Encephalotrigeminal Angiomatosis with Facial Lobular Capillary Hemangioma: An Unusual Case Report

Abstract

Encephalotrigeminal angiomatosis, also called Sturge–Weber syndrome (SWS), is a syndrome of etiology which is not yet clear. It is a nonhereditary condition. The clinical features include pigmentation over the facial skin known as port-wine stain, abnormalities of ocular region, and central nervous system involvement as leptomeningeal angioma. In this manuscript, we present a rare case report with an unusual combination of SWS with facial lobular capillary hemangioma.

Keywords: *Calcifications, capillary hemangioma, facial pigmentation, Sturge–Weber syndrome*

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Introduction

Encephalotrigeminal angiomatosis, also called Sturge–Weber syndrome (SWS), was first described in 1860 by Schirmer, and then in 1879 by Sturge who associated changes on skin and ophthalmia to neurological symptoms. Later in 1929, radiologic features were described by Weber.[1] It is rare "affecting 1 in 100,000 South Asian population,"[2] nonhereditary developmental condition characterized by angiomatosis of face, ocular involvement, mental retardation, and seizures.[3] Port-wine stains (nevus flammeus) and strawberry nevi (infantile hemangiomas) are commonly occurring capillary vascular malformations. Port-wine stains remain throughout life, but most infantile hemangiomas regress on their own.[4] Here, we describe the unusual case of co-occurrence of SWS and facial lobular capillary hemangioma.

Case Report

A 12-year-old male patient reported to the department of oral medicine and radiology with a very slow-growing, ulcerated but painless growth on the chin and lower lip area for 6 years. The patient had a history of similar growth on the left ear, which healed itself with scar formation.

Medical history revealed frequent episodes of migraine, but there was no history of epileptic seizures and natal history was not relevant. On general examination, there were neither any signs of developmental delay in motor and speech function nor any evidence of cardiovascular and respiratory disease.

Examination of the extraoral region revealed an asymmetrical face due to the enlarged left side, especially lips, nose, and zygomatic area. Port-wine stains were seen over the whole left-sided face, extending to the right side at the forehead, nose, lip, chin, and cheek region [Figure 1]. A pedunculated, round growth having diameter approximately 2.5 cm was present on the chin area below the lower lip [Figure 1]. This erythematous growth was firm, smooth, noncompressible, nonpulsatile, and nontender on palpation, but blanching was noted on digital pressure. Gingiva, labial, buccal, and palatal mucosa of the left side of the oral cavity was found to have erythematous enlargement. Initial glaucoma signs were noted on ocular examination.

Skull radiography after computed tomography brain revealed abnormal, well-defined, 0.5-cm-sized calcified areas with perilesional areas of cerebrospinal fluid Hu (gliosis) in the right occipital and parieto-occipital parafalcine regions of brain parenchyma, suggestive of calcified lesions with perilesional gliosis [Figure 2].

An excisional biopsy of the growth on the chin was performed. The specimen showed parakeratinized, atrophic, stratified squamous epithelium. Lesional connective tissue was predominantly cellular,

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showing small, uniform vascular channels having lobular architecture which are surrounded by large vessels. The capillaries varied in their appearance from plump, solid nest of endothelial cells without lumina to larger vessels lined with prominent endothelial cells. Endothelial cells showed tufting and mitotic activity. The surrounding connective tissue showed fibroblast, collagen fibers, and areas of hemorrhage. The histopathological diagnosis was lobular capillary hemangioma [Figure 3].

Based on clinical, radiographic, and histopathological findings, encephalotrigeminal angiomatosis with facial capillary lobular hemangioma was given as the final diagnosis.

Discussion

Encephalotrigeminal angiomatosis is a rare disorder without racial predilection but equally affects males and females. The clinical features of the syndrome are "angioma of the leptomeninges epilepsy, port-wine stain, ocular involvement, dermal angiomas, mental retardation, hemiplegia, and abnormalities in skull radiographs."[5]

The Roach Scale[6] is used for the classification of this syndrome depending on the involvement of central nervous system (CNS) and facial angiomas. The classification is as follows:

1. Type I – Facial and leptomeningeal angiomas which may have glaucoma
2. Type II – Only facial angioma, no CNS involvement and may have glaucoma
3. Type III – Leptomeningeal angiomas and no glaucoma.

