Case Report

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Encephalotrigeminal Angiomatosis: A Case Report

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ABSTRACT

Background: Encephalotrigeminal Angiomatosis is an infrequent congenital disorder of vasculature of eyes, face and brain. It manifests with glaucoma, port wine stain over the face and seizures. It is commonly known as Sturge-Webber Syndrome (SWS) and comes under the group of "Phakomatosis". There are three classic variants of Sturge-Weber Syndrome based on involvement of the eye, skin and central nervous system. They are - Complete Trisymptomatic, Incomplete Bisymptomatic (either oculocutaneous or neurocutaneous) and Incomplete Monosymptomatic (only neural or cutaneous involvement). This is a case report of a 23-year-old male patient with loss of vision in the right eye, port wine strain involving the complete right side of face and bilaterally over the neck, hypertrophy of the face with history of seizures. The intraoral findings were hemangioma of the right of soft palate, over the entire right buccal mucosa, right side of upper and lower labial mucosa, with hypertrophy of the right side of maxilla and deranged occlusion. Various radiological and hematological investigations like IOPAR, OPG, CT, CECT, MRI were performed. All the radiological features are suggestive of Sturge-Webber Syndrome.

Keywords: Encephalotrigeminal Angiomatosis, trigeminal nerve, dentistry.

INTRODUCTION

Encephalotrigeminal Angiomatosis or Encephalofacial Angiomatosis, more commonly known as Struge-Weber Syndrome (SWS) is a rare, non-familial, congenital disorder which is classified under the spectrum of Phakomatoses. [1,2,3] Phakomatoses (mother spot disease) or Neurocutaneous syndromes affect the central nervous system (CNS), skin, viscera and other connective tissues can also be involved. [4] More than 30 different conditions are included in this spectrum of disease. [4]

Struge-Weber syndrome is a congenital disorder involving the vasculature of meninges, brain, face, eyes and is often associated with neurological complications such as seizures, mental retardation, contralateral hemiparesis and glaucoma. [5] SWS occur sporadically with equal prevalence in males and females and across all races; the prevalence rate is 1 in 20,000-50,000 births. [6]

SWS was first described in 1860 by Schimrer, later in 1879 by William Struge and then in 1897 by Kalischner. The first radiographic evidence i.e. intracranial calcification was described by Weber and Dimitri in the year 1922 and thereby labelling it as a complete syndrome. [3] In 1934, Krabbe proved that calcification seen on roentgenograms of skull were actually present in the atrophic cerebral cortex. [3]

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Somatic mutations in some affected areas lead to the development of SWS. [5] It is caused primarily by a somatic activating mutation in the GNAQ gene encoding for the G protein alpha subunit Gαq, R183Q. [6] There are 3 classical variants of SWS:

1. Complete Trisymptomatic: All the 3 organ systems are involved – eye, skin and CNS [3]

2. Incomplete Bisymptomatic: Involvement of 2 organ systems; either oculocutaneous or neurocutaneous. [3]

3. Incomplete Monosymptomatic: Only the neural or cutaneous system is involved [3]

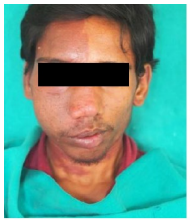
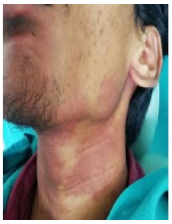
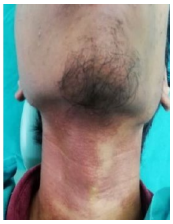

Fig 1: Portwine stain on right side of face. Fig 2: Portwine stain on left lower third of face and neck. Fig 3: Portwine stain on anterior surface of neck.

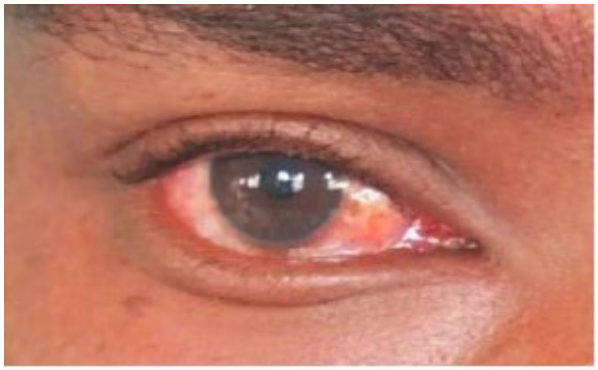

Fig 4: Redness of the sclera of right eye.

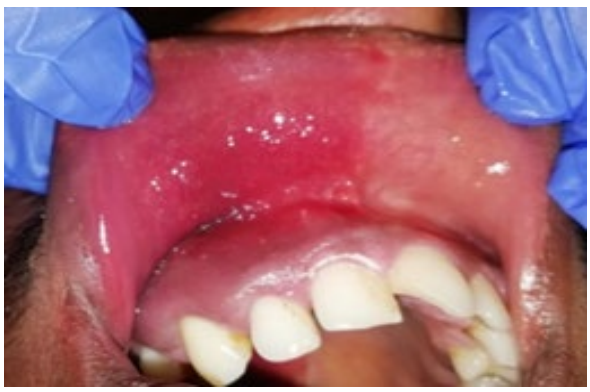

Fig 5: Upper labial mucosa.

