Negative findings of regional cerebral blood flow with ^{99m}Tc-ethyl cysteinate dimmer in Susac's syndrome

To the Editor: We would like to present a case of Susac's syndrome and explain the role of brain single photon emission tomography (SPET) with acetazolamid enhancement in this rare and important disease.

Susac's syndrome (SS) consists of a noninflammatory microangiopathy which results in a triad of, encephalopathy, branch retinal artery occlusions, and hearing loss[1]. Other neurological manifestations due to multifocal vasculitis are extremely variable and include ataxia, corticospinal tract signs, urinary incontinence and vertigo [2]. The etiology of such an occlusive microangiopathy is unknown and its predilection for the brain, retina, and cochlear has not been adequately elucidated. Brain biopsies have reported sclerosis of the media and adventitia of cortical and pial vessel [3, 4].

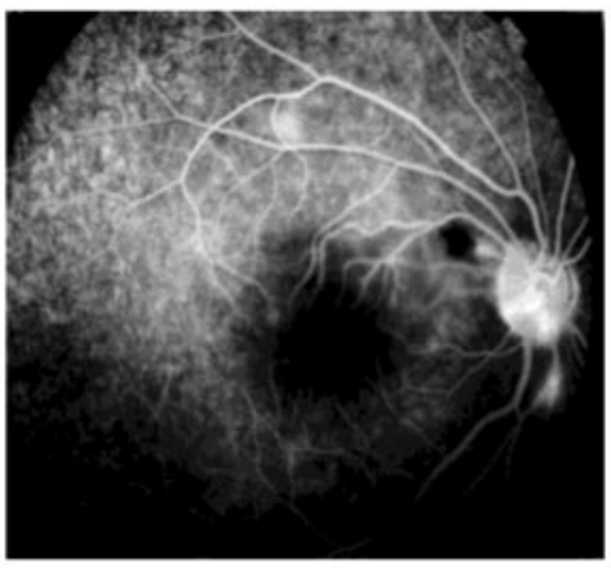

Figure 1. Fluorescein angiogram showing the segmental arteriolar branch occlusion seen in Susac's retinopathy

The differential diagnosis of SS is broad [5] and includes, multiple sclerosis, aseptic meningitis, systemic lupus erythematosus, Bechet's disease, acute and chronic encephalitis, thromboembolic stroke and complicated migraines. Other diseases like sarcoidosis, tuberculosis, syphilis, systemic lupus erythematosus, antiphospholipid antibody syndrome [APS] and lymphomas should be kept in mind [6].

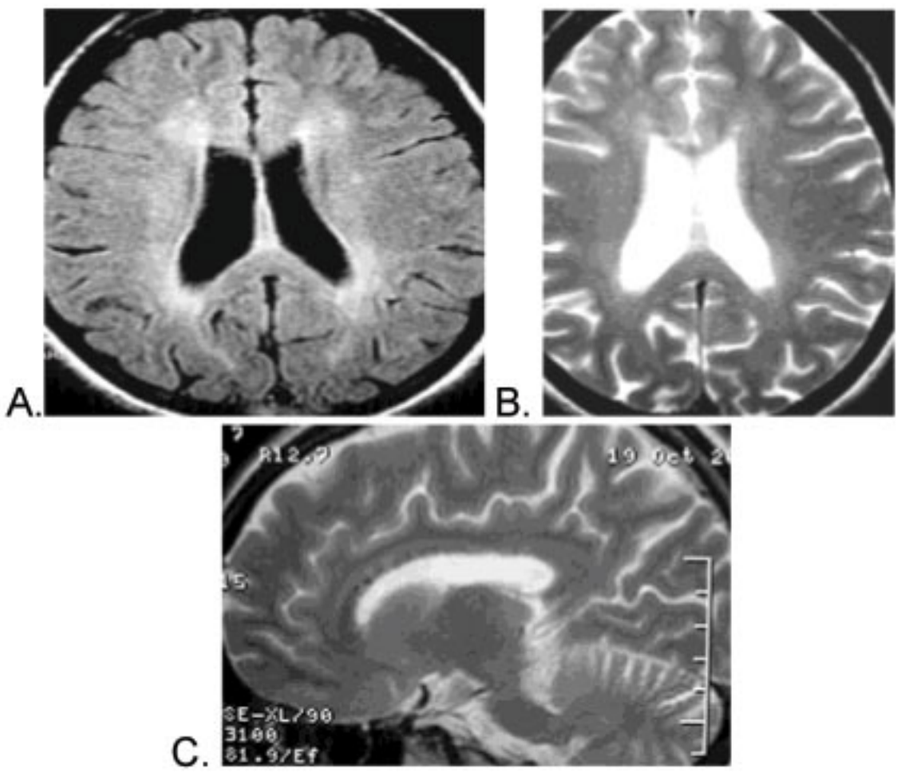

Figure 2. Magnetic resonance imaging of brain showing a few small high signal intensity foci in periventricular white matter on FLAIR (A), T2 weighted images (B) and also in corpus callosum on T2 weighted image (C).These lesions are bright spots on the aforementioned images.

Our case was a 27 years old lady presented with visual impairment of the right eye. The patient had no fever on admission and a general physical condition was normal. Laboratory tests including erythrocyte sedimentation rate, C reactive protein, glycemia, cholesterol, triglyceride levels, liver function tests and urine analysis were within normal limits. Antibody screening tests, i.e. antinuclear antibodies, anti-netrophyl cytoplasmic antibodies, anti-DNA, antismooth muscle antibodies (ASMA) and anti β2 microglubolin, rheumatoid factor and lupus erythematosus cells were negative.

