A Patient With Susac Syndrome and Bipolar Disorder

To the Editor: Susac syndrome is a rare disease first described in 1979 by Susac et al.[1] To date, more than 200 cases have been published worldwide.[2] The typical clinical manifestation is characterized by the triad encephalopathy, branch retinal artery occlusion, and sensorineural hearing loss, which can occur simultaneously or subsequently in unpredictable order.[3] Susac syndrome mainly affects women (3:1) between the ages of 20 and 40 years, but the age range extends from 7 to 72 years.[4] Susac syndrome is considered to be a vasculopathy, and recently antiendothelial antibodies were reported.[5] Yet, the precise etiology remains unclear. During the encephalopathic phase of the disease, many patients suffer from temporary psychiatric symptoms.[6,7] Here, we report the case of a Swiss patient who developed a comorbid psychiatric disorder, possibly secondary to Susac syndrome. To our knowledge, this is the first case to be reported in the literature describing such a medical condition.

Case report. Ms A, a 52-year-old white Swiss woman, was admitted to our department of general psychiatry in 2004, 2005, and 2010 (for roughly 1 month each time) under conditions of involuntary commitment presenting with mood disturbance, mania, and delusions. She reported feelings of persecution and delusions of grandeur and showed aggressive behavior (setting fires, physical violence). Aside from a neurologic report in 1994, when a certain affective lability was noticed, her psychiatric history was otherwise unremarkable. The family history revealed an affective disorder, ie, major depression, in her sister and a propensity to cardiovascular disease. Her father and brother died of myocardial infarction at the ages of 39 and 38 years, respectively.

Her medical history showed acute bilateral hearing loss between the ages of 20 and 30 years, intermittent visual disturbances, and recurrent facial anesthesia. Cerebrospinal fluid samples exhibited elevated protein, and antinuclear antibodies were positive in serologic samples. Neurocranial magnetic resonance imaging studies in 1997, 2004, and 2009 displayed progressive cortical atrophy, markedly in the temporal and insular region, as well as white matter degeneration (corpus callosum), whereas hyperintense lesions on T2-weighted images were scarce. Neuropsychological testing repeatedly detected attentional deficits and impairment of verbal memory. Fluorescein angiography revealed retinal vasculitis, and electroencephalogram changes were ascertained. On the basis of these results, the diagnosis of Susac syndrome was retrospectively established by the department of neurology. From a psychiatric perspective, the patient's symptoms were indicative of a bipolar spectrum disorder, and, according to *ICD-10*[8] and *DSM-IV*,[9] the occurrence of psychotic symptoms justified the differential diagnosis of schizoaffective disorder because the delusions partly outlasted the manic symptoms. During the last admission in 2010, we treated the patient with risperidone 6 mg/d and quetiapine 100 mg, which led to a steady regression of psychotic symptoms. Despite the involuntary inpatient settings, she developed compliance and cooperation. However, she remained in denial about her psychiatric diagnosis.

We can only speculate whether the development of the bipolar disorder is related to Susac syndrome, but the timeline indicates that psychiatric symptoms of clinical relevance emerged 25 years after the first signs attributable to Susac syndrome. When we focus on gross, pathological neuroanatomy as provided by magnetic resonance imaging, we find gray and white matter abnormalities. A literature search revealed that these abnormalities are relevant to the pathophysiology of bipolar disorder,[10,11] and our attention is particularly drawn to the corpus callosum, the structure of which is regularly altered in Susac syndrome. Assuming that white matter abnormalities in the corpus callosum are at least partly involved in the pathogenesis of bipolar disorder, the patient's psychiatric disorder is possibly secondary to Susac syndrome. There is limited evidence on schizoaffective disorder and its possible neurobiological underpinnings. Regarding discussions about the unknown future of the category schizoaffective disorder in *ICD-11* and *DSM-5* and propositions to apply a dimensional approach to psychopathology,[12] we consider our differential diagnosis to be of less clinical importance, for it would not have prompted us to a different treatment. This case illustrates the necessity for an interdisciplinary approach to Susac syndrome and, vice versa, a thorough workup of patients suffering from bipolar disorder.

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Potential conflicts of interest: None reported.
Funding/support: None reported.
Published online: August 9, 2012.
Prim Care Companion CNS Disord 2012;14(4):doi:10.4088/PCC.12l01350