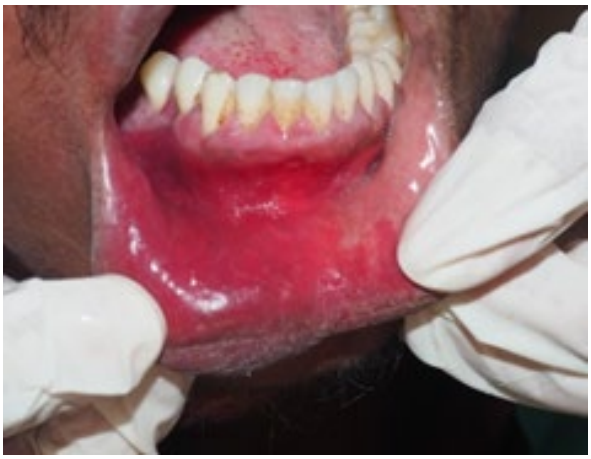

Fig 6: Lower labial mucosa.

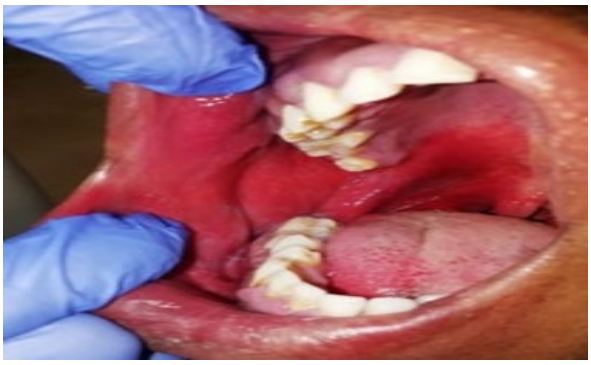

Fig 7: Right buccal mucosa.

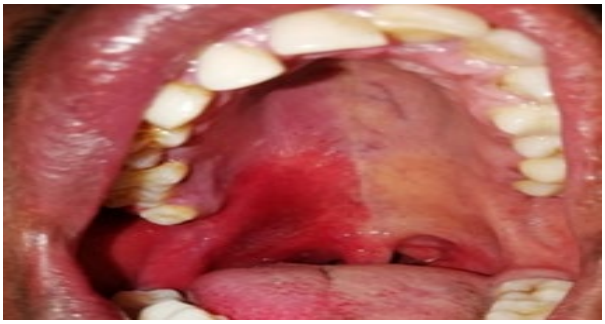

Fig 8: hard and soft palate.

CASE REPORT

A 23 year old male patient reported to the department of Oral Medicine and Radiology, with a complaint of pain in the right side upper jaw back tooth region from 10 days. Patient's past medical history revealed that he was born with a reddish birthmark/ patch on the right side of face which gradually increased in its size, extended up to the left side of ear and neck. Patient had also revealed that he had intermittent redness in the right eye since 5-6 years and had decreased vision since 2

years, which gradually led to loss of vision in the right eye except light perception.

Patient had nearly 7-10 episodes of seizures since 1 year of age and all are accompanied with extreme weakness in the lower limbs, dizziness and occasional vomiting. Extraoral examination reveals facial asymmetry due to hemi facial hypertrophy of the right side of face. Port wine stain is seen over the entire right side of face and neck including the ear, extending from the hairline to 1cm above sternum/ clavicle (fig.-1). On the left side, the stain is seen over the neck and in the lower third of face up to the ear lobule (fig-2). The stain is seen only on the anterior surface of neck, no involvement of the posterior surface of neck (fig-3). Redness was seen in the right eye (fig-4) with protrusion of the right eye indicative of Buphthalmous.

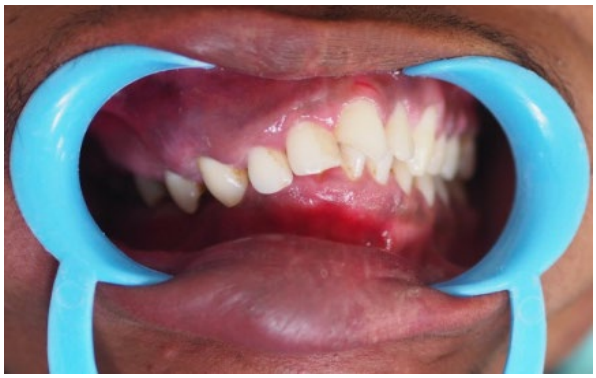

Fig 9: Deranged occlusion on right side.

