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International Journal of Current Research
Vol. 7, Issue, 11, pp.22768-22770, November, 2015

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

ISSN: 0975-833X

CASE REPORT

ENCEPHALOTRIGEMINAL ANGIOMATOSIS: A CASE REPORT AND REVIEW

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ARTICLE INFO

Article History:
Received 12th August, 2015
Received in revised form
18th September, 2015
Accepted 05th October, 2015
Published online 30th November, 2015

Key words:

Epilepsy,
Sturge-Weber Syndrome,
Port-Wine Hemangioma,
Mother's spot,
Phakomatosis.

ABSTRACT

Sturge Weber Syndrome is a sporadic neurocutaneous disease characterized by facial port-wine stain, ocular abnormalities (glaucoma and choroidal hemangioma) and leptomeningeal angioma. Port-wine stain and abnormal findings like soft tissue calcification or bony changes in skull radiographs, mental retardation, ocular involvement and hemiplegia. Oral manifestations of the disease may vary considerably and changes in morphology and histology of gingiva, periodontium and pulp have been reported. However the most common feature is a gingival hemangiomatous lesion usually restricted to ipsilateral maxilla, mandible, floor of mouth, lips, cheeks, palate and tongue.

***Citation*:** Dr. Saloni Avinav, Dr. Anita Das, Dr. Sonali Sharma, 2015. "Encephalotrigeminal angiomatosis: A Case Report and Review" *International Journal of Current Research*, 7, (11), 22768-22770.

INTRODUCTION

Hamartmatous lesions are (from Greek hamartion "bodily defect") benign (noncancerous) tumor like malformation made up of an abnormal mixture of cells and tissues found in areas of the body where growth occurs (Seth Rohit, 2015). Sturge Weber Syndrome is a sporadic neurocutaneous condition which is characterized by facial port-wine stain, ocular abnormalities (glaucoma and choroidal hemangioma) and leptomeningeal angioma (Khambete *et al.,* 2011). An 11 year old boy visited the department of Oral Medicine & Radiology with a chief complaint of malpositioned teeth of upper front jaw region. His past dental history was non-significant. His past medical history reveled that at the patient is suffering from epilepsy since he was 9 month of age and since then patient is on medication (tab phenytoin) for the same. His vital signs were within normal range. On Extra oral examination, it was found that there was facial hemihypertrophy of the right side with an erythematous macule over the same side of the face extending supero-inferiorly from hairline to the line joining the right corner of mouth to the right ear.

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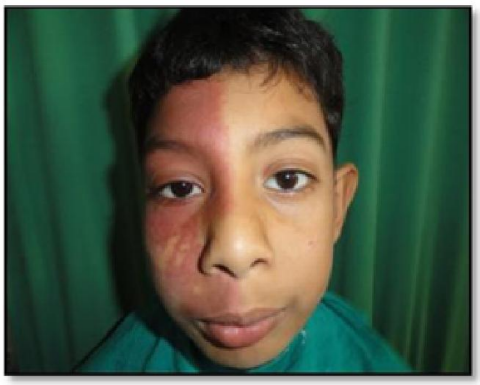
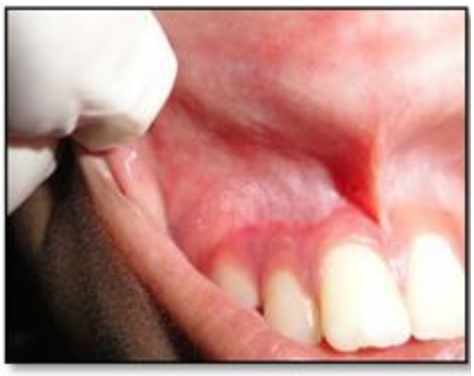
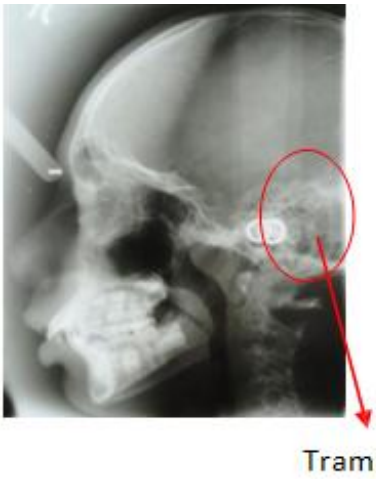
On intraoral examination, right maxillary arch was found to be enlarged as compared to left side and there was generalized gingival inflammation. On the hard tissue examination it was seen that the maxillary anterior teeth were proclined with Angle's class II malocclussion. Suspecting any other illness diascopy was done for the skin lesion. It came out to be positive. Radiographic investigations were performed over the patient. Patient was subjected to lateral Cephalogram. Lateral Cephalometric revealed Tram line calcifications on the skull in posterior parietal lobe. So, correlating past medical history, clinical examination and investigation, diagnosis of Sturge Weber syndrome was made.

