Case Report

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Pyogenic Granuloma, an Oral Manifestation of Sturge-Weber Syndrome: A Case Report

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ABSTRACT

Background: A 45 years old female reported to the department of periodontology, with a solitary, oval, pedunculated hemangioma (?) on the gingiva, as the chief complain. The outgrowth was reddish in color, with ulcerative surface and was bleeding profusely on probing. Panoramic radiography, ultrasonography and haematological parameters such as CBC and PT were done. Laser diode (LD) excision was the surgeon's choice. Post-operative histopathological report of the outgrowth revealed that the connective tissue was interspersed with numerous small and large budding capillaries (angiogenic activity). The blood vessels were distributed in a clustered pattern, as an identified feature of a typical granuloma. So the case was confirmed as the oral lobular pyogenic granuloma (LPG). LPG was co-related with the past medical history, findings of present physical examination and clinical reports. The Port-Wine Stain birthmark of right face, earlier epileptic episodes and funduscopic findings were gone in favor of Sturge-Weber Syndrome (SWS). Thus the LPG was finally diagnosed as the intraoral manifestation of SWS. SWS is a rare congenital non-fatal neurocutanious disorder caused by somatic mosaic mutation.

Keywords: Pyogenic granuloma, Sturge-Weber Syndrome, The Port-Wine Stain.

INTRODUCTION

The pyogenic granuloma (PG) is often noticed as a small benign vascular tumor of the skin or mucosal membrane, may be in response to localized irritation or trauma. It has been proved that, nothing like pyogenic or pus forming, nor the tumor is granulomatous hence the terminology PG is a misnomer, but it is popularly accepted as the pyogenic granuloma across the world. [1]. PG may be an inflammatory hyperplasia consequence of the traumatic injury, slow persistent local irritation or may be the angiogenic outcome of estrogenic effect. The tumor is characterized with rapid proliferating vascular networks and fibroblastic connective tissue [2]. Pyogenic granuloma also synonyms with lobular capillary hemangioma, Crocker and Hartzell's disease, granuloma pyogenicum, granuloma pediculatum benignum, benign vascular tumor and granuloma gravidarum [3,4]. It mostly appears as a painless, pedunculated or sessile erythematic mass of gingival tissue. PG is described in two histological forms viz. lobular capillary hemangioma (LCH) and the non-lobular capillary hemangioma (non-LCH)[4]. Etiology of the PG is not yet fully understood and remains an etiopathological enigma. Several reports describe the PG as an extra oral cutaneous or mucosal layer hemangioma of the proximal aero-digestive tracts [2,3]. PG at other places of human body have been rarely reported [5]. Sometimes oral pyogenic granuloma have been co-related with pre existing Strurge–Weber Syndrome (SWS) and diagnosed as the oral manifestation of the same [7,8]. Sturge-Weber Syndrome is a rare congenital disorder, it involves cutaneous, CNS and optalamic systems.

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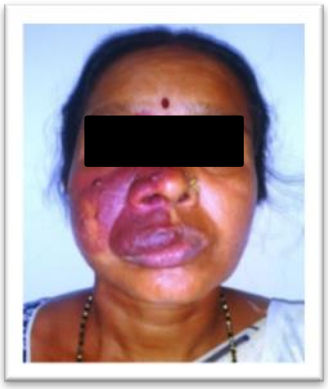

Fig 1: The subject with well-defined Port Wine Satin (PWS) birthmark extending up to the vermilion border of upper lip (arrow).

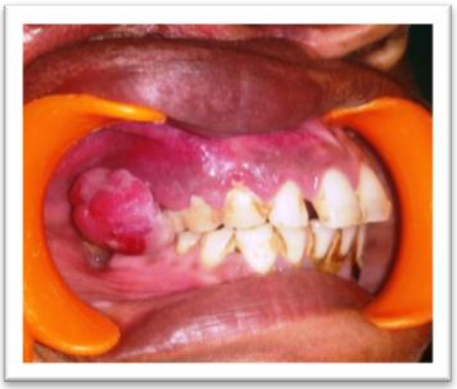

Fig 2 The Lobular capillary haemangioma of maxillary gingiva in relation to # 16; Note the irregular border of the haemangioma (arrow).

