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Alarming Skin Tatoo with Periodontal Link: Sturge Weber Syndrome

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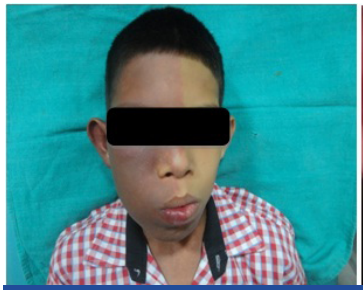
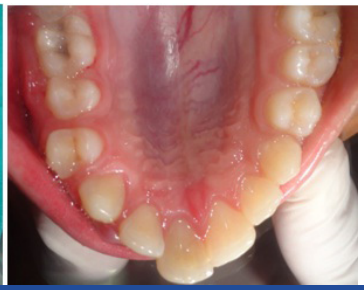
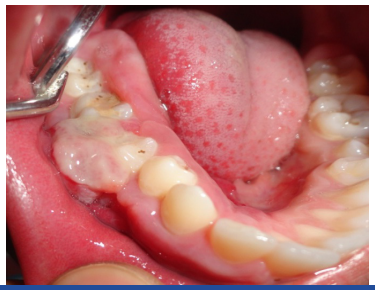
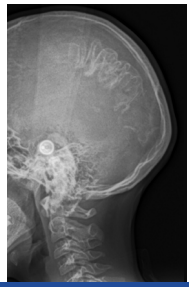
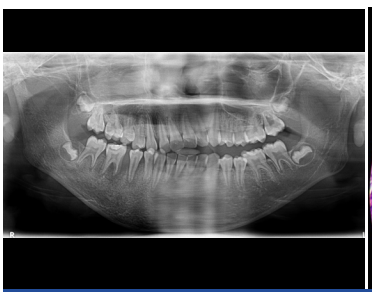
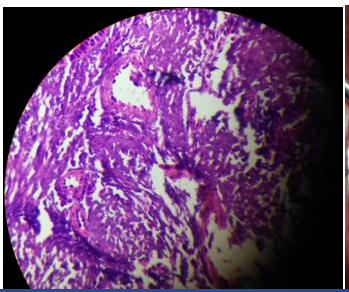
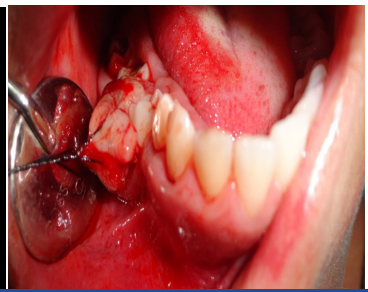
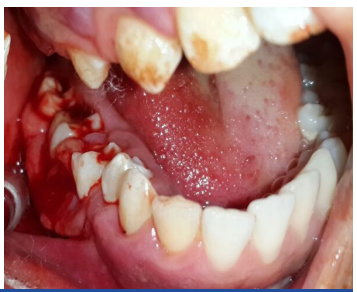
A 12-year-old child with unilateral reddish cutaneous capillary malformation on the face reported to the Department of Pedodontics and Preventive Dentistry at Seema Dental College and Hospital, Rishikesh complaining of swelling in mandibular right back tooth region for the last 15 days which was associated with gradual increase in the size. Patient's past medical history revealed seizures since five months of age and was on long term antiepileptic treatment with multiple antiepileptic drugs (Carbmazepine, Clonazepam). He was born full term and delivered normally. His family history was non contributory. On extra oral examination, Port-wine stain was observed on the right side of the face involving ear and lip on the affected side and crossing the midline in the lower jaw [Table/Fig-1]. Port-wine stains were noticeable on the affected side of the palate also [Table/Fig-2]. His mouth was deviated towards the right side of the face with unilateral open bite on the left side. Intra oral examination revealed firm, non-tender gingival enlargement involving right upper and lower quadrant of the oral cavity (in relation to second premolar and first permanent molar of the affected side) which on probing, bleeded profusely [Table/Fig-3]. Supra gingival calculus and plaque was noticed in both of the arches. Tooth mobility was present in relation to second premolar and first molar on the affected side. Patient was asked to undergo investigations including OrthoPantomoGraph (OPG), complete hemogram, lateral cephalometric radiograph and histopathological investigations. The complete hemogram showed normal hemoglobin levels with no other abnormality in the total or differential white blood cell counts. Bleeding and clotting time were within the normal limits. Anteroposterior view of skull revealed tram track calcification caused by calcification in apposing gyri [Table/Fig-4]. Orthopantomogram revealed bone loss in relation to 46 which was more prevalent in relation to mesial root [Table/Fig-5]. Histopathological investigations suggested "Lobulated Capillary Haemangioma" [Table/Fig-6].

Management: The complete treatment procedures were explained to the patient, a written informed consent was obtained from the patient, and a medical consent was obtained from the general physician.