Three months after initiation of the disease, the patient complained of left ear hearing loss and audiometric evaluation demonstrated sensorineural hearing loss and impaired word discrimination. Subsequently, the patient had intermittent sensory and motor signs that resulted in hospital admission where she was treated with corticosteroids.

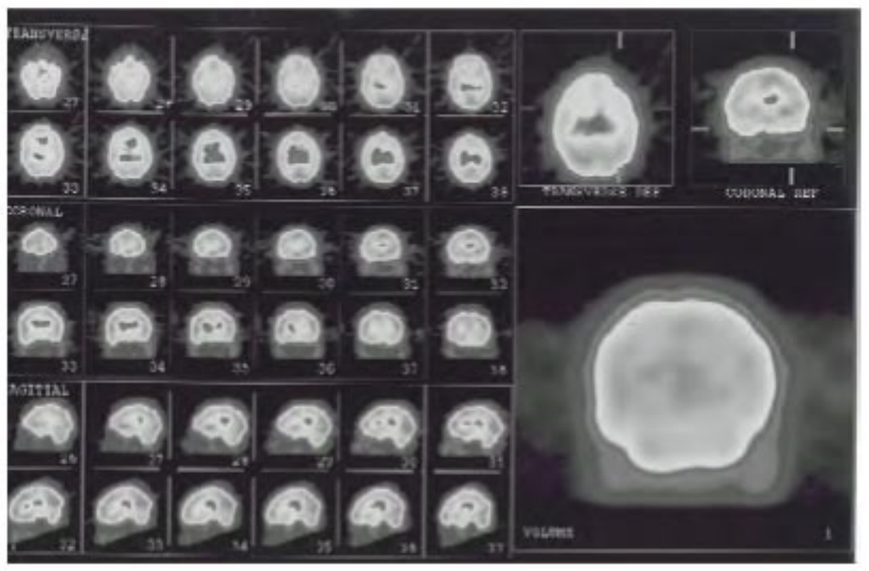

Figure 3. Brain SPET with ^{99m}Tc-ECD shows no remarkable abnormality.

The acquired electroencephalogram showed no abnormality. The ophthalmologic examination revealed mild pigmentation on the right macula. A primary test showed central scotoma on the right macula congruent with multiple retinal arteriolar branch occlusions on fluorescein angiography (Fig. 1). She was subjected to brain MRI twice with an interval of 4 months that showed only a few tiny high signal intensity foci in the periventricular white matter on the FLAIR and T2 images (Fig. 2a, 2b and 2c); These lesions are bright spots on the aforementioned images. Cervical MRI was unremarkable.

The cerebral vascular reactivity assessed with acetazolamide brain perfusion SPET with technetium-99m ethyl cysternate dimer (^{99m}Tc-ECD) was also performed and revealed no perfusion abnormality (Fig. 3). Because the acetazolamide stress brain perfusion SPET was within normal limits, basal brain perfusion SPET was not important.

Susac et al in 2003 reported the largest series of cases of a syndrome that bears Susac's name that is 27 cases studied by magnetic resonance imaging (MRI). They reviewed that the character of lesions on FLAIR and on T2 sequences consisted of small well-demarcated, spherical, high-signal-intensity lesions located in the body and splenium of corpus callosum and further attributed to microinfarctions [7]. The size of the callosal lesions is usually 3-7mm, suggesting occlusion of small (under 100μm), precapillary arterioles [8]. This

small size is beyond the resolution of arteriography, which is almost always normal in patients with SS [7, 9].

Diffusion-weighted imaging and apparent diffusion coefficients have been proved to be sensitive to histologic and physiologic changes associated with brain infarction and useful in differentiating acute from chronic lesions in Susac's syndrome [10, 11]. On the other hand, Xu et al (2004) reported one case of Susac's syndrome in which perfusion MRI, did not reveal any areas of decreased perfusion. The authors suggested that their study was performed months after the initial presentation, when the patient was in remission and presumably, no longer suffering from cerebral ischemia [12].

Others have reported brain SPET hypoperfusion in 100% and 45% of SLE patients with and without abnormal brain MRI, respectively [13]. Others have reported APS cases who showed hypoperfusion lesions on brain SPET with technetium-99m hexamethyl-propylene amine oxime and no obvious abnormalities on MRI [14, 15]. Microscopic brain examination of APS patients shows occlusion of small brain vessels caused by fibrin thrombi or endothelial proliferation in the vessel lumen [16] which explain the hypoperfusion findings of APS patients on brain SPET [14, 15].

As our knowledge, in the first study with fluoro-deoxy-glucose positron emission tomography (FDG-PET) , marked hypometabolism in the right frontal, parietal and temporal lobes was shown[17]. In that case, the hypometabolism on PET was more extensive rather than on conventional MRI, suggesting that positron emission tomography (PET) may be complementary to other diagnostic techniques in the evaluation of SS.

In our case, unlike to a connective tissue disorder with neurologic manifestations and usually an abnormal perfusion scan, brain SPET with ^{99m}Tc-ECD and acetazolamid enhancement were normal. The small volume involvement of brain parenchyma and also the early stage of the disease might explain the absence of such abnormalities in the acetazolamid-challenged SPET. Furthermore, the study was performed after corticosteroid treatment, which may have lead to patrol remission during which the patient was not suffering from cerebral ischemia. *In conclusion,* brain SPET with ^{99m}Tc-ECD-acetazolamid enhancement in this case of SS was normal. The small volume involvement of brain parenchyma, the early stage of the disease and treatment with steroids might explain normal findings.

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