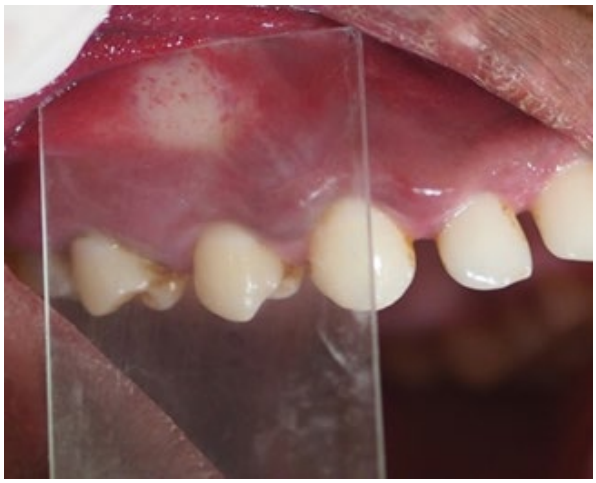

Fig 10: Diascopy.

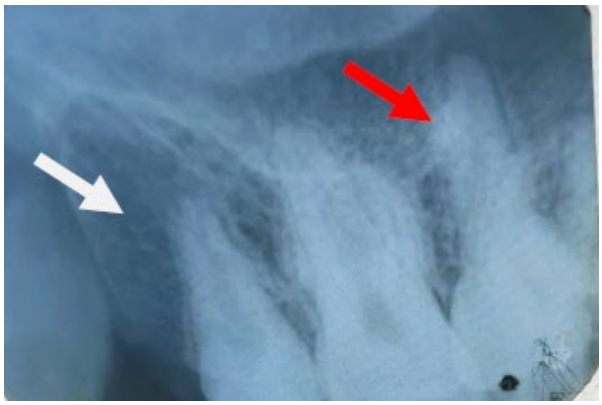

Fig 11: IOPAR of 16 tooth region.

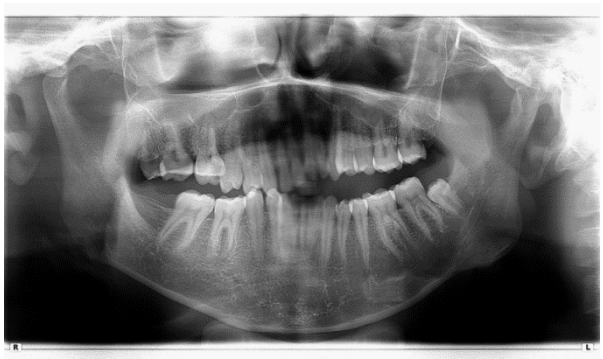

Fig 12: Orthopantamogram.

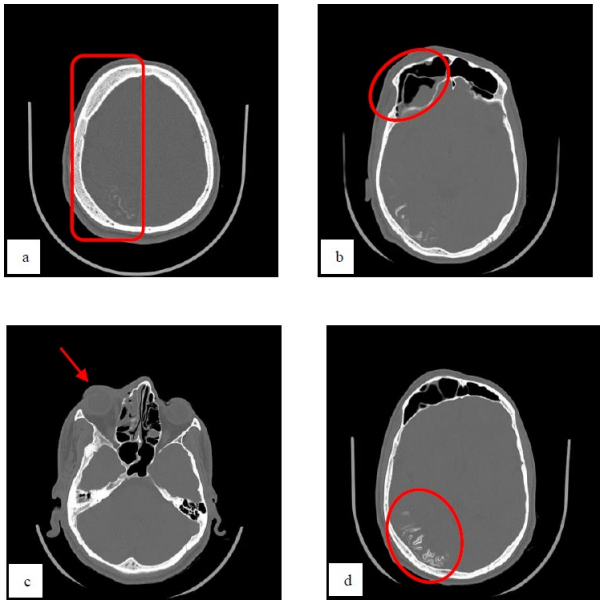

Fig 13: Computed Tomography – Axial Sections of Brain (a) cerebral hemiatropy with calvarial thickening (b) polypoidal thickening in the right frontal sinus (c) protrusion of the right eyeball (d) Cortical and sub-cortical tram track like calcifications.

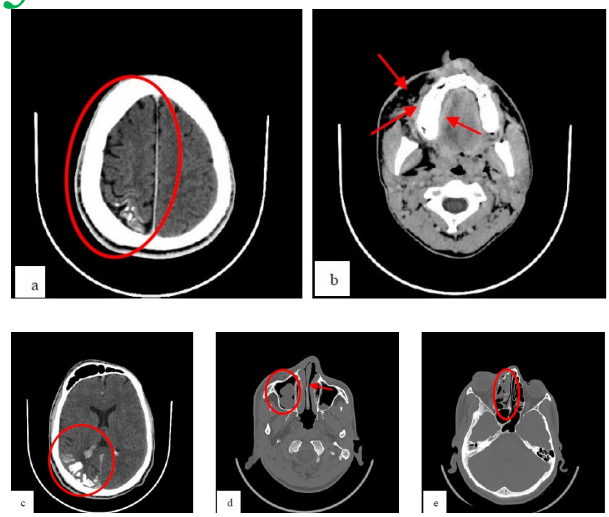

Fig 14: Contrast Enhanced Computed Tomography – Axial Sections of Brain. (a) leptomeningeal enhancement in the right cerebral hemisphere (b) subcutaneous vascular channels in face and hemihypertrophy of right side of maxilla. (c) cortical and sub-cortical tram track like calcifications and enlargement of the right choroidal plexus (d) Polypoidal mucosal thickening in bilateral maxillary sinuses & deviated nasal septum (red arrow) (e) Polypoidal mucosal thickening - ethmoidal sinuses.