As seen in our case report, the 12-year-old child had unilateral facial angiomas without epilepsy and mental retardation since birth. Hence, according to the Roach classification, it may be Type II. The port-wine stains were seen over one side of the face, but in 50% of cases, it may extend over the midline of face. About 33% of the cases show bilateral involvement.[7] In the present case, port-wine stains were extended from the left side of the face to the right side. Almost bilateral involvement was noted.

Lobular capillary hemangioma frequently develops within port-wine-stained skin.[8] In the present case, a proliferative vascular lesion was seen over the chin area. The lesion was small measuring approximately 2.5 cm in diameter, and not appeared to be life threatening on presentation. The radiograph did not reveal any kind of abnormality or bony involvement. Hence, simple excision of the small growth under necessary precaution was performed, which was histopathologically examined and finally diagnosed as facial lobular capillary hemangioma.

Lobular capillary hemangioma is also known as pyogenic granuloma. It is a misnomer, as neither it is pus filled nor has granulation tissue. It is a reactive lesion which is proliferating in nature with more vascular component because of which it is classified as a vascular neoplasm.[9]

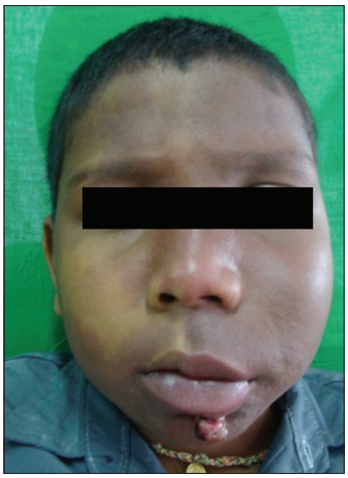

Figure 1: Port-wine stains seen over the left side of the face, extending to the right side at the forehead, nose, lip, chin, and cheek regions and a pedunculated, round growth having diameter approximately 2.5 cm on the chin area below the lower lip

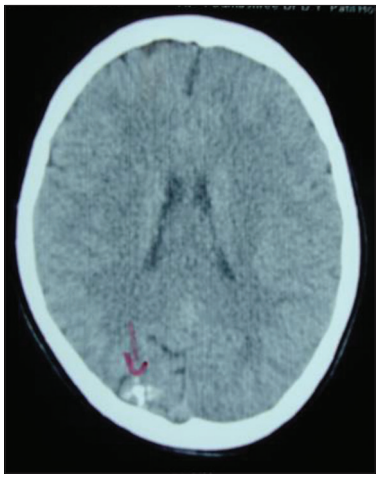

Figure 2: Computed tomography brain revealing abnormal, well-defined 0.5-cm-sized calcified lesions with perilesional gliosis

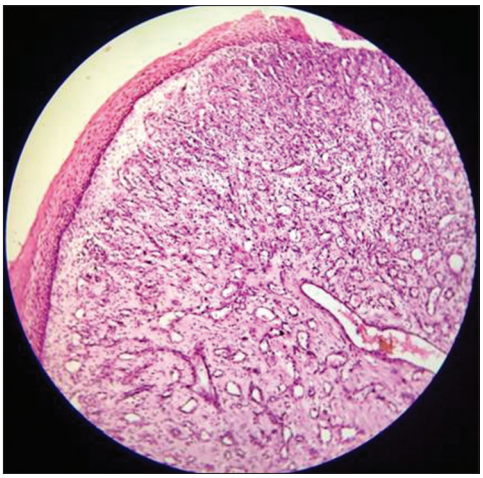

Figure 3: Histopathology showing features of lobular capillary hemangioma

This acquired vascular disorder manifesting in the skin and mucous membranes primarily affects infants and children. However, it is frequently misdiagnosed as infantile hemangioma. The prevalence of this disorder is approximately 12% in infancy, and 42% during the first 5 years of life. The most common locations of lobular capillary hemangiomas are on the head and neck. It rapidly enlarges to a size of around 6.5 mm, usually has a pedunculated base, and is prone to bleeding if there is erosion, which is difficult to be controlled.[10]

Intraoral examination in this case revealed hypervascular changes to the ipsilateral gingiva, which is the characteristic feature observed in SWS due to increased vascular component.

Conclusion

It is a rare vascular disorder and has higher tendency of bleeding because of which it stands apart from the routine cases. The management of such condition is dependent on its manifestation and severity of its clinical presentation, and it requires multidisciplinary approaches for its management. Dentists should be aware of such syndromes and their possible complications so that necessary precautionary measures can be taken during dental procedures to avoid complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that name and initial will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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