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A deep insight on psychological aspect in patients with Sturge-Weber syndrome

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Abstract:

Studies on Sturge-Weber syndrome (SWS) have already focused on various medical aspects of this syndrome. This review is focusing on the psychological aspects of Sturge-Weber syndrome patients, the behaviors of patients, and the effects of the disease on the children. This disorder is characterized by angiomas, glaucoma, and seizures with a birthmark. Patients with this syndrome have behavioral problems, a lack of social skills, and a lack of intellectual or academic skills. These patients are usually introverted because of a lack of warmth and limited communication. Friends or classmates tease them by calling them "ugly." All this leads to a lack of social skills, and they mostly keep their feelings to themselves and prefer to be alone. Because of all this mocking by classmates, they keep isolating themselves. All this causes psychological problems in children. They start behaving differently, have suicidal tendencies, and fight with their friends who bully them. Psychological problems are mostly seen in children who are lacking in intelligence and functioning and who have seizure disorders, mostly in those who have frequent seizures. Problems like anxiety, depression, mood changes, anger, and violence are also observed in these patients. All this is always ignored by the parents as well as the doctors. This review aims to focus on the psychological aspect of the patients with SWS, to inform the parents about the behavior of their children with SWS, and to pay more attention to the psychological problems. This review is focusing on the importance of the psychological behavior of patients with SWS, how to diagnose it at an early stage, and how to treat and take care of children with SWS.

Keywords:

Anxiety, depression, psychological, suicidal tendencies

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Introduction

Schirmer was the one who provided detailed descriptions of SWS in 1860.[1] William Allen Sturge first identified this syndrome in a girl who was 6 years old, and he further described SWS-related other manifestations, which are neurological, dermatological, and ophthalmic.[1] It is also called encephalo trigeminal angiomatosis. It is a congenital neuro-dermatological disorder. The cephalic venous microvasculature is impacted by the uncommon, sporadic neurocutaneous condition known as Sturge-Weber syndrome (SWS).[2] It occurs with a frequency of 1 per 50,000. In SWS, blood vessels grow too much, forming growths that are referred to as angiomas. It is characterized by an intracranial vascular anomaly, leptomeningeal angiomatosis, involving the occipital and parietal lobes. Leptomeningeal angiomatosis causes chronic ischemia, which can cause cognitive alterations, visual field cuts, epilepsy, headaches, and intellectual disabilities.[3] Three major features of this are a red or pink birthmark, which is known as a port-wine birthmark; a brain abnormality known as leptomeningeal angiomatosis; and increased eye pressure (glaucoma). The common symptoms and signs are seizures, facial cutaneous vascular malformations, and

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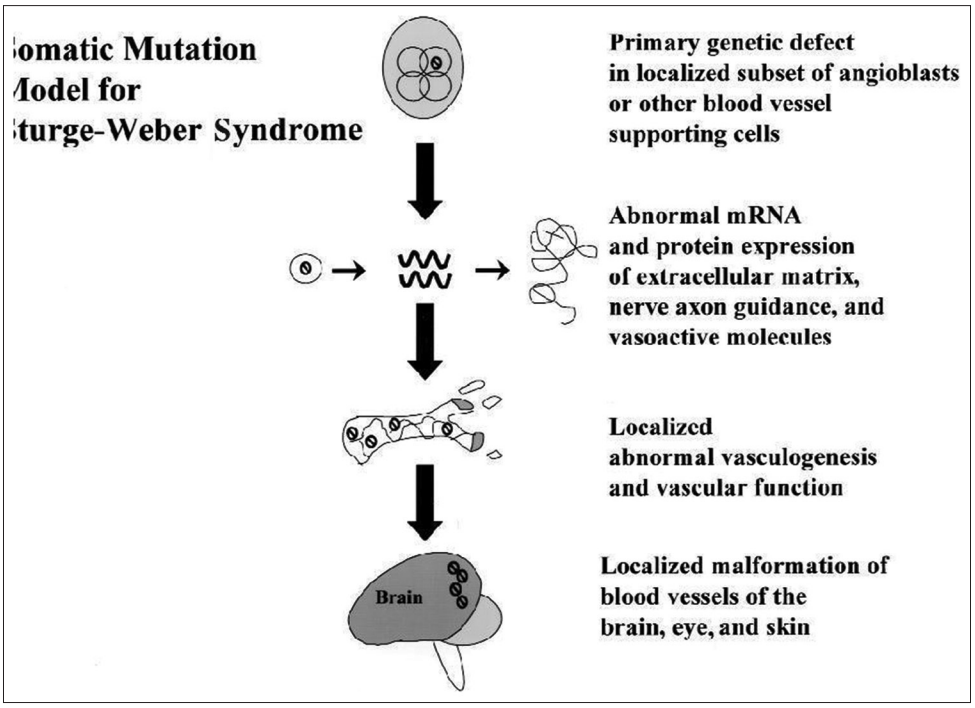

