Unusual association of diseases/symptoms

Sturge–Weber syndrome: neurology-psychiatry interface

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Correspondence to Professor Amin Gadit, amin.muhammad@med.mun.ca**Summary**

This is a case of a 22-year-old male who was born with a port-wine stain on right side of his face, developed seizures at the age of 2, was not able to complete formal education in a school. MRI revealed intracranial calcification and left-sided brain atrophy. He had diagnosis of Sturge–Weber syndrome. Since the time of adolescence, he developed psychiatric problems and hence was treated with psychotropic medications. This case remains under the domain of psychiatry as well as neurology and calls for joint management. A number of presentations with Sturge–Weber syndrome have been reported in the literature that poses problems in terms of appropriate medical management. A classical syndrome like this invites serious attention of both psychiatrists and neurologists to devise appropriate drug treatment with a view of interactions, side effects, complications and prognosis.

BACKGROUND

There has always been a debate about some cases that fit well into either psychiatry or neurology. Sturge–Weber syndrome comes under the umbrella of neurology but its psychiatric manifestations are dealt with by psychiatrists. Trainees in psychiatry tend to ignore such cases or do not consider it important for psychiatrists to deal with it. Once in the membership examination of the Royal College of Psychiatrists, a patient with seizure disorder, mental retardation, port-wine stain and behavioural disorder was kept as a long case. The candidate appearing in the examination found it most difficult to handle in terms of history, management and prognosis. It transpired that for psychiatrists, it was important to have awareness about neurology related to psychiatry. Such cases come under the treatment of either specialty. Sturge–Weber syndrome is more like a spot diagnosis with typical clinical features. We thought a case like this would academically be stimulating and will lead future psychiatrists and neurologists to be familiar with many such cases that come under the umbrella of both specialties.

CASE PRESENTATION

This 22-year-old patient was born as a result of complicated pregnancy. His milestones were grossly delayed. He had a port-wine stain on right side of his face since birth. At the age of 2, he developed seizures that were controlled with medications. He was unable to attend regular school and was assessed to be functioning at IQ level of 70. He was assessed first time as an adolescent aged 17 with behavioural problems, withdrawal and attempts at self harm. Since then he was referred simultaneously to the neurologist and ophthalmologist for recurrence of seizures and vision problems. The patient was commenced on citalopram 20 mg once daily and clonazepam 0.5 mg twice daily. His medical history indicates that he suffers from glaucoma, hypertension and hypothyroidism. He was never treated for any psychiatric illness in the past. In

his family, both his parents are alive and healthy; he has two brothers and two sisters. One of his paternal uncles suffers from epilepsy. His maternal aunt is being treated for major depressive disorder. He spends most of his time at home, never employed, does not socialise much and does not abuse any drugs. His full mental status examination remained inconclusive. However, there was objective evidence for depression and anxiety. There was a major problem with communication in terms of comprehension and verbalisation.

INVESTIGATIONS

- ▶ MRI: evidence of intracranial calcification and left sided brain atrophy
- ▶ EEG: epileptogenic activity in right temporo-parietal region
- ▶ Ophthalmology: Evidence of high intraocular pressure
- ▶ High TSH levels
- ▶ IQ test: 70.

DIFFERENTIAL DIAGNOSIS

With classic features of Sturge–Weber syndrome, there was no provision for any differential diagnosis.

TREATMENT

- ▶ Citalopram 20 mg once daily
- ▶ Clonazepam 0.5 mg twice daily
- ▶ Thyroxine 50 µg per day
- ▶ Lamotrigine 200 mg twice daily.

OUTCOME AND FOLLOW-UP

The patient is jointly being followed by neurologist and the psychiatrist. His depressive and anxiety symptoms have shown some improvement. However, he remains a challenge in terms of clinical management. His progress is being monitored with outpatient psychiatric appointment once in a month for the next 3 months.

DISCUSSION

Sturge–Weber syndrome is also known as encephalotrigeminal angiomatosis which is a rare congenital neurological and skin disorder. It is often associated with port-wine stains of the face, glaucoma, seizures and ipsilateral leptomeningeal angioma. Mental retardation is a consequent feature in view of gross brain involvement. Psychiatric problems are not uncommon among such patients. A study showed mood disorder (31%), disruptive behaviour disorder (25%) and adjustment disorder (25%) among a small group of outpatients aged 3–34 years.¹ A 23-month-old girl suffering from Sturge–Weber syndrome without facial angioma was presented with severe tantrums. She went into remission after undergoing brain surgery and being commenced on anticonvulsants.² A variant of classical presentation was seen in an 11-year-old male with a serious bilateral glaucoma associated with a bilateral facial nevus flammeus and cerebral calcifications without neurological symptoms.³

In a case report, an older patient suffering from the same disease presented with episodic ‘angry’ slapping symptoms to a psychiatric facility that were attributed to complex partial seizures that improved in frequency with an increase in the dose of the antiepileptic medication.⁴

There are multiple issues that may pose challenge in managing such cases. With seizures, mental handicap, no structured daily activity and limited understanding, there is every likelihood of mental health problems. Expression of depressive features in such patients may be quite misleading. Often the presentation is in the form of disturbed or aggressive behaviour. The effect of antidepressant on the seizure threshold and drug-interaction with antiepileptic drugs may compound the problem. The increase in frequency of seizures because of depression or antidepressant

medication, the issue of brain damage, uncontrollable seizures because of treatment failure and non-compliance with medications and psychiatric problems can lead to a management dilemma and calls for comprehensive care planning.

The presentation of this syndrome may vary in terms of clinical problems but the issue of appropriate treatment remains a challenge for the concerned physicians.

Learning points

- ▶ Some clinical presentations are common to both psychiatry and neurology.
- ▶ The psychiatrists and neurologists should be well-versed with cases common to both specialties.
- ▶ There are psychiatric manifestations of neurological disorders and vice versa.
- ▶ The psychiatric treatment of neurological disorders is complex.

Competing interests None.

Patient consent Obtained.

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