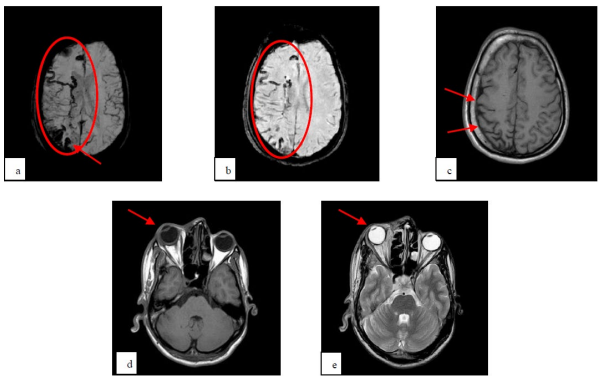

Fig 15: Magnetic Resonance Imaging- axial sections. (a) SWI imaging showing multiple 'black blood' vascular channels with blooming artifacts (red arrow). (b) VenoBOLD images showing multiple tortuous venous channels. (c) increase in calvarial thickness and reduced cerebral volume. (d) T1 weighted image showing proptosis. (e) T2 weighted image showing proptosis.

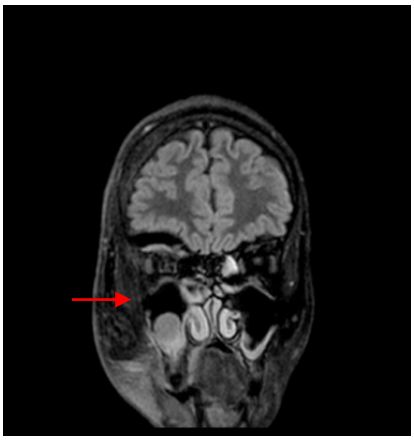

Fig 16: MRI coronal section showing subcutaneous enlargement of right side face.

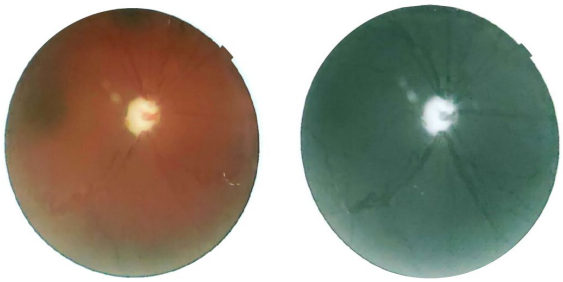

Fig 17: Optical Coherence Tomography of right eye.

Intraoral examination reveals the upper and lower labial mucosa (fig-5&6) showed reddish discolouration on the right side without crossing the midline. Reddish discolouration is seen over the right buccal mucosa (fig-7), right side of posterior part of hard palate, soft palate and uvula (fig-8) extending to the midline. Hypertrophy was seen in the right half of maxilla involving the alveolar ridge, leading to derangement in occlusion with upper right teeth completely overlapping the lower right teeth (fig-9). Teeth indentations were seen over the right lower alveolar mucosa distal to 47. Tongue is normal in its size and colour. The gingiva on the right side shows increase in size and the colour of the gingiva was reddish purple. On palpation, the hypertrophied swelling of right half of maxilla was non-tender, ill-defined, hard in consistency and non pulsatile. Deep dental caries was present in relation to 16 tooth and it was also tender on percussion. On performing diascopy, blanching was observed (fig-10) and rebound phenomenon (return of colour) was also observed on removal of the glass slide/ pressure. All the required haematological and radiological investigations were performed. The haematological investigations such as complete

blood picture, bleeding time and clotting time were unremarkable. Radiographic investigations included conventional and advanced radiographic imaging techniques. Conventional techniques were intraoral periapical radiograph in relation to 16 tooth region, orthopantomogram and lateral cephalogram. Advanced imaging procedures included Contrast Enhanced Computed Tomography (CECT), Magnetic Resonance Imaging (MRI) and Optical Coherence Tomography (OCT) for both eyes.

Intraoral periapical radiograph (IOPAR) of the right upper first molar tooth region revealed widening of PDL space and loss in continuity of lamina dura in the apical third of root of right upper first molar with a diffuse ill-defined radiolucency at the apex; these features suggestive of periapical abscess (fig-11 red arrow). Bony trabeculae show sparse arrangement with spacing in between them (fig-11 white arrow). Orthopantomogram reveals haziness with entire right maxillary sinus, mucosal thickening seen in the inferior aspect of left maxillary sinus, supraerupted teeth 11,12,13,14,15 & Deep dental caries involving pulp leading to periapical abscess in relation to 16 (fig-12).

