

Concurrent Occurrence of Lobular Capillary Haemangioma and Port-Wine Stain: A Case Report and Literature Review

Review began 01/04/2022

Review ended 01/09/2022

Published 05/06/2023

© Copyright 2023

Priya et al. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Sasti Priya¹, Karthik Rajaram Mohan¹, Ravikumar Pethagounder Thangavelu¹, Saramma Mathew Fenn¹, Kumar Appusamy¹

1. Oral Medicine and Radiology, Vinayaka Mission's Sankarachariyar Dental College, Vinayaka Mission's Research Foundation-Deemed University, Salem, IND

Corresponding author: Karthik Rajaram Mohan, drkarthik@vmsdc.edu.in

Abstract

A port-wine stain is a type of non-neoplastic hamartomatous malformation of capillary blood vessels, resulting from ectatic capillaries present from birth. Lobular capillary hemangioma is a form of capillary hemangioma that occurs from hamartomatous malformation of capillaries. In our report, we discuss the rare case of both port-wine stain and capillary haemangioma on the gingiva in a 22-year-old young male.

Categories: Dentistry, Oral Medicine

Keywords: hmme-pdt therapy, port-wine stain, pulsed-dye lasers, hamartoma, capillary haemangioma

Introduction

A port-wine stain is a hamartomatous malformation of the capillary blood vessels. Such lesions are present from birth but become more apparent after the pubertal age [1]. Port-wine stains do not undergo regressive changes, unlike other haemangiomas such as salmon patches, which undergo involution or regressive changes by the formation of fibrous scar or fine telangiectasia on the surface of the skin where they occur. Some persist as nodular growth due to the presence of ectatic blood vessels in them [2]. Dysregulation of vascular mitogen-activated protein kinase (MAPK) and/or phosphoinositide 3-kinase (PI3K) signalling during human embryonic development, as well as somatic mutations of *GNAQ* and *PI3K*, play a role in the pathogenesis and progression of Port Wine Stain [1].

The term "lobular capillary haemangioma" represents a form of hamartomatous malformation affecting the capillary blood vessels. Lobular capillary haemangioma occurs as bright red, raised nodular growth on the gingiva, and has an increased tendency to bleed [3]. Lobular capillary haemangioma occurs concurrently in patients with port-wine stain. It can occur in Sturge-Weber syndrome, in which patients have a history of seizures and a risk of increased intraocular pressure (glaucoma). The cerebral cortex underlying the leptomeningeal (arachnoid and pia mater) vascular malformations usually become dystrophic, non-functional, and calcified producing a tram-track sign in the lateral cephalometric skull radiographs [4]. The concurrent occurrence in a patient of such a lobular capillary haemangioma with port-wine stain is discussed here.

Case Presentation

A 22-year-old male reported with a chief complaint of growth on the gums, present from birth. On eliciting the history of presenting illness, it started as a small growth involving the gingiva and has gradually progressed to the present size. The growth became more prominent over the period of the past seven months. The growth interfered with speech and he had mild discomfort of bleeding, only while brushing his teeth. On extraoral examination, a patch with a pinkish hue was seen on the skin of the face near the chin and below the lower lip on the right side (Figure 1).

How to cite this article

Priya S, Rajaram Mohan K, Pethagounder Thangavelu R, et al. (May 06, 2023) Concurrent Occurrence of Lobular Capillary Haemangioma and Port-Wine Stain: A Case Report and Literature Review. Cureus 15(5): e38642. DOI 10.7759/cureus.38642



FIGURE 1: Extraoral examination revealed a port-wine stain below the right side vermilion border of the lower lip

History revealed he had no episodes of any seizures. General examination revealed his vitals are stable. On intraoral examination, a growth was seen on the lingual aspect of the gingiva in relation to the 26, 27, 28, 29 tooth regions. It measured approximately 1.5 x 2 cm in diameter approximately; the surface showed discrete areas of purplish hue. The right side of the floor of the mouth and the ventral surface of the tongue appeared bright red, not crossing the midline (Figure 2).



FIGURE 2: Intraoral clinical photograph revealed a growth with purplish hue on the gingiva on the lingual aspect in relation to 26, 27, 28, 29 regions and bright, fiery-red area only on the right side ventral surface and floor of the mouth not crossing the midline

On palpation, the growth was firm in consistency, non-tender, sessile, non-pedunculated, compressible, and reducible, showing an increased tendency to bleed. The growth showed a positive modifieddiascopy test when performed with the back of the mouth mirror (Video 1).