Figure 1: Pathogenesis of Sturge Weber syndrome[8]

glaucoma. Leptomeningeal angiomatosis is caused by ischemia brought on by stasis, which culminates in calcification and laminar cortical effects.[2] The syndrome has a range that includes the birthmark linked to both the brain and skin, which occurs in around 10% of cases.[4] It is caused by a mutation of the GNAQ gene. This type of gene mutation occurs randomly during the development of an embryo, affecting only certain tissues in the body. It affects the small blood vessels called capillaries. Problems with capillaries cause port wine stains or marks to form. It has broadly three types: type 1 dermatoneurological, type 2 dermato-ocular, and type 3 pure neurological. Patients may also face symptoms like seizures, developmental delays, headaches, migraines, hypothyroidism, soft tissue hypertrophy, etc., Some patients with SWS are mentally challenged as well. When we see the psychosocial aspect of SWS, some patients with mild impairments sometimes adjust to normal living, but patients with more devastating symptoms are less confident, irritable, introverted, deficient in social skills, etc.[5] In the study conducted by Lynn Chapieski, we collected psychological and medical data from 79 children and adolescents with SWS and some of their siblings. It is seen that the patients have more problems than their siblings in the behavioral domain, which includes social skills, mood, academics, and functioning. The parents and teachers in this study reported that their children have developmental and other problems. They also reported lower levels of intellectual functioning. In some cases, the parents reported complaints of anger and aggressiveness in schools when they saw other normal children, usually when other kids bully them on their appearance and start behaving aggressively with their classmates.[1] Similarly, the siblings of these patients feel ashamed about their appearance in front of their friends. In some cases, more problems are faced by them during their marriage, too. Another study by Elizabeth Turin has stated that there is a significant association between disruptive behavior disorder and SWS patients. This study presents behavioral, emotional, and psychiatric features and their associations with neurologic symptoms. This study presented behavioral and neurological features in 16 patients, of whom 75% had moderate to severe neurologic symptoms and 95% had psychiatric disorders, and the reason is mainly due to their appearance. All these problems lead to suicidal feelings and behavioral changes in them. Psychological problems can be dangerous if not taken care of.

Some children have behavioral problems. Half of the patients at SWS are mentally challenged. It is seen that most of the patients with SWS behave differently; they have problems with understanding, are slow learners, lack social skills, have anxiety, depression, a lack of intellectual ability, aggressive mood swings, violence, and anger issues.[1] All these psychological problems are because of their illness, which goes unnoticed by their parents and carers. Due to all these symptoms, some children also face psychosocial problems. Children become introverts; their classmates may taunt them about their appearance and social disabilities.[4] These kinds of kids also need more attention, and a lack of attention can also cause behavioral problems. Also, the parents reported

depressed moods and emotional distress. All this may lead to depression and suicidal tendencies, leading to psychological problems. They may want to fight with their own friends who bully them, behave differently, isolate themselves, and stop talking to family and friends. Suicide risk in patients with unilateral right brain involvement has been noticed.[6] To avoid this, proper communication is also important between the patients, their parents, and the doctors.[7] It helps in developing good relationships and satisfaction in patients as a sense of humanity.[7] All these things and behaviors of patients with SWS are most of the time ignored by their parents and families. This review focuses on the psychology of these patients to educate their parents about their behavior and how to treat them in the early stages.