CT (fig-13) and CECT of brain (fig-14) reveals leptomeningeal enhancing thickening over the right parietal lobe and multiple prominent transparenchymal venous channels in the right cerebral hemisphere (fig-14a). Multiple prominent enhancing subcutaneous vascular channels were noted in the right side of face (fig-14b). Cerebral hemiatrophy noted on the right side predominantly in high frontal, parietal and occipital regions with compensatory adjacent calvarial thickening (fig-13a). Cortical and sub-cortical tram track like calcifications were seen involving right temporal, parietal and occipital lobes (fig-13d, 14c). Frontal sinus enlargement was noted(fig-13b, 14a, 14c). Polypoidal mucosal thickening noted in bilateral frontal, ethmoidal and maxillary sinuses (fig-13b, 14d, 14e). Deviation of the nasal septum was present (fig-14d). Asymmetry in maxilla noted on right side with mild displacement towards left side (fig-14b). Hypertrophy of maxilla is noted with irregular appearance of the external cortical surface (fig-14b). Protrusion of the right orbit was noted (fig-13c).

MRI (fig-15) of brain reveals multiple tortuous vascular channels in the perimesencephalic cistern and periependymal regions of the right lateral ventricle and right parasagittal frontal regions (fig-15a, 15b). Subtle volume loss was noted in the right temporo-occipital and parietal region in the form of dilated cortical sulci, when compared to the opposite side (fig-15d). Gyriform blooming artifacts are seen at right temporo-occipital and parietal regions, which could represent calcifications (fig-15c). Proptosis of right eyeball noted (fig-15e, 15f). Subcutaneous enlargement of right side face is noted (fig-16).

Optical Coherence Tomography of the right eye showed blurring of the optic disc margins along with disc pallor (cup:disc ratio was 0.6:1). Multiple tortuous blood vessels were noted suggestive of choroidal hemangioma. Intraocular pressure was raised to 36 in the right eye, and it was 16 in the left eye. These features were suggestive of glaucoma. (fig-17).

Patient was initially prescribed with medication for his tooth pain & referred for endodontic therapy. After ophthalmic examination he was advised acetazolamide, 250mg (Dimox) and 0.15% w/v bromonidine tartrate and 0.5% w/v timolol malate (Brimolo T) ophthalmic solutions for glaucoma correction. He was also given 0.5% w/v loeprednol etabonate (L-Pred) for symptomatic relief (redness of eye).

DISCUSSION

Encephalotrigeminal Angiomatosis is an uncommon, congenital and sporadically occurring group of disorders which are collectively classified under Phakomatosis ie, mother spot (birthmark) diseases. The color of the birthmark may vary from pink to deep purple depending upon the plethora of capillaries. [7] Prevalence is equal in both sexes and affects about 0.3-0.5% of newborns. [8, 9]

Diverse pathophysiogenetic mechanisms have been suggested for the vascular malformations in SWS such as venous dysplasia of the emissary veins in the intracranial circulation, neural crest alterations leading to alteration of autonomic perivascular nerves, and mutation of the GNAQ gene. [10] A somatic mutation of GNAQ gene which is present on chromosome 9q21 has been regarded to cause SWS. [10] Signals for formation of protein guanine nucleotide binding protein G (q) subunit alpha

(Gαq) is provided by GNAQ gene. [7] Development and function of blood vessels is regulated by a group of proteins that include Gαq protein. [7, 9] The GNAQ gene mutation that causes SWS results in the production of a protein with impaired function which causes the abnormal and excessive formation of vessels before birth. [7, 9]

SWS as proposed by Howard Royle to be the consequence of a defect in development during the 1st month of intrauterine life (IUL), where the vascular plexus persists around the cephalic end of the neural tube. [3, 5] This plexus originates in the 6th week of IUL but usually undergoes regression in the 9th week of IUL. [5] The appearance of facial nevus and cerebral angiomatosis is thought to be due to the development of facial skin from the ectoderm covering this plexus. [5]

Monte A Del postulated that insult to the precursors of tissues originating from promesencephalic and mesencephalic neural crest develop into vascular malformations in the meninges, eye, and the dermis. [3]

The hallmark features seen in this syndrome are malformations of the vasculature of the leptomeninges, facial skin and the eye. The leptomeningeal angiomatosis is commonly seen involving the occipital and posterior parietal lobes. Ipsilateral lobe involvement is quintessential, although there have been reports of involvement of the contralateral lobes. [1, 7, 10] Nevus flammeus or facial nevus is often seen in the distribution of the ipsilateral trigeminal nerve course along all the 3 of its branches. [1] Other associated traits include glaucoma, seizures, headaches, neurological and behavioral deficits. [1, 8, 11] Contralateral hemiparesis, hemiatrophy and homonymous hemianopia have also been manifested. [1]

The clinical features can be descrided as follows:

1. NEVUS FLAMMEUS

Nevus flammeus or facial nevus or portwine stain is macular progressive lesions that are generally but not always present right from birth. [1, 12] Portwine stain is named after Portuguese fortified red wine which has the same colour as the macular lesion in SWS. [8] Classically it is manifested along one or more branches of the trigeminal nerve. [3] Enjolaras et al observed that SWS typically shows involvement of the Ophthalmic division (V1) of the trigeminal nerve. They have concluded that the involvement of V1 is essential for establishing the diagnosis of SWS. [1, 3] Bioxeda et al observed that extrafacial involvement was common when mandibular division (V3) was involved. [1, 3]

The portwine stain is usually seen in one side of the face and rarely occurs on both sides. [1] The colour may vary from light pink to deep purple which is determined by the myriad proliferation of capillaries beneath the dermis. [1, 3, 10] The portwine stain may also be visualized on other parts of the body, like upper limb, although rare. [1] The extent and size of the portwine stain cannot be used to determine the amount of brain involvement.