DISCUSSION

The incidence of Sturge Weber Syndrome is 1/50,000 live births (http://www.patient.co.uk/doctor/sturge-weber-syndrome #), although it is more often underreported and is found worldwide. The primary defect is developmental, affecting the precursors of tissues that originate in promesencephalic and mesencephalic neural crest that lead to malformations in the CNS, eye, and skin (BIDMC, 2008). Generally, the condition is easily diagnosed at birth or in early infancy based on the external clinical signs alone. However, the development of morbidity from secondary changes and complications occurs throughout life (Monte, 2015).







first trimester of pregnancy (http://rarediseasesnetwork.epi.usf.edu/BVMC/professional/SWS/index.htm). During development at 4-5 weeks gestation, a primordial sinusoidal vascular plexus forms around the cephalic portion of the neural tube and under the ectoderm that later becomes facial skin. This vascular plexus normally regresses at 9 weeks of gestation (BIDMC, 2008) but in Sturge Weber Syndrome the cortical bridging veins fails to form, the vascular plexus persist and remaining veins become engorged with redirected blood flow (BIDMC, 2008) eventually culminates. According to Monte A Del Monte *et al.* (Monte, 2015) it is classified into

1. Complete trisymptomatic

When all three organ i.e. eye, skin, and CNS are involved.

2. Incomplete bisymptomatic

When the involvement is either oculocutaneous or neurocutaneous.

3. Incomplete monosymptomatic

When there is only neural or cutaneous involvement.

Our present case is type 2 (incomplete bisymptomatic). Patients with sturge weber syndrome are born with dermal capillary vascular malformation of the face known as Port wine stain or nevus flammeus. Lesions are generally flat,deep/purple in color. Intraoral involvement in sturge-weber syndrome in common, resulting in hypervascular changes in ipsilateral mucosa. Angiomatosis can involve lips, causing macrochelia, resulting in hemihypertrophy of buccal mucosa, palate and floor of the mouth (http://medind.nic.in/jao/t05/i2/jaot05i2p99.pdf, 2005). Macroglossia and maxillary bone hypertrophy are found in some patients leading to malocclusion and facial asymmetry (Khambete *et al.,* 2011). Gingival enlagement can vary from light vascular that can resemble Pyogenic granulomas to monstrous over growth making closure almost impossible (http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3467913/).

No.	Clinical Manifestation	Incidence (%)	Present Case
1	Epilepsy	80	+
2	Port-Wine Stain	76	+
3	Abnormal Radiographic Findings	63	+
4	Mental Retardation	54	-
5	Oral Manifestations	38	+
6	Hemiparesis	37	+
7	Ocular Manifestations	37	-

The lesions of Sturge Weber Syndrome are regarded as dysplasia of the embryonal vascular system resulting in hemangiomatosis according to Howard Royle (Sathawane *et al*., 2006). The localized abnormalities of blood vessel development and function affecting the facial skin, eye and brain suggest a developmental disruption occurring in the

The characteristic calcification seen in the leptomeninges consists of double-contoured “tram- lines “Trolly – track” or “Tram track (Robert *et al*., 1995; Hedge *et al*., 2007). These are seen best in the lateral skull view, with the affected side closest to the film (Robert *et al*., 1995). Basilar or Anteroposterior view may show the distribution from

superficial to deeper parts of the leptomeninges. The malformations consist of simple vascular structures situated along the space between the piamater and the arachnoid membrane (http://radiolopolis.com/radiology-cases/radiology-images-radiology-teaching-files/neuroradiology/sturge-weber-syndrome-3488.html). Gyriform cortical calcifications on CT and plain film due to altered vessel wall permeability leakage of calcium phosphate or carbonate with subsequent secondary crystallization within perivascular parenchyma (Audrey S. Wang, 2008). Other radiographic investigation done is

- Angiography
- CT scan
- MRI
- MRI with godolinium
- Single photon emission computed tomography (SPECT)
- Positron emission tomography (PET) (Hedge *et al.,* 2007)

Treatment

Sturge-Weber syndrome is incurable. People do not die from it, it is not fatal. Laser treatment may be used to remove or lighten the birthmark. Anticonvulsants can be prescribedrtoma for seizures. In patients with glaucoma, an annual eye-check is advised with prescription of eye drops. In serious cases surgical intervention may be required. For muscle weakness physiotherapy may be indicated to help strengthen the muscles. For developmental delays, the child may require individualized or specialized tuitions (http://www.medicalnews today.com/articles/220430.php). The dental rehabilitation of patients with Sturge–Weber disease is a complex process requiring initial conservative management and later surgery. Poor oral hygiene leading to secondary inflammatory gingival enlargement is often encountered. Patient education and implementation of preventive procedures should be done. A thorough plaque control regimen can go a long way in avoiding gingivectomy which is risky in these pat ients. Hancock *et al* achieved regression of gingival enlargement by vigorous plaque control measures alone (Manivannan *et al*., 2012).

Conclusion

Encephalotrigeminal angiomatosis i.e. Sturge-Weber syndrome whose exact etiopathogenesis is not yet known and it is inferred that the deformity develops during early gestation and display multidisciplinary clinical manifestation mainly involving brain, eye, skin and oral cavity.

Early and prompt diagnosis in dental office is critical since it allows the control of future complication. Proper and advance radiographic technique like imaging helps dentist in diagnosis due to the presence of characteristics calcification i.e. tramline in lateral skull view.

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