

Sturge-Weber syndrome with pyogenic granuloma

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Abstract

Vascular lesions represent one of the rare disorders affecting overall quality of life of a child. A wide variety of these conditions are known, ranging from a simple nevus to life-threatening hemangiomas. These conditions make the treatment options more complex due to the fear of uncontrollable bleeding. The present case is one of the rare combinations of Sturge-Weber syndrome and pyogenic granuloma. Conditions of importance and treatment options keeping hemangioma in mind are discussed.

Keywords: Hemangioma, portwine stain, pyogenic granuloma, Sturge-Weber syndrome

Introduction

Congenital deformities represent major psychologically disturbing problems for both parents and children. Encephalotrigeminal angiomatosis, or Sturge-Weber Syndrome (SWS), is an uncommon nonhereditary developmental condition with hemangioma as a disturbing feature.^[1] The pathognomonic features of this syndrome are venous angiomas of the leptomeninges extending over the cerebral cortex with ipsilateral angiomatous lesions, unilateral facial nevus along the distribution of one or more divisions of the trigeminal nerve extending up to the midline and epileptic convulsions. Commonly the clinical features are convulsions, hemangiomas, abnormal radiographic findings, ocular involvement, and hemiplegia.^[2] On the basis of systems involved SWS is classified as complete trisymptomatic when all three organ systems (eye, skin, and CNS) are involved, incomplete bisymptomatic when the involvement is either oculo-cutaneous or neuro-cutaneous and incomplete monosymptomatic when there is only neural or cutaneous involvement.^[3]

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Presenting themselves as one of the benign conditions their entire course runs from 1 month to 10 years after which they usually regress. Hemangiomas appear during first 2-3 weeks of infancy and are most commonly seen in females than males (3:1 to 5:1). The incidence in Caucasian infants ranges from 10 to 12% and lower in dark skinned infants. They may be solitary or present in many locations measuring 0.5 cm to an extensive area. The life cycle of majority of hemangiomas is characterized by rapid proliferative phase seen during first 2 years followed by a ceased growth lasting for 7-8 years followed by the phase of involution over next few years.^[4]

In the present case Hemangiomatous lesion divided the face in the two halves from midline with prominent portwine stains and was associated with pyogenic granulomatous lesions. The clinical findings and management are discussed.

Case Report

A 12-year-old girl reported to the department of Pedodontics RDC Loni with a complaint of swelling in the lower front gum region since 3 months. Clinically there was an isolated gingival swelling in the lower left anterior region between central and lateral incisors (31 and 32). The swelling was red and shiny in appearance measuring about 2 × 1 cm in size extending from mesial surface of central incisor to the mesial half of canine on the labial side and was extending on the lingual side of incisors measuring 0.8 × 0.5 cm in size. Both the swellings were connected at the interdental area of central and lateral incisors [Figures 1a and b]. On palpation the swelling was a pedunculated mass, soft and easily movable from the base. On probing gently, the lesion showed considerable amount of bleeding. Extraorally the face was asymmetrical with slight enlargement on the right side of the face. Remarkably the enlarged right half of the face was bluish red in color. There was a prominent scar on the right side of the face in upper buccal region [Figure 1c]. Parents revealed a history of surgery 7 years ago for a red swelling in the scarred area.

Intraorally right half of the mouth was markedly red affecting labial and buccal mucosa, buccal vestibule, floor of

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Figure 1a: Localized gingival enlargement between 31 and 32



Figure 1b: Labial and lingual swelling connected at the interdental area of 31 and 32



Figure 1c: Enlarged right half of the face reddish in color with arrow showing scarred area

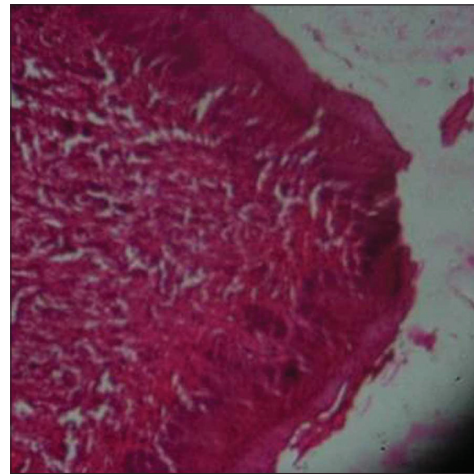


Figure 2: Histopathology showing multiple endothelium lined spaces and connective tissue fibers suggestive of pyogenic granuloma



Figure 3: Six months follow up showing satisfactory healing

mouth, tongue and palate while left side did not show any abnormalities. Intraoral periapical radiographs of the 31, 32, and 33 region and OPG were advised to rule out any bony involvement. The radiographs did not reveal any significant

problems except presence of initial bone loss from the interdental area of 31 and 32.

Since the lesion had a pedunculated base with little bony involvement excisional biopsy was carried out carefully followed by bony curettage. The entire procedure was performed under local anesthesia. Histopathological examination of the excised mass showed numerous endothelium lined spaces, abundant fibroblasts, collagen fibers, and polymorphonuclear leucocytes revealing features of pyogenic granuloma [Figure 2]. At the end of 1 month, the lesion had uneventful healing [Figure 3]. Six-month follow-up was quite satisfactory and there was no evidence of any recurrence. During surgery and curettage the amount of bleeding was controllable and no additional steps were required to stop the bleeding. This could have been due to the presence of granuloma on the left side which was not affected by the vascular lesion.

Discussion

In the present era of science, anomalies are toward fading side but some anomalies are really disturbing.^[2,3] Although majority of hemangiomas recede over a period of 10-15 years, in few cases they may continue to exist and disturb the esthetics and function.^[3] Since SWS is a disease with hemangiomatosis, such growths on gingival can be expected. This complication of gingival enlargement could be due to increased vascular component and precipitated by local irritation by plaque and calculus.^[4] Regardless of these speculations, the true etiology remains unknown. A significant increase in the numbers of mast cells in pyogenic granuloma with similar vascular lesions has been reported. The average “density” of mast cells (MCs) per mm square appearing in the central region of the pyogenic granuloma is shown to be higher compared to adjacent nevus and normal skin indicating that MCs are closely associated with angiogenesis in pyogenic granuloma.^[5]

The diagnostic tools for such cases include ultrasound a relatively economic non-invasive technique, CT scanning, and MRI.^[6]

Multidisciplinary treatment approach including plastic surgeons, pediatric surgeons, anesthetists, hematologists, etc., is required to manage such cases. Corticosteroids either systemically or intralesionally can be used. Oral Prednisone 1-2 mg per kg for 6 months and Triamcinolone given intralesionally 1 to 2 mg per kg per month for 6 months has documented good relief from the condition. Interferon-alfa 2a, Bleomycin, and cyclophosphatamine are also used.^[7] Use of Nd:YAG laser is commonly followed surgical procedure by many plastic and reconstructive surgeons. The lesions which are superficially situated and smaller in size are best managed by Nd:YAG laser. But the effectiveness of lasers in deeply situated skeletally involving hemangiomas still requires extensive study.^[8]

In the present case, hemangioma with portwine stains present on the right half of the face was an unusual finding. The patient had undergone cosmetic surgery for the same; however, the buccal aspect still showed signs of scarring. Surprisingly the mass representing pyogenic granuloma was seen on left unaffected area of lower jaw making it difficult to limit the diagnosis of hemangioma on other side. Keeping in mind the potential complication in the form of hemorrhage all precautions were taken and emergency team was ready if difficult situation would exist. Eventually the case was managed satisfactorily under local anesthesia with no complications.

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