VIDEO 1: Modified diascopy test is positive for blanching

View video here: <https://vimeo.com/662387206>

The differential diagnosis for the growth on the gingiva includes fibroma, peripheral ossifying fibroma, pyogenic granuloma, peripheral giant cell granuloma. Fibroma is a universal tumour that can occur anywhere in the body. Usually, fibroma inside the oral cavity appears clinically as a growth showing the same colour as adjacent mucosa. Pyogenic granuloma usually occurs from local irritating factors such as supragingival and subgingival calculus deposits [5]. In our case, local factors such as supragingival calculus are not present, hence pyogenic granuloma is ruled out. Peripheral ossifying fibroma can also occur as a sessile mass involving the gingiva [5]. Since there is a bright reddish area involving only the right lateral half of the floor of the mouth in relation to the vicinity of the growth in the 28, 29 regions, a feeding artery is suspected of supplying the growth on the gingiva. Hence, a contrast-enhanced CT is advised. The arterial phase of contrast-enhanced CT axial, coronal, sagittal sections revealed an enhanced contrast suggestive of a feeding artery supplying the growth on the gingiva in relation to the 27, 28, 29 regions (Figure 3).

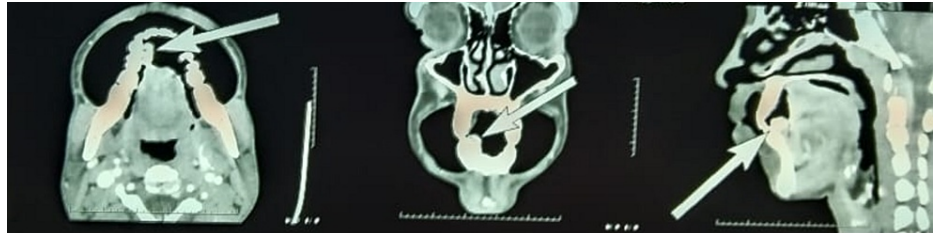


FIGURE 3: Contrast-enhanced CT (axial, coronal, and sagittal section) revealed a feeding artery on the gingiva on the lingual aspect in relation to 27, 28, 29 regions

The growth on the gingiva is excised after presurgical precautions such as careful clamping of the major feeding vessels (lingual artery) supplying the growth on the gingiva. The compression was provided with the help of cotton gauze in the region for 15 minutes. The excised growth was sent for histopathological examination, which revealed numerous capillaries, each lined by a single layer of endothelial cells (Figure 4).

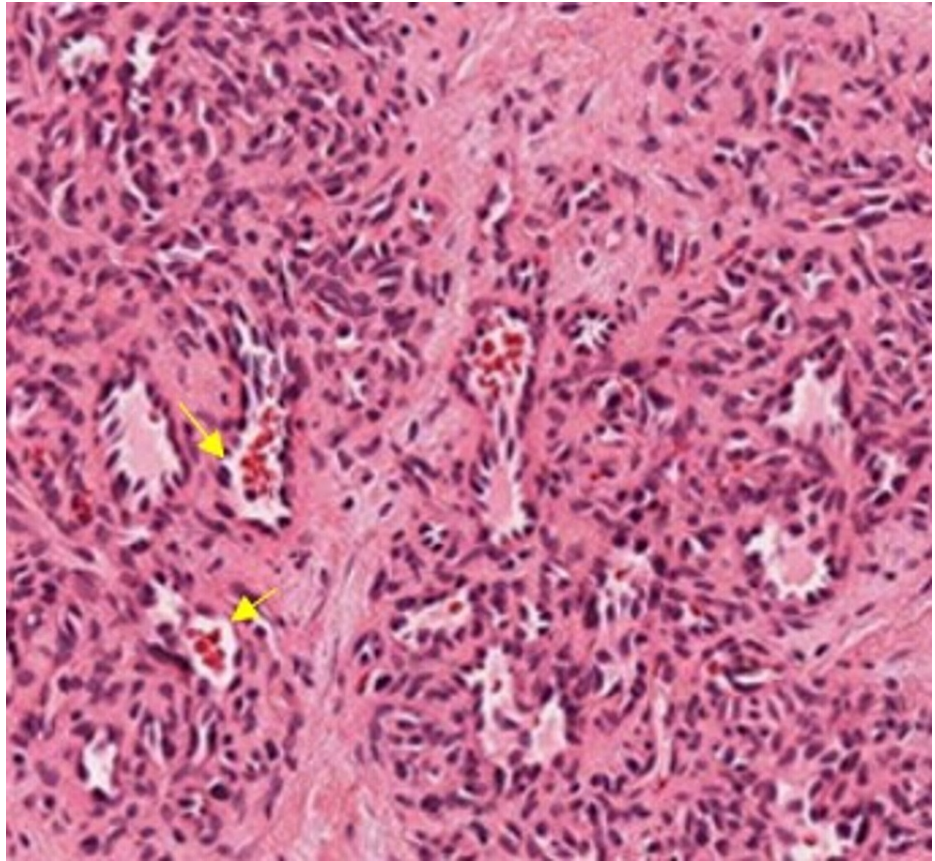


FIGURE 4: Histopathological photomicrograph revealed numerous capillaries, each lined by a single layer of endothelial cells (yellow arrows)

The follow-up intraoral photograph after a month revealed a mild inflammatory swelling on the lingual aspect of marginal gingiva and interdental papilla in relation to 28, 29 region (Figure 5).