Pathogenesis

Unlike most phakomatoses, SWS is sporadic; it has no definite, identifiable hereditary component. A gene mutation is seen as a nucleotide transition in GNAQ on chromosome 9q21. GNAQ codes for a guanine nucleotide protein, and it is a part of the trimeric G protein complex, which couples with G protein-coupled receptors like the endothelial receptor, the angiotensin II receptor type, vasopressin type 1A and type 1B receptors, and others. Mutations in this GNAQ gene can cause Sturge-Weber syndrome [Figure 1].[8]

Complications

Neurologic: Children suffering from SWS have neurologic anomalies like epilepsy, mental retardation, attention deficit hyperactivity disorder, and migraine.[3] Epilepsy occurs in 75–90% of patients with SWS. Focal seizures are also seen in patients with SWS. Fever and infection are seen as the aggravating factors for the onset of seizure. Seizures like generalized tonic-clonic seizures, myoclonic seizures, and absence seizures are seen in SWS. Microcirculatory stress and hypoxia are more likely to cause seizures. Half of the SWS patients are mentally retarded, whereas others have learning disabilities, behavioral problems, and attention disorders.[9] Seizures begin in infancy with other focal deficits. Most of the patients have had seizures since the age of 2. 30–45% of patients with SWS have headaches. The temporal relationship between headaches, migraines, and seizures is relatable to the pathogenesis of SWS.[10] Developmental delays and mentally challenged are seen in 50% of patients with SWS. Attention deficit hyperactivity disorder (ADHD) is a severe condition in SWS. ADHD is a chronic condition that has symptoms of attention difficulty, hyperactivity, and impulsiveness. It is mostly seen in childhood and persists into adulthood as well. It results in low self-esteem, difficulty in school, and other problems.

Headache is the second most common symptom of SWS, seen most commonly in children than pathogens.[11]

Ocular: Vascular anomalies of the episclera, conjunctivitis, and retina arise as ocular complications. The most common complication is glaucoma, which occurs in 30–70% of patients.[12] It is also a chronic condition that is caused by damage to the optic nerve, which can lead to loss of vision. 60% of glaucoma occurs in infancy because the eye is susceptible to increased intraocular pressure; 40% occurs in childhood and early adulthood. Enlarged corneal diameters and myopia in infants are the early onset of glaucoma, and in the late onset, there is no eye enlargement.[13,14] Choroidal hemangiomas may be present in 71% of SWS patients. Iris heterochromia, retinal detachment, strabismus, and homozygous hemianopia are also ocular complications of SWS.

Oral: Unilateral hemangiomatous lesions are seen in the maxillary and mandibular regions, gingivitis, lips, palatine region, tongue, etc., Unilateral hyperplasia is present on the gingiva due to an increased vascular component and demonstrates bleeding due to minimal trauma.[15] Gingival overgrowth is seen and is most common in the ipsilateral maxilla and mandible.[15] Port wine stains and gingival enlargement are also seen. Some patients also give the history of drug-induced gingival enlargement and swollen gums.[16]

