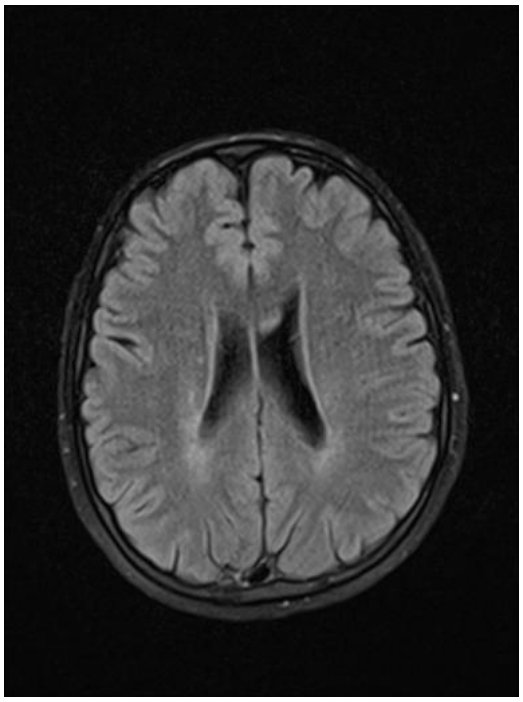

Figure 2a

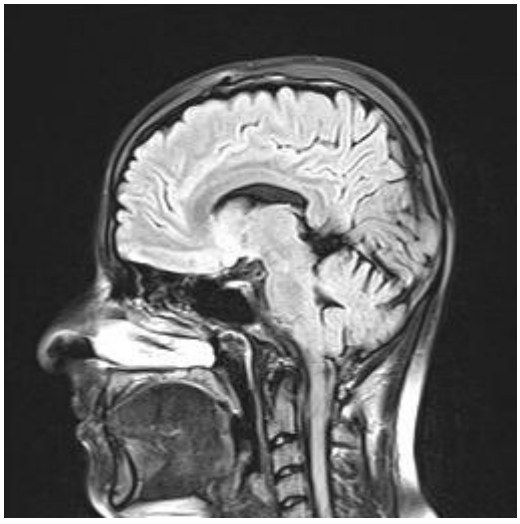

Figure 2b

Figure 2 (a and b): MRI Brain Showing Increased Signal Intensities in Deep Peri-Ventricular White Matter and Corpus Callosum

DISCUSSION

Retinal artery occlusion generally occurs in older males with an average age of 60. Only 10% are below 40 years[4]. Retinal artery occlusion in young people is multifactorial in etiology. The reported factors are hypercoagulable state, such as homocystinemia, hyperlipidemia, coagulation disorders, antiphospholipid antibody syndrome, vasospasm as in migraine, use of oral contraceptive pills, hormonal therapy, etc[6-8]. Cardiac conditions, such as valvular heart disease

and rheumatic heart disease are also reported to cause retinal arterial occlusions[5]. Apart from this, smoking, alcoholism, sickle cell disease and cosmetic filler injections in periocular areas are other risk factors[9-11]. √

Susac syndrome is a rare microangiopathy affecting the arterioles of the brain, retina and cochlea; it is presumed to be of an autoimmune origin. It is characterized by a clinical triad of subacute encephalopathy, visual loss secondary to retinal branch occlusions and sensorineural hearing loss; it is more common in females[12]. These cases might not be initially reported with full blown clinical features. Analyzing the subclinical cases, it was found that 97% of the patients did not have the clinical triad at the time of onset. In some patients, the triad could reveal itself after a delay of weeks and up to two years or more[13].

Some cases of retinal artery occlusions are without any identifiable causes or risk factors[14]. Two of such reports were in healthy pregnant females[15,16]. However, some of these cases were not evaluated by MRI brain scan.

CONCLUSION

To our knowledge, there is no reported case of Susac syndrome from Bahrain. Even though the full clinical triad is missing and considering the percentage of incomplete triad at the time of presentation and negative laboratory reports for other pathologies in this case, Susac syndrome is a very likely diagnosis. The case needs further follow-up over a longer period to confirm the diagnosis. √

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