KJO
pISSN: 1011-8942 eISSN: 2092-9382

Korean J Ophthalmol 2014;28(1):113-114
http://dx.doi.org/10.3341/kjo.2014.28.1.113

Letter to the Editor

Sneddon's Syndrome versus Susac Syndrome

Dear Editor,
I read with interest the article titled "Acute central retinal artery occlusion associated with livedoid vasculopathy: a variant of Sneddon's syndrome [1] and there may be a different diagnosis. Since a few cases of young patients have been identified with Susac syndrome with livedo, could this case represent a case of this disorder? Did the patient have an magnetic resonance imaging scan of the brain and were there any callosal lesions? What did the fluorescein angiogram of the left (the normal) eye show? This was not included in the paper. This author will state that central retinal artery occlusion is very rare in Susac syndrome but not impossible. Has this patient's hearing been checked?

Robert A. Egan
Oregon Neurology, Tualatin, OR, USA

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Author Reply

Central Retinal Artery Occlusion in a Young Female: Sneddon's Syndrome versus Susac Syndrome?

Dear Editor,
We appreciate the comments provided by Dr. Egan on our article entitled "Acute central retinal artery occlusion associated with livedoid vasculopathy: a variant of Sneddon's syndrome" [1]. He indicated that we had not provided adequate information to eliminate the possibility of Susac syndrome. This patient may have demonstrated symptoms similar to those associated with Susac syndrome such as livedo reticularis and retinal artery occlusion in young women without prior medical history. However, we did not consider Susac syndrome as the definite diagnosis for the following reasons: first, the patient had not demonstrated any symptoms or signs of inner ear or brain involvement for 8 years since first developing livedo reticularis. Second, the patient presented with central retinal artery occlusion (CRAO) and not with branch retinal artery occlusion or arterial wall hyperfluorescence, which are typically observed in patients with Susac syndrome [2]. Third, the intimal proliferation observed via skin biopsy is consistent with Sneddon's syndrome rather than with Susac syndrome [3,4]. Patients with Susac syndrome can remain asymptomatic even though subtle abnormalities can be observed on magnetic resonance images or in audiogram findings. Unfortunately, we did not perform either of these tests. Fundus fluorescein angiography of the left eye was completely normal.
Prior to the report demonstrating skin involvement proven by histological examination, skin involvement was not considered to be a symptom of Susac syndrome [4]. However, in the reported case, apparent psychomotor symptoms developed 2 weeks after skin involvement [4]. Another case with Susac syndrome initially presented with unilateral CRAO as a sole symptom; however, the patient later developed apparent hearing loss, encephalopathy, and contralateral branch retinal artery occlusion without skin involvement [5]. In conclusion, although we cannot completely eliminate the slight possibility of Susac syndrome, clinical manifestations were limited to CRAO along with livedo reticularis and intimal proliferation observed on skin biopsy, and these manifestations indicate a diagnosis of Sneddon's syndrome in this young woman with livedoid vasculopathy. We would like to thank Dr. Egan for his comments, which were helpful in the differential diagnosis of Sneddon's syndrome-associated CRAO along with providing additional information regarding Susac syndrome.

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