Minor traumas can also result in more bleeding of the gums. Oral symptoms usually occur on one side of the mouth. There is often a difference observed between the "normal" and the affected side in the mouth as to when the teeth come out and also their development. The teeth can come out too late or too early. Some patients are without birth defects, have spaces between teeth, and also be misaligned. The malformation of jaw is the reason for the misalignment of the teeth and the asymmetrical growth of the jaw. Position of the teeth can also be influenced by the pressure caused by the excess growth of blood vessels. This abnormal growth can put pressure on teeth and cause them to be pushed out of position. It is helpful to have the orthodontist inspect the milk teeth when SWS is indicated. Having SWS is not a contraindication for orthodontic treatment, and it is advisable to check for proper oral hygiene. The presence of braces makes cleaning the teeth more difficult, and as mentioned earlier, oral hygiene is more difficult because of the presence of hypertrophy of the gums. In older patients, orthodontics is sometimes combined with oral surgery. Possible problems can then be detected earlier and be simply treated, and prevented, in a timely fashion. Proper dental hygiene with assistance by a dentist or dental hygienist is extremely important. This syndrome can be treated with anti-epileptics although these medicines have secondary effects and excessive growth of gums.

Orofacial: An ipsilateral angiomatous lesion of the face is sometimes accompanied by angiomatous lesions of the

skin, jaw, and oral cavity. Angiomatosis and skin changes follow the maxillary and trigeminal nerve distributions. A bluish-red discoloration (port wine) appearance or birthmark is seen extra orally on the forehead and eye region.

Psychological: Psychological problems are always seen in patients with SWS, which are mostly ignored by their parents and families. Problems like anxiety, depression, mood changes, anger issues, violence, emotional disorders, behavioral disorders, sleep disorders etc are most commonly seen. They have behavioral problems because they are mostly isolated by their friends and people around them; friends and batch mates bully them for their looks.

Investigation

Sturge-Weber syndrome can be diagnosed using

1. MRI of the brain with and without contrast

Best structural imaging is best seen by magnetic resonance imaging; it allows all aspects of SWS except for calcification, which is best seen using computed tomography.[17] MRI is used to see the intracranial extent and distribution of leptomeningeal angiomatosis with gadolinium-DTPA.[18] Magnetic resonance venography can also be used. MRI shows accelerated myelination, no venous occlusion, an enlarged plexus, and gadolinium enhancement.

2. A CT of the brain is needed to detect the calcium deposit. CT shows calcifications of the brain that are not visible on normal radiographs.[19]

A CT scan reveals calcification, tram track calcification, cortical atrophy, abnormal draining veins, an enlarged choroid plexus, contrast enhancement, and, during seizures, blood-brain barrier breakdowns.

3. EEG (Electroencephalogram)

It is used for children who have seizures.

The use of EEG in diagnosing brain illnesses, particularly those involving seizures and epilepsy disorders, reveals variations in brain activity. It is also used to identify other disorders of the brain, such as migraines, vertigo, brain tumors, and others. Using tiny metal discs or electrodes placed on the scalp, it measures the electrical activity of the brain. It is a minimally invasive process. Finding epilepsy is the main goal. The brain signal associated with anxiety and depression is also identified by EEG.

EEG exhibits polymorphic delta activity, epileptiform characteristics, and reduced background activity. Generalized tonic-clonic seizures, myoclonic seizures, and absence seizures are the types of seizures that are visible on an EEG. It is a painless operation that is safe to do.

Sturge Weber syndrome EEG score[20]

0 Normal
1 Focal asymmetry (slowing or loss of normal background activity)
2 Sporadic, unilaterally sharp waves (often occipital)
3 Frequent, at times semi rhythmic, unilateral runs of spikes (often occipital). Occasionally, with secondary generalization[20]

