NEVUS FLAMMEUS WITH NEVUS OF OTA
- A CASE REPORT

ABSTRACT:
Pigmented areas of the face and neck region are complex presentations of clinical dilemmas. They manifest in various forms and in unusual location and hence pose great challenges to the clinician during management. Presented herewith is a case report of a 6 year old boy, showing signs of Nevus Flammeus & Nevus of Ota. Management of such a patient involves challenges in view of control of hemorrhage. The child reported with trauma to the chin after 3 days and was diagnosed to have intrusive luxation of tooth 41. The tooth was kept under periodic observation and follow-up to monitor the eruption of tooth. It is important for dental surgeons to identify such cases so that appropriate referral to the physician can be made to avoid future complications and to provide comprehensive health care to the child.
key words: Nevus of Ota, Nevus Flammeus, Portwine stains, Management

INTRODUCTION:
Portwine stains represent hamartomatous capillary malformations and are named so due to the deep red hue that they leave on the skin or mucosa. Their presence can signify a developmental anomaly involving the central neural axis. They are present at birth and are most commonly seen on face. Early stains are flat and pink in appearance, later colour may deepen to dark red. Nevus of Ota, which was originally described by Ota and Tanino in 1939, is a hamartoma of dermal melanocytes. Clinically, nevus of Ota presents as a blue or gray patch on the face, which is congenital or acquired and is within the distribution of the ophthalnic and maxillary branches of the trigeminal nerve. The nevus can be unilateral or bilateral and, in addition to skin, it may involve the ocular and oral mucosal surfaces. A large variation has been reported in pattern of occurrence of these facial nevi. The etiology and pathogenesis of nevi of Ota are unknown. Although unconfirmed it is assumed that they represent melanocytes that have not migrated completely from the neural crest to the epidermis during the embryonic stage. The variable prevalence among different populations suggests genetic influences, although familial cases of nevus of Ota are exceedingly rare. Nevi of Ota occur frequently in Asian population with an estimated prevalence of 0.2 – 0.6%. The
male to female ratio is 1:4.8. Various cutaneous and leptomeningeal conditions are associated with nevus of Ota viz. nevus of Ito, nevus flammeus (portwine stains), Sturge-Weber syndrome, neurofibromatosis and leptomeningeal melanosis.

Recognition of the above said conditions in the dental set up is important not only to prevent complications but also for proper referral to concerned specialist due to multiple organ involvement.

Presented herewith is a case report of a patient with portwine stains and ocular manifestation of nevus of Ota.

CASE REPORT:

A six year old boy was referred to the department of Pediatric Dentistry, for the management of mobile teeth following trauma. History revealed, that the patient had trauma to the face 3 days back while playing for which he had reported to a general hospital. There was no history of unconsciousness, vomiting or epistaxis. There was history of bleeding from gums. In view of the trauma, sutures were placed on the chin and labial mucosa after which he was referred to us. Past medical history revealed that the patient had been posted for tonsillectomy which was cancelled due to risk of prolonged bleeding. Family history was non contributory.
Extra oral examination revealed that the patient had a dressing on the chin (Fig-2). It was distinctly observed that the patient had bilateral grayish pigmentation of the conjunctiva with irregular boundaries (Fig-3). Intra oral examination showed a single suture in relation to 82 (Fig-4) in the labial vestibule. On clinical examination it was observed that 72 and 82 had grade I mobility, 31 had grade II mobility and 41 was intruded. It was also observed that 55, 75, 84 and 85 had dental caries (Fig-5). General examination showed that there were well defined erythematous macules and patches on right side of the chest (Fig-6), right arm and hand involving the palm (Fig-7). Similar lesions were observed on the right foot extending to the dorsum of the foot (Fig-8). Macules showed blanching on pressure. It was decided to carry out radiographic examination of the patient in view of trauma. X-ray skull (Fig-9) was taken at the medical hospital which did not show any evidence of fracture. OPG and IOPA of the region showed intrusion of 41 (Fig-10). There was no other evidence of alveolar or jaw fracture. As the patient did not require any active intervention for trauma it was decided to keep the patient under observation for the same. However, in view of the medical condition the patient was referred to a pediatrician and an ophthalmologist. All the investigations had been carried out, blood profile was normal. 3-D CT scan of face and head showed minimal stranding in the sub cutaneous tissue on the left side, no other abnormality was detected on the scan (Fig-11). Based on the clinical findings and other investigations the patient was diagnosed to have Portwine stains with nevus of Ota. Consent was taken to carry out dental restorations. Patient was taken up for dental restorations in 55,75,84,85, (Fig-12, 13) and had been kept under observation by periodic follow up to monitor eruption of 41 (Fig-14).

**DISCUSSION:**

Nevus flammeus is a vascular birthmark made of enlarged capillaries in the skin, which produces a reddish-purple discolouration of skin5. Portwine stains may be one of a group of symptoms and signs perhaps of Struge-Weber syndrome, Klippel-Trenaunay syndrome6,7. The management may be complicated due to risk of bleeding. Nevus of Ota can cause facial disfigurement, resulting in emotional and psychological stress. In rare cases, melanoma which can be life threatening, has been reported to arise from nevus of Ota. Glaucoma also has been occasionally associated with the lesion8. Routine ophthalmologic follow up is necessary. Treatment of portwine stains includes cryotherapy, surgery and radiation. LASER’s have however made a big impact on treatment because they are excellent for destroying the tiny vessels9. The flash lamp pumped dye laser, yellow laser light, is successful in infants and children.

For nevus of Ota laser surgery, cryotherapy, microsurgery, dermabrasion, sequential dry ice epidermal peeling have been advocated9,10. Failure to recognize the potential for malignant transformation11 of nevi of Ota may result in medico-legal complications. From the dental point of view, emphasis should be made on preventive measures to avoid complex treatment.

**REFERENCES:**