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 acetzolamid 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].

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-nitrophil cytoplasmic antibodies, anti-DNA, antismooth muscle antibodies (ASMA) and anti β2 microglobulin, 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.

The differential diagnosis of SS is broad [5] and includes, multiple sclerosis, aseptic meningitis, systemic lupus erythematosus, Bechet’s disease, acute and chronic encephalitis, thrombocytopenic 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].

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-nitrophil cytoplasmic antibodies, anti-DNA, antismooth muscle antibodies (ASMA) and anti β2 microglobulin, 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.

The differential diagnosis of SS is broad [5] and includes, multiple sclerosis, aseptic meningitis, systemic lupus erythematosus, Bechet’s disease, acute and chronic encephalitis, thrombocytopenic 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].

The differential diagnosis of SS is broad [5] and includes, multiple sclerosis, aseptic meningitis, systemic lupus erythematosus, Bechet’s disease, acute and chronic encephalitis, thrombocytopenic 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].

The differential diagnosis of SS is broad [5] and includes, multiple sclerosis, aseptic meningitis, systemic lupus erythematosus, Bechet’s disease, acute and chronic encephalitis, thrombocytopenic 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].

The differential diagnosis of SS is broad [5] and includes, multiple sclerosis, aseptic meningitis, systemic lupus erythematosus, Bechet’s disease, acute and chronic encephalitis, thrombocytopenic 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].
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
coefficient values 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-
doxy-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
correlation with 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 99mTc-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 99mTc-
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.

Bibliography

outcome in Susac syndrome. Medicine (Baltimore). 2007; 86:
93-102.
2. O’Halloran HS, Pearson PA, Lee WB et al. Microangiopathy
of the brain, retina, and cochlea (Susac syndrome): A report of
five cases and a review of the literature. Ophthalmology. 1998;
105: 1038-44.
imaging demonstrates fiber impairment in Susac syndrome.
disturbance and hearing loss-recognizing the symptoms of
16: 789-90.
7. Susac JO, Murtagh FR, Egan RA et al. MRI findings in
8. Moody DM, Bell MA, Challa VR. Features of the cerebral
vascular pattern that predict vulnerability to perfusion or
oxygenation deficiency: an anatomic study. AJNR Am J
9. Saw VP, Canty PA, Green CM et al. Susac syndrome:
Microangiopathy of the retina, cochlea and brain. Clin
10. Allmendinger AM, Spokker V, Destani S, CT and MRI imaging
of Susac syndrome in a young male presenting with acute
11. White ML, Zhang Y, Smoker WR. Evolution of lesions in
Susac syndrome at serial MRI imaging with diffusion-weighted
imaging and apparent diffusion coefficient values. AJNR Am J
12. Xu MS, Tan CB, Umapathi T, Lim CC. Susac syndrome:
13. Chen JJ, Yen RF, Kao A et al. Abnormal Regional cerebral
blood flow found by technetium-99m etyl cistate dimer brain
single photon emission computed tomography in systemic lupus
erythematosus patients with normal brain MRI findings. Clin
cerebral blood flow with 99mTc-HMPAO in primary
antiphospholipid antibody syndrome. J Nucl Med. 1999; 40:
1446-50.
15. Kato T, Morita A, Matsunoto Y. Hypoperfusion of brain single
photon emission computerized tomography in patients with
16. Hughson MD, McCarthy GA, Sholler CM, Brunnback RA.
Thrombotic cerebral arteriopathy in patients with the
report and PET imaging findings. Acta Neurol Belg. 2009;

Majid Assadi, MD, Hooman Salimipour, MD, Iraj
Nabioupour, MD, Reza Nemati, MD, Jamshid
Saberifarid, MD, Mohammad Seyedabadi, PhD

1. Bushehr Research Center for Nuclear Medicine, The
Persian Gulf Biomedical Sciences Institute, Bushehr
University of Medical Sciences, Bushehr, Iran
2. Department of Neurology and 3. Radiology, Faculty of
Medicine, Bushehr University of Medical Sciences,
Bushehr, Iran

Majid Assadi, MD,
Bushehr Research Center for Nuclear Medicine, The
Persian Gulf Biomedical Sciences Institute, Boostan 19 Alley, Imam Khomeini Street,
Bushehr, Iran
Tel: 0098-771-2580169
Fax: 0098-771-2541828
E-mail: assadipo@yahoo.com

Hellenic Journal of Nuclear Medicine • September - December 2010 292