1. Ophthalmic examination to rule out glaucoma
   Eye angle exam
   Corneal thickness measurement
   Visual field test
   Dilated eye exam
2. An eye exam including pressure measurements
3. CSF analysis to check for elevated proteins
   CSF analysis is a test in which a group of laboratory tests are performed to measure the chemicals in cerebrospinal fluid.
   Increased protein levels may cause tumors, bleeding, inflammation, or injuries to the nerves.
4. Radiograph
   Radiographs show tram track lesions.
5. Angiography
   Angiography is an imaging technique used to visualize the lumen of blood vessels and other organs of the body, mainly the arteries, veins, and heart chambers; aberrant tortuous vessels; and non-filling dural sinuses. Angiography shows the venous stasis, which is a feature, or SWS. Angiography is mainly used to check the health of blood vessels and the flow of blood through them.
6. A single-photon emission computed tomography (SPECT) scan is done to analyze body organs, tissues, and bones. It is used to detect altered blood flow in the brain and diagnose brain disorders. SPECT scan initial hyperperfusion is followed by hypoperfusion. SPECT reveals hyperperfusion and hypoperfusion. SPECT is an important tool in the presurgical evaluation of SWS. It is seen that the area with intense hyperperfusion reflects the ictal onset zone.[21] The disadvantage of a SPECT scan is that it produces low-resolution images that are prone to artefacts and takes a long time to scan.

Spin-echo T1- and T2-weighted pictures, together with the administration of gadolinium contrast, are included in the whole evaluation of SWS. Find out how extensive the vascular irregularity is. T1-weighted images are essential.[22,23] Areas of hypoperfusion in patients with SWS can be seen on advanced single-photon emission computed tomography. MRI and single-photon emission computed tomography, if used in conjunction, can show more remote areas of involvement.[24] EEG is the most important modality in the diagnosis of SWS, followed by MRI and CT scanning.

Treatment

Treatment for SWS is symptomatic, that is, there is no specific treatment for this disease, but symptoms of it are treated. In newborns with SWS, impaired blood flow and convulsions can result in brain injury. Brain involvement can be treated with anticonvulsants or low-dose aspirin. Studies have shown that low-dose aspirin is beneficial in reducing seizures and stroke-like episodes. Medical care includes seizure anticonvulsants, symptomatic and prophylactic therapy for headache, and laser therapy. Medicines mostly given for seizures are oxcarbazepine, valproic acid derivatives, phenytoin, carbamazepine, etc., The parents of patients with seizures should be educated on how to recognize and treat seizures.[7] Sturge-Weber syndrome epilepsy can be challenging to manage since it manifests as clusters of seizures and bouts of status epilepticus.[25] To permanently exclude seizures, one should provide rectal diazepam for infants who are older than 3 months. Drug-resistant patients are treated with surgery for the removal of epileptic brain tissue. 30% to 50% of seizures are refractory. Surgery is necessary: VNS, hemispherectomy, corpus colostomy, and focal cortical resection stimulation of the vagus nerve. Seizures with SWS can be improved by hemispherectomy.[26]

Glaucoma medications: Almost 30% to 71% of Sturge-Weber syndrome patients suffer from glaucoma. It can be treated with topical medication as well as surgically. The most common topical medication is latanoprost.[27] Beta antagonist eye drops for the production of aqueous fluid, adrenaline eye drops and motivation eye drops for the drainage of fluid, and carbonic anhydrase inhibitors eye drops, commonly non-specific beta-blockers or prostaglandin analog drops, are mostly first-line medications to reduce intraocular pressure. Individuals with glaucoma should refrain from eating a lot of trans fats since they harm the visual nerve. The treatment of this SWS condition is frequently challenging. The potential for hemorrhage from the dilated aberrant episcleral and choroidal arteries makes surgery in these eyes highly risky.[28] Headaches and migraines are treated with preventive medications. Triggers for migraine should be identified and treated accordingly. Preventive medications like valproate and gabapentin are seen as effective medications in cases of both seizures and migraine.