2. NEUROLOGICAL ABNORMALITIES:

Focal motor seizures are frequently present in patients with SWS. Seizures usually develop in the first year after birth. Management of seizures is one of the important aspects in the management of SWS. [1, 3]

Angioma (excessive blood vessel growth) on the surface of brain, more commonly in the parietal or the occipital regions leading to abnormal brain function is the primary neurological symptom. [3] Intractable seizures might lead to motor deficits and mental decline. [1] Cortical irritability due to cerebral angiomas, through mechanisms of hypoxia, ischemia and gliosis results in seizure. [1, 3]

The median age of onset of seizures was 6 months, but seizures can manifest anytime ranging from birth to 23 years of age, according to a survey of cases by Struge-Weber foundation. [3] It was also reported in the survey that about 75% had onset of seizures during the first year of life, 86% before the age of 2 years and 95% before the age of 5 years. [3]

Onset of seizures before the age of 2 years has a greater chance for development of refractory epilepsy and mental retardation as refractory seizures have more extensive brain involvement. [2, 3] Prolonged seizures leads to neurologic injury which is secondary to metabolic disturbances like hypoxia, hypoglycemia, hypotension, ischemia and hyperthermia. [3] Episodes of status epilepticus prove to be very dangerous in SWS.

Transient weakness may frequently occur with seizures and it increases with recurrent seizures. Transient hemiplegia maybe accompanied by migraine episodes. [3] As observed by Cho et al, an important distinguishing factor is migraines which are associated with prolonged visual auras and hemiplegic attacks in SWS. [6] In the study by Cho et al, a positive association between seizures and the presence of a port-wine birthmark on the forehead and scalp was also observed. [6]

3. OCULAR MANIFESTATIONS:

Choroidal angioma is a classic ophthalmic lesion. Choroidal, conjunctival hemangiomas and heterochromia of iris and glaucoma are the ocular abnormalities in SWS. [12] Glaucoma i.e. incresead intraocular pressure is seen in almost 30% of cases with SWS. [3] Glaucoma is usually absent when nevus is not present in V1 distribution; therefore patients without nevus also may not have glaucoma. [13] Its onset is bimodal, early onset is between birth to 4 years of age and in late onset glaucoma may develop around the time of adolescence. [1, 3]

Early onset glaucoma manifests as Buphthalmous, due to abnormality in the anterior chamber angle and outflow tract. [1] Late onset glaucoma presents with elevated episcleral venous pressure which might show little or no enlargement. [1]

Diffuse choroidal vascular malformations are yet another characteristic feature of SWS.[1] These cause progressive secondary changes in the overlying retina such as retinal pigment degeneration, fibrous metaplasia and cystic retinal degeneration leading to loss of vision and visual field defects. [1, 3] Continuous exudation from the malformation might lead to retinal detachment. [1]

4. ORAL FEATURES:

Oral manifestations of SWS were seen in about 38% of cases in a review of 111 cases by Gorlin and Pindborg. [1] Lips and cheeks are most commonly affected and it rarely involves the palate and mandibular gingiva. [1, 3] The lesions present as red to purple flat patches that show blanching on pressure i.e. diascopy is positive. [1, 3] The gingiva is usually red and enlarged, may present as monstrous masses hindering mouth closure. [3] The color typically stops at the midline of maxilla. [3] The severity of lesion can vary from a minor vascular hyperplasia to a large angiomatous lesion. [14] Portwine stain lesion of oral mucosa along with the hypervascular changes is also observed. [14]

Gingival haemorrhage with minor trauma is a characteristic feature of SWS. [14] These vascular lesions are prone to risk of severe intra and postoperative hemorrhage and hence quarantine special attention during oral surgical procedures where bleeding is expected, such as periodontal surgery, tooth extraction etc. [1] Analysis of panoramic and intraoral radiographs suggest the involvement of alveolar, periodontal and pulpal structures, and alteration of vascularity in the jaws which might result in altered eruption sequences and also early eruption of permanent teeth thereby causing malocclusion. [1]

Ipsilateral cheeks, palate and rarely tongue appear reddish purple and show hypertrophy.[5] Macrochelia is also frequently associated. Hard tissue involvement presents as bone growth and results in malocclusion and facial asymmetry. [5]

IMAGING

The typical imaging features of SWS reflect the presence of angiomatosis and the sequelae of related hemodynamic disturbances. [15] MR imaging is currently the imaging investigation of choice, with pial enhancement on post gadolinium sequences considered the gold standard in assessing the extent of disease and regarded by some as being the most important criterion in the radiologic diagnosis. [15]