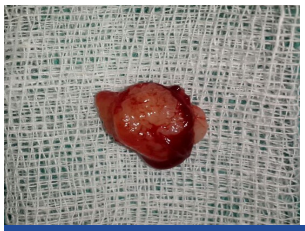
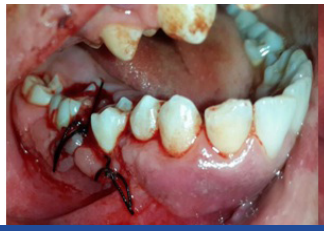
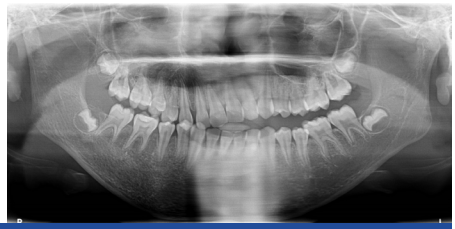
First line treatment of the patient included thorough scaling and root planing to remove any local irritating factors that may have been responsible for the superimposed gingival inflammation. Patient was educated and motivated for maintaining proper oral hygiene. Under strict aseptic conditions using local anaesthesia (Lignocaine and adrenaline 1:80,000), inferior alveolar nerve block was given and a 3-0 suture was passed through the swelling to enscribe it and hold it tight for easy excision [Table/Fig-7] and the mass was then completely excised in relation to buccal side of 45, 46 [Table/Fig-8,9] and stored in formalin solution for further histopathological investigations. To control the excessive hemorrhage sutures were given to achieve hemostasis [Table/Fig-10,11] and post operative instructions were given to the patient. Patient was recalled after one week and sutures were removed followed by an uneventful healing.

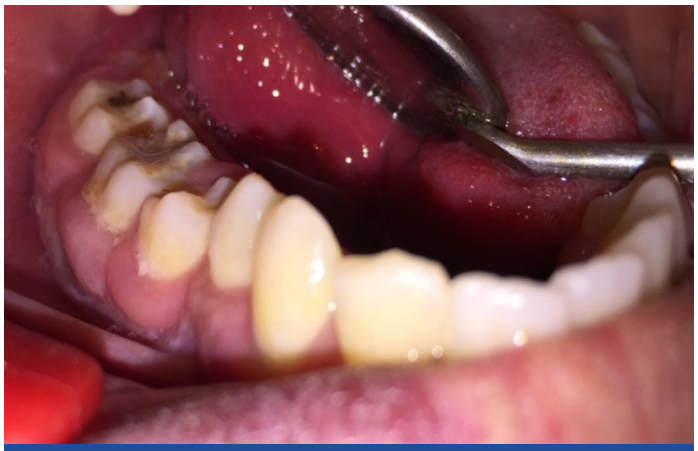
[Table/Fig-1]: Clinical picture showing port wine stain involving the right side of the face. **[Table/Fig-2]:** Port wine stain present on the palate of the affected side.
[Table/Fig-3]: Clinical picture demonstrating swollen gingival. **[Table/Fig-4]:** Lateral cepahlogram depicting tram track lines.

[Table/Fig-5]: Preoperative OPG revealing bone loss i.r.t. 46 **[Table/Fig-6]:** Histopathological slide exhibiting lobulated capillary haemangioma. 40X magnification.
[Table/Fig-7]: Enscribing the swelling using 3-0 suture. **[Table/Fig-8]:** Removal of gingival swelling.

[Table/Fig-9]: Picture showing excised mass. **[Table/Fig-10]:** Suturing done at the site of excision. **[Table/Fig-11]:** Clinical picture depicting hemostasis using 3-0 suture. **[Table/Fig-12]:** Post operative OPG revealing healthy bone formation in relation to 46 after six months follow-up.



[Table/Fig-13]: Follow up after six months.

Post operatively patient was kept under antibiotic coverage and was guided to use 0.2% chlorhexidine mouthrinse for two weeks. After a follow up of uneventful six months, it was seen that there was deposition of normal healthy bone in relation to 46 using OPG [Table/Fig-12] with no recurrence after six months of follow up [Table/Fig-13].

DISCUSSION

Sturge Weber Syndrome occurs with a frequency of approximately 1 per 50,000 [1]. The British physician William A. Sturge was the first person to describe this condition and the other physician Frederick Parkes Weber demonstrated its intracranial calcifications [2]. The Roach Scale is used for classification which is as follows: Type I-Presents with facial and leptomeningeal angiomas. Ocular involvement is normally noted within the first year of life. Type 2-involves a facial angioma and the possibility of glaucoma, but no evidence of intracranial disease. Type 3-It is commonly noted to have a leptomeningeal angioma, with no facial involvement and usually no development of glaucoma. Hence, according to the classification the present case refers to Type 1 of Roach Scale. In Sturge Weber syndrome angiomas are formed due to the failure of regression of vascular plexus around cephalic portion of neural tube which is destined to become facial skin [3]. In our case, the patient showed nevus flammeus on the right side of the face extending over the midline.

In 38% of the cases, oral manifestations are reported which involves hemangiomatous lesion in the lip, oral mucosa, gum, tongue and palatine region. The oral manifestations are generally unilateral finishing abruptly in the middle line [4]. The gingival overgrowth can be drug induced because the affected individuals usually have epileptic seizures and frequently are taking antiepileptic medications [5]. In the present case, it was also noted that one of the drug used for treating the patient was carbamezepine, which could be the reason for gingival overgrowth and further the poor oral hygiene maintenance due to mildly challenged cognitive functioning would have intensified the inflammation and gingival hyperplasia.

CONCLUSION

Although a rare disease, SWS is important to be recognized and treated properly by the dental professionals because of the increased risk of hemorrhage. To reduce the risk of hemorrhage, consultation with a physician, periodic systemic evaluation and meticulous oral examination are critical.

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