For the port wine birthmark, there is no specific medication, and it is only treated with laser surgery. Lasers can be used to lighten the birthmark. It is less damaging sometimes and does not cause scarring of the skin. Sometimes the pigmentation gets darker due to layers; that is why it is not a permanent treatment.[25]

Counseling is the best option to treat patients with anxiety and depression. Counseling patients about their lifestyle could be very effective for the behavioral problems seen in patients with SWS.[29] Motivate them to do things of their interest; ask them to involve them in more activities that will help them live better. Most important is to educate their parents about their behavior and to guide them to pay more attention to the psychological behavior of these children; proper treatment should be taken at an early stage.[30] Sleep deprivation and exhaustion, changes in routine, stress, and minor head trauma were found to be the most frequent precipitating causes for headaches, and this information is helpful in counseling patients and their families.[31]

Young patients with SWS are at risk for various problems like behavioral and emotional functioning. Studies have shown that these children have developmental and social problems. Psychological problems are a part of an individual's life, usually seen in one's work, health problems, relationships, etc., Studies have shown that patients suffering from SWS can experience psychological problems. They face problems since childhood; children feel shameful because of their appearance. Initially, they do not feel much about their birthmark, but as they grow up and start going to school, their friends and colleagues start bullying them, calling them "ugly." Due to all this bullying or mocking, they become introverted, aggressive, and irritable. They present with a port wine stain over their forehead. Girls face more problems throughout their lives. Girls are always conscious of their looks, and having a birthmark on their face makes them feel shameful. These are major problems they face during their marriage. Studies have shown that these patients are mostly very aggressive, and sometimes they become introverts.[5] Children always have a fear that friends or classmates will harm them because of their looks. They used to spend all their time alone at home and become agitated and nervous. They withdraw themselves from friends and all the other activities. As they grow, they start having fears of not being accepted by society. They feel like they are unwanted. They start feeling sad or down. These children need more attention, but their parents are not more attentive toward them, which may be the reason for their psychological behavior. All of which lead to confused thinking, a reduced ability to concentrate, and an inability to cope with daily problems or stress. They have trouble understanding and always use to relate their situation to others, thinking why they are different from others. Extreme mood changes between high and low can also be seen. These patients have suicidal tendencies, too. Suicidal thoughts and behavior are common in some of the patients with SWS.[6] They think of hurting themselves or sometimes the ones who used to bully them. Psychological problems are also seen in this patient, like anxiety, depression, schizophrenia, eating disorders, a feeling of guilt, sleeping problems,

excessive anger, violence, etc., Behavioral issues like emotional disorders and dissociative disorders are also seen. Children with SWS have learning problems or developmental delays. Angiomas in parts of the brain affect learning. Their communication skills are lost; they keep their feelings to themselves. For this, children should be taken for regular checkups to ensure they are learning, talking, and playing in the right ways according to their age. Behavioral disorders, also called disruptive behavioral disorders, are the most common reason that parents are guided to take their kids for health assessments and treatment. Counseling can be done for patients with anxiety and depression. Counseling can be very useful for these patients. They are motivated to do things that are in their interests. Care should be taken that they are communicating properly with friends and parents, sharing their feelings, and not being isolated, so that they become free and confident. Communication helps in more serious physical illnesses as well, to make them feel good, to be empathetic, and to build their confidence. Patients with severe illnesses need patient–doctor communications that make their health more effective. Such patients with psychological behavior can be taken care of. Communication can help these children build their confidence. They should pay more attention and get routine checkups and medical care. Psychological aspects are never taken care of in patients with SWS and are always ignored; this should be given more attention. And this area should be more focused, and more research should be done.

Conclusion

After studying the articles, it is apparent that the psychological aspect is not more focused on and is always ignored by parents and families. This review is focusing on the psychological aspect of patients with SWS. Patients have various psychological behaviors that should be evaluated and taken care of. Behavioral problems like suicidal feelings, anxiety, depression, and mood disorders are seen in these patients, and parents and carers are educated to take proper care of them. They deserve more attention and care. Psychological problems are one of the serious issues in SWS and should be properly evaluated and addressed. More importance should be given to this psychological aspect. And more research and study should be done on this aspect.

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Conflicts of interest

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