Imaging techniques that can be done to establish diagnosis of SWS include skull radiographs, angiography, CT scan, MRI, MRI with gadolinium, single positron emission computed tomography (SPECT) and positron emission tomography (PET). [3, 16]

1. Skull radiographs: The archetypal finding on skull radiographs, especially lateral skull radiograph are “tram-line” or “tram-track” or “trolley-track” or “railroad track” calcifications. [1, 3, 5] These are often a late finding and may not be present in the initial stage of disease. [1, 3] This sign is produced by cortical calcifications that result from leptomeningeal vascular malformations. [1] Theses vascular malformations consist of simple vascular

structure situated along the space between piamater and the arachnoid membrane. [1]

Other indirect signs that may be seen include cranial asymmetry, thickening of the cranial diploe and increased sinus sizes on the side of the portwine stain. [5] Orthopantomograms may show unerupted teeth and osteohypertrophy of the involved side. [5]

2. Angiography: It demonstrates a lack of superficial cortical veins, nonfilling of dural sinus and abnormal tortuous veins. [3]

3. CT scan: Neonates and infants may show calcifications on CT images. [1] Ipsilateral cortical atrophy, ipsilateral choroid plexus enlargement, abnormal draining veins and a breakdown of the blood-brain barrier with seizures. [1, 3] Cortical and intracranial calcifications underneath the leptomeningeal angiomas are best seen on CT scans. [5] The have a characteristic gyriform or "S" shape. [5, 17] They may be found at the cortex as well as at the meningeal arteries cortical, and subcortical veins. [5] They are commonly associated with cortical atrophy in the same area. [5] Atrophy and calcifications are considered to be an indirect consequence of chronic ischemia of the cortex due to vascular stasis in the area of leptomeningeal angioma. [5] An ipsilateral prominent choroid plexus may be seen on contrast-CT, with or without calcifications. [5]

CT is better than MR in detecting the presence and extent of cortical calcifications, and equal to MR in showing the prominent choroid plexus. [18]

4. MRI and MRI with gadolinum: Magnetic resonance imaging without contrast may show cerebral atrophy both in T1- and T2-weighted sequences, diploic prominence, and enlarged choroid plexus. [5] Calcifications may be seen in T2-weighted, especially, in gradient-echo images, as areas of low signal. [5] Accelerated myelination may produce the areas of hypointensity in T2-weighted. Hyperintensity on T2 weighted representing areas of gliosis may also be seen. [5]

MRI with gadolinium enhancement shows pial angioma. [1, 3] It may also show accelerated myelination around the leptomeningeal angioma, enlarged choroid plexus whose size correlates with the extent of leptomeningeal angioma. [3]

MR imaging, with and without contrast administration, is better in showing the extent and degree of patency of the leptomeningeal malformation, the degree of parenchymal atrophy, the changes in the affected gray and white matter, the parenchymal venous anomalies, and the diploetic prominence on the affected side. [18]

5. Single positron emission computed tomography (SPECT): It measures the cerebral blood flow and it demonstrates hypoperfusion in the area of pial angioma, and hence can detect a latent angioma which may not be seen in other techniques. [1, 3, 19] Pinton elicited that the cortex is hypoperfused during the first year of life before the first seizure and hypoperfusion after first year of age, even in the absence of epilepsy. [1, 3]

6. Positron emission tomography (PET): This demonstrates metabolic abnormalities in the structurally affected hemispheres that extend beyond the anatomic abnormalities detected by CT scan. [1, 3, 19]

7. EEG (Electro Encephalogram): This modality is used to show reduced background activity, Polymorphic delta activity and Epileptiform discharge. [19] The posterior head region (PHR) most commonly shows epileptiform discharges followed by occipital lobe and then temporal, parietal, and frontal lobes with decreasing frequency, respectively. [13] Some patients have frontal spikes, which can be explained by suppression of the background activity in the PHR. [13]

• Medical Treatment of SWS [22]

The current approach of medical treatment is anticonvulsant treatment and anti-platelet treatment with prophylactic low dose aspirin. In view of the possibility of neurological deficit with seizure clusters, episodes of seizures should be treated aggressively and prolonged seizures avoided where possible. Maintenance anticonvulsant treatment is indicated in infants and children with seizures and should be considered after one seizure episode. At present, there is no evidence base to suggest one particular anti-epileptic drug above another, thus use of local clinical guidelines is recommended. Acute rescue treatment of seizures is with benzodiazepines or if

ineffective, intravenous phenytoin or phenobarbitone is recommended. The rationale for the use of low dose aspirin is that venous stasis may lead to the potential for thrombus formation. Maria et al. reported fewer stroke like episodes in a retrospective series of children with SWS treated with aspirin prophylaxis compared to those not receiving treatment. In this series aspirin was well-tolerated.

Table 1: Differential Diagnosis.