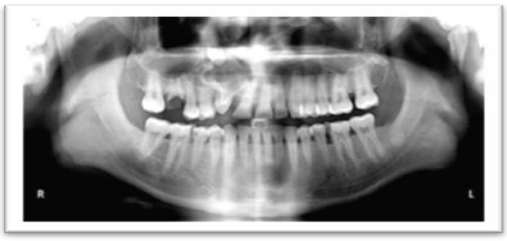

Fig 3: The panoramic radiograph (Orthopantogram) showing traumatic tooth root stump (arrow)

The factor (s) that induces SWS is not fully understood but it is due to a developmental disorder during first few weeks of embryogenesis. Normally the vascular plexus that develops around the cephalic region of neural tube regress gradually by ninth week of intrauterine life. Persistent vascular plexus may lead to neurocutanious-ocular disorder [9]. SWS has several other synonyms like

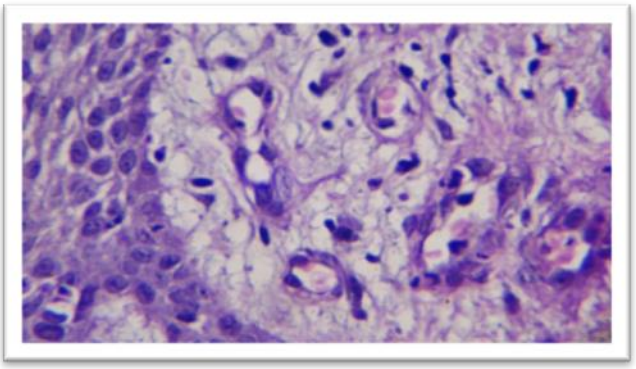

Fig 4: Histopathology.

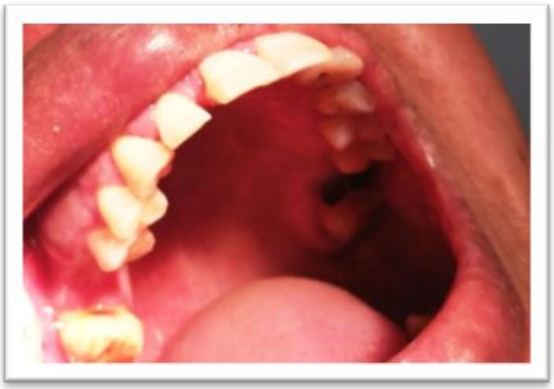

Fig 5: Post operative follow up after 8 month. Note the completely healed up maxillary gingiva at pyogenic granuloma site (arrow).

dimitri disease, encephalofacial angiomatosis, encephalotrigeminal angiomatosis, leptomeningeal angiomatosis. Sturge-Kalischer-Weber syndrome, Sturge-Weber-Krabbe syndrome and Sturge-Weber phakomatosis. Based on the systemic involvement several authors classified SWS as trisymptomatic (skin, brain and eye), bisymptomatic (any two of the systems, may be oculo-cutaneous or neuro-cutaneous) and monosymptomatic (any one of the system) [10]. In this classification the former one is treated as complete SWS and latter two are considered as incomplete. However several case studies have reported the common symptoms of SWS as exogenous hemangioma (cutaneous capillary malformation). Port Wine Satin (PWS) birthmark or nevus flammeus, glaucoma and periodical seizures are also attributed as some important symptom of SWS [11,12]. The present report is based on a case study, where a subject came to the Hi-Tech Dental college and Hospital, Bhubaneswar with chief complain of growth in her maxillary right posterior tooth region since last 8

months. The case was finally diagnosed and treated as Strurge–Weber Syndrome (SWS), associated with angiomatosis as oral manifestation.

CASE REPORT

A 45 year old female patient reported to us with a chief complain of growth in her maxillary right posterior tooth region since 8 month. She gave a history of traumatic extraction in that particular region, after which the growth started. Extra oral physical examination could show the facial asymmetry with swelled up right cheek and upper lip. There was a prominent unilateral Port Wine Satin (PWS) on the right side of face extending to the vermilion border of upper lip [Fig. 1]. Intraoral physical examination could locate a solitary, oval, pedunculated , well defined swelling in the gingiva in relation to # 16, covering the crowns of 15, 16,17 and measured about 14 x 15 mm in size. It was shiny reddish in colour with ulcerative irregular surface[Fig. 2]. The above unilateral erythematous gingiva on the maxillary site was partially covered by white slough. On palpation, the swelling was found soft but non-tender. There was profuse bleeding while probing the structure. Oral hygiene was poor with extrinsic stain on the tooth surface with calculus.

Among other important case histories, the patient expressed decreased vision on right eye since last 2 months. She also revealed about her recurrent seizures of childhood, but that was reduced with increase of age. The patient was subjected to some clinical tests such as complete blood count (CBC), prothrombin time (PT) retinography and ophthalmoscopy, Orthopantomogram (OPG), ultrasonography (US) of right side of face with doppler. Findings of all the tests are summarized (Table 1). Clinical findings and the past medical history prompted the surgeon for excision of the PG and its maintenance. The post excision tumor was passed through the routine histopathological screening and the case was finally diagnosed as the intraoral pyogenic granuloma with Sturge-Weber Syndrome.

Surgical intervention and treatment procedure.