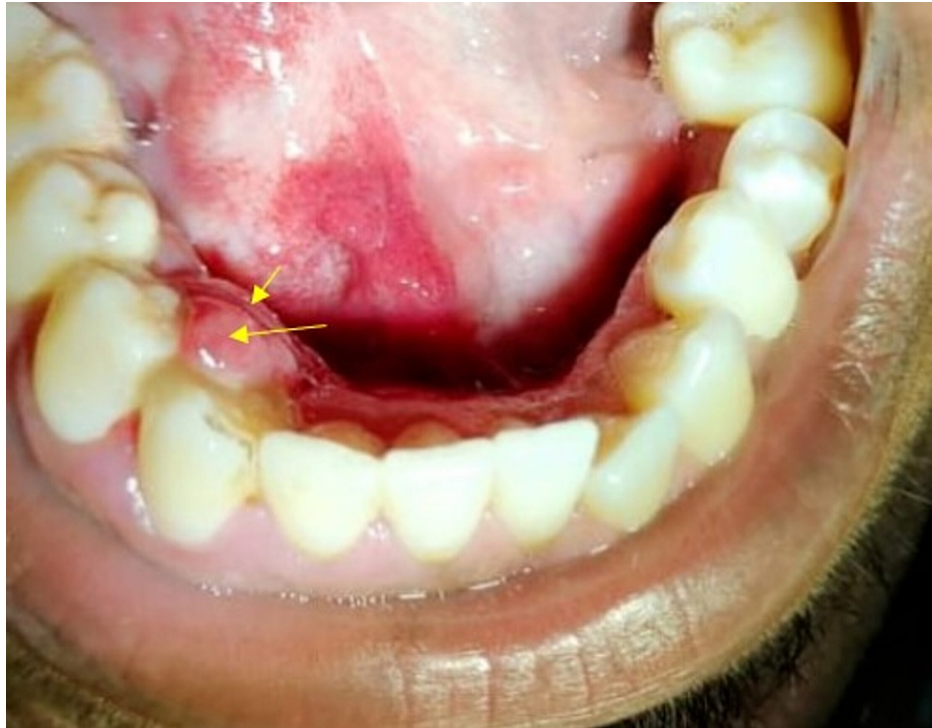


FIGURE 5: Postoperative intraoral clinical photograph in the one-month follow-up

The other management options for capillary haemangioma include: Tixel-induced rapamycin delivery following pulsed dye laser, hematoporphyrin monomethyl ether photodynamic therapy (HMME-PDT), sirolimus drug therapy [6-8].

Discussion

The port-wine stain is a non-neoplastic hamartomatous proliferation of capillaries resulting in nodular growth, as a result of ectatic blood vessels [2,9-12]. A port-wine stain clinically presents from birth and such lesions do not regress or undergo involution over a period of time. Port-wine stains are formed by somatic mutations of *GNAQ1*, *P13K* and dysregulation of vascular MAPK and/or PI3K signalling vascular pathway during human embryonic development [1].

Port-wine stain was first described as granuloma pyogenicum. It is a form of nevus flammeus affecting the capillaries in the dermis and is more common on the external surface of the facial skin. They are distributed along the dermatomal branches supplied by the trigeminal nerve. They occur clinically as asymptomatic, nodular growth involving the gingiva, non-tender, compressible, and reducible on palpation and also showing a positive modified diascopy test, on the application of pressure (by the back end of the mouth mirror). Such growth was friable and tendency to bleed [6]. The unilateral distribution of the reddish colour not crossing the midline in relation to the right ventral surface of the tongue and floor of the mouth provided a clue to the diagnosis. Such encountered growth must be carefully dealt with and a proper clinical and radiographic examination should be done before a surgical excision is attempted [7,8,13-15]. The various investigations and management modalities for port-wine stain are listed in Table 1.

| Investigation modalities | Management modalities |
|---|--|
| Dark ground illumination with confocal reflectance microscopy | Pulsed dye lasers (wavelength=585 nm) |
| Artificial RBCs - Haemoglobin vesicles | Cryotherapy: Cryoprobes with cryogen- liquid freeze-dried Nitrