ness, the patient started exhibiting disinhibited behaviors such as disrobing in front of others, manipulating her genitals, and making sexual advances toward her father. She would lick any object lying on the ground and whenever she got an opportunity, she would rush to the toilet and try to put urine and feces in to her mouth (urophagia and coprophagia, respectively). She also exhibited hyperphagia and would eat three times the usual amount of meal at a time. She was impulsive and would try to snatch any object and slap others without reasons. She would misidentify parents and relatives and would call them by different names. A meaningful mental state examination was not possible in view of her irrelevant speech and impulsive and disorganized behavior. Her neurologic examination did not show any long tract signs. Her blood work up was unremarkable and so was an EEG. However, a head CT scan showed diffuse mild cerebral atrophy with no focal abnormalities.

The patient was started on carbamazepine titrated up to 600 mg per day without any improvement in symptoms. Three weeks later, risperidone 4 mg per day was added, but only mild improvement was noticed. Six weeks after this combination, sertraline 25 mg was added which was increased to 100 mg over a period of 2 weeks. Surprisingly, 6 weeks after commencing sertraline, all her symptoms improved remarkably, including hypersexuality, disinhibited behavior, hyperorality, urophagia and coprophagia. On follow-up 2 months later, she had stopped taking sertraline for 2 weeks (while continuing other drugs) and had worsening of the above-mentioned symptoms; reinstituting sertraline again ameliorated the symptoms. The patient was doing well for next 6 months till her last follow-up.

Our case exhibited most of the symptoms of KBS. Although KBS has been shown to be associated with demonstrable bilateral lesions in the anterior temporal horn or amygdale [4], other authors have reported that lesions in the amygdala are not necessary for KBS in animals or humans [1]. This is consistent with our patient who showed diffuse brain atrophy but no localized lesions. Carbamazepine and antipsychotics have been previously reported to be effective in managing some of the behavioral symptoms of KBS [2]. There is only one report describing the successful use of selective serotonin reuptake inhibitors (SSRIs) in two patients with KBS secondary to head trauma [5]. In one of these cases, the patient didn't respond to carbamazepine alone but responded only after being started on fluoxetine. In the second case, the patient responded to a combination of antipsychotics and sertraline. That sertraline may have an independent effect on symptoms of KBS is further bolstered by relapse of symptoms on withdrawing this drug and their resolution again on rechallenge in our patient. Some of the symptoms of KBS are compulsive in quality and may reflect a disturbance of aggression regulation and impulse control. It may be worth exploring if the known efficacy of SSRIs against these symptoms cluster may play a role in treating KBS.

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Brief psychotic disorder associated with Sturge–Weber syndrome

Keywords: Sturge–Weber syndrome; Phakomatoses; Brief psychotic disorder

Sturge–Weber syndrome (SWS) is a rare, sporadic neurocutaneous syndrome with an estimated frequency of one per 50,000 live births [5]. Although mental retardation is a well-studied sequelae of SWS [5], little attention has drawn on the psychological well being of SWS patients. SWS is devastating, especially when children experience recurrent seizures, pervasive learning and behavioral problems, and disabling visual impairment [5], all predisposing to problems in behavioral functioning [1].

Despite the above, literature on psychotic symptoms in these patients is scarce.

We report on the case of a 33-year-old male with an extended nevus covering his whole face except for a small area surrounding his right eye and vascular deformities extending to the left temporo-parietal lobes. He was suffering from glaucoma and was on carbamazepine for complicated seizures. The patient had no personal or family history of major psychiatric disorder and had a normal psychosocial development. He was working as a freelancing software engineer but

had little social life. He presented with psychomotor agitation and persecutory delusion, which had persisted for about 10 days. He was diagnosed as brief psychotic disorder and was prescribed penfluridol 20 mg/week, as he refused daily schemes. Despite the resolution of symptoms, the patient did not return to the prior level of functioning. After 3 months, and despite pharmacotherapy, the patient relapsed again, only to improve again with an increase in penfluridol dosage. Fortunately, the antipsychotic caused no worsening of the seizures.

The young people with SWS who appear to be at most risk for a broad range of behavioral and emotional problems are those with lower levels of intellectual functioning, seizure disorders and more frequent seizures [1]. As the area of the face covered with a port-wine stain increased, so did parent and teacher reports of emotional distress and teacher reports of social problems but only for children who were at least 10 years old [1].

Sing Lee reported [2] on three SWS patients. The first one was first diagnosed with paranoid disorder, a diagnosis that changed to obsessive–compulsive disorder 2 years later [3]. The second one was mentally retarded with a commorbid paranoid disorder and the third patient, also mentally retarded, suffered from a mood disorder [2]. Of these patients, the last two had left hemisphere lesions (same lateralization of lesion as our patient). The small number of reported cases prohibits safe conclusions on the existence of any relationship between lateralization of the lesions and the emergence of psyhcopathology.

The high incidence of schizophrenia permits the possibility of an accidental comorbidity of SWS and schizophrenic disorder. Nevertheless, we feel that psychosis is underreported in this group of patients. This could be due to the atypical presentation of psychiatric symptoms in patients with mental retardation, who represent more than 50% of SWS patients. [4] The clinician must keep in mind that, in the mentally retarded, the line between behavioral and psychotic symptoms is rather thin, depending heavily on the ability of the patient to express oneself fluently.

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