The following treatment plan was chosen to address the issues. Supra gingival and subgingival scaling was done. Oral hygiene prophylactic and antibiotic coverage was given before excision.

The soft tissue overgrowth was excised with the help of a laser diode, (LD). The LD had the advantage of minimal incision, minimal bleeding, limiting mechanical trauma, effective removal of affected tissue and reduced post-operative pain. Operation was conducted under local anesthesia. Extraction of offending root stump # 16 was instantly done. Antibacterial coverage and oral antiseptic mouthrinse was recommended for easy healing. Scaling and root planning was also done to maintain the oral hygine. The patient is under maintenance therapy with regular intermittent follow up over one year.

Table 1 Clinical investigation result of the subject

Sl. No	Name of the investigation	Result	Remark if any
1	Complete blood count (CBC)	All the parameters are in normal range except Hb (11%)	This may be due to poor health status
2	Prothrombin time (PT)	Normal 11 Sec.	Range 10-14 Sec
3	Retinography and funduscopy	Redness of conjectiva and dilated bv (left) and hemianopia (right)	It support the decrease visual complaint
4	Panoramic Radiograph (Orthopantogram)	Presence of root stumps # 16, with no bone loss	Traumatic condition
5	Ultrasonography (doppler)	Soft tissue dense mass lesion of size 3 x 1 cm over right maxilla	Impression of PG
6.	Rutine histopatholgical (H&E) test of PG(?)	Parakertanized stratified squamous epithelium and fibrovascular connective tissue. budding capillaries in clustered pattern	Impresion of angiogenic (?) and haemangioma (?)

DISCUSSION

SWS is a rare congenital neurocutanious disorder. It is often recognized by the appearance of and Port-Wine Stain (hemangioma) on the dermal sites, epileptic episodes and glaucoma. In a recent review Anne M.Comi [13] described the Sturge–Weber syndrome as the third most important rare

neurocutaneous disorder appears sporadically 1 in 20 000 live births. He opined that an activating mutation in GNAQ causes somatic mosaic mutation which is expressed as both SWS and isolated Port-Wine Stain (PWS) birthmarks. He also added that a new born with PWS birthmarks may have 15-50% chances to develop SWS. The present patient with some of the above mentioned characteristics is identified as a subject of SWS. Such rare incidences have been reviewed and reported many other workers [10,12,15]. SWS is classified as complete if there is CNS, cutaneous and ophthalmic involvement and incomplete with absence of one or more characters [7,8,11]. Pyogenic granuloma in oral cavity has been reported as an oral manifestation of SWS patient of all ages and both the sexes. Generally, the granuloma is ipsilateral to PWS birthmarks in the anterior maxillary position. The present study corroborates the locational position of the PG as above. There are reports of mandibular involvement [16], multiple intra oral occurrence [17], and extra oral location of PG [6,18] in isolated cases. The patient in her case history reported the traumatic tooth extraction quite before the appearance of the PG. The OPG also evident the retention of root stump of the same. Akheel and Tomar [19] in their review attributed that one third of the PG ever reported, occurred after the traumatic injury. Hence the present case history of trauma for this lesion might have contributed factors like of persistent irritation and local mild infection. The epidemiology od SWS and associated PG shows the female dominance. This may be due to the vascular endothelial growth factor activity of estrogen. Moreover the immunohistochemical studies on estrogen and progesterone receptor expression in gingival lesions have strongly supported the angiogenic activity of the estrogen. [20, 21]. Histopathology section of the post excised PG of the present patient revealed that the fibro-vascular connective tissue. Epithelium was hyperplastic, characterized by elongated rete ridges. Connective tissue also shows the collagen fibers network interpreted with numerous small and large budding capillaries (angiogenic activity). The engorged RBC, were noticed in the epithelial cells. Blood vessels which were showing clustered patterned distribution were satisfying the haemangioma condition. However, all these histological findings agreed with the finding of earlier workers those who described the typical pyogenic granuloma [1,5,22]. The ocular examination findings of the present subjects is recorded in term of redness of conjunctiva with dilated blood vessel (left eye) and hemianopia (right eye). This findings were almost similar as reported by Neerupakan *et al.* [12]. Therefore the possibility of ocular insolvent in the present study cannot be rule out. Laser diode, (LD) excision was the surgeon choice. Scaling of teeth, postoperative antibiotic coverage and oral prophylactic could maintain and manage the patient over the period of one year. Although there are report of recurrent granuloma as satellite oral lesion attributing fluty surgical management[23], the present patient has not developed any complicacy after the surgical intervention.

CONCLUSION

SWS is a rare congenital non- fatal neurocutanious and ocular disorder. Pyogenic granuloma is a non-neoplastic growth which may be associated with SWS as an intraoral manifestation. It needs a surgical intervention, treatment and routine follow up for sustainable maintenance.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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