S.No	Syndrome	Differentiating Features
1	Rendu-Osler-Weber syndrome	Autosomal dominant, multiple telangectasis on skin, significant oral involvement, occasional GI tract involvement, arteriovensous malformations
2	Maffucci syndrome	Hemangiomas of the mucous membrane and dyschondroplasia. asymmetric enchondromas and haemangiomas, affecting the skin and skeletal system.
3	Klippel Trenaumy-Weber syndrome	Portwine stain, varicose veins, and bony and soft tissue hypertrophy affecting the extremities
4	Beckwith-Wideman syndrome	Capillary malformation in forehead and upper eyelid region which mimics portwine stain. Hemihypertrophy, skin angiomatosis, omphalocele and macroglossia
5	von Hippel-Lindau syndrome	Hemangiomas of the cerebellum or retina, cysts of viscera
6	Shapiro-Shulman syndrome	Bilateral facial nevi and abnormal venous drainage
7	Divry-Van Bogaert syndrome	Leptomeningeal angioma (non-calcifying) with diffuse sclerosis, progressive neurological disorder, and livedo reticularis

Treatment:

• Surgical Treatment in SWS [22]

Surgical treatments for SWS have been used for over 30 years. The rationale behind their use has been the traditional epilepsy surgery model of removal or disconnection of the epileptogenic tissue, which in most cases requires hemispherectomy. Lobar or multilobar resection can however be successful in appropriately selected children. Seizures often reduce in frequency, the progressive cognitive decline may be halted or show some recovery. Acquired motor deficits in infants or children with an established hemiplegia following hemispherectomy are often minimal. The timing of such surgery appears to be crucial. Treatment very early in life, often before one year, appears to be more successful in abolishing seizures and ensuring more normal development. The difficulty with advocating early surgery for children with SWS, is predicting those that will follow a course with severe episodes of encephalopathy in order to select those children who might benefit from early hemispherectomy. In those children where involvement is more localized, lobar or multilobar excision may be associated with improvement in seizures and may minimize post-operative deficits. In a recent series of 14 operated cases, in seven cases the angioma was entirely removed; complete and lasting seizure control was obtained. In the remaining cases, only one achieved complete seizure control.

Corpus callosotomy may lead to improvement for children who have drop seizures as the major seizure type. Epilepsy surgery has not generally been used for those with bilateral angiomas. However, Tuxhorn and Pannek report hemispherectomy with improvement in seizure control and improved function in three cases with bilateral angiomas. It is recommended that infants and children with SWS are evaluated at a center with expertise in pediatric neurosurgery if seizures occur or if seizures are associated with developmental delay or progressive neurological deficit.

• Symptomatic Treatment in SWS:

Symptomatic treatment is usually followed for struge weber syndrome. [3] Management of SWS requires a multidisciplinary approach and it largely depends on the organs that are affected. [1, 19] A team of neurologists, ophthalmologist, dentists, dermatologist, plastic surgeons, neuroendocrinologists, psychologists and nurses are required based on the type of SWS. [3, 19] A multitude of modalities such as sclerotheraphy, embolization, laser photocoagulation, medical management and surgical excision have been employed. [3]

Scelrotheraphy advocates the direct injection of sclerosing agents into the lesion which act on endothelial surface by causing destruction of thrombosis and thereby resulting in fibrosis and obliteration of vascular lumen. [8] Categorization of

MRI findings in vascular malformations also provides information for the prediction of the probability of therapeutic success from sclerotherapy. Grade I lesions are well defined and less than 5 cm. Grade II lesions are either poorly defined or greater than 5 cm. Grade III lesions are poorly defined and greater than 5 cm. Lower grade lesions have better prognosis. Choice of the sclerosing agent depends on clinician's preference, dimensions and extent of the lesion, and draining veins within the lesions. [8]

Laser therapy can be used to lighten or remove the portwine stain, and these are advised early on for better results. [3] This modality is effective only on surface lesions that are less than 3-4 mm in size. [8]

Management options for choroidal hemangioma that might lead to loss of vision includes laser, photodynamic therapy and radiotherapy. [11] The first choice of drugs used are beta blockers followed by adrenergic drops or carbonic anhydrase inhibitor drops. [19]

The present proporsal for management of seizures is use of anticonvulsants and aspirin as maintenance doses. [19, 22] Infants with seizure attacks should be maintained on anticonvulsants, the choice of anticonvulsant is uncertain. [22] Aspirin (3-5 mg/kg/day) may be used to maintain venous stasis by preventing thrombus formation. [19, 22] Localized focal cortical resections, corpus callosotomy, hemispherectomy, and vagal nerve stimulation are surgical modalities that have been tried on patients with persistent seizures. [19, 22]

CONCLUSION

Encephalotrigeminal Angiomatosis or Struge-Weber syndrome is an uncommon and rare congenital disorder. Every arteriovenous malformation should be assessed for SWS using advanced imaging procedures to establish a diagnosis. The management of this condition is very challenging and it should include a multidisciplinary effort. As dentists, it is very important for us to recognize and understand this condition so as to prevent hemorrhage. Proper counseling to parents of children identified with this syndrome can help to prevent complications and also improve the quality of life of the patient. Dentist should be aware of the hemorrhage that can occur in this condition and take precautions. More focus should be laid on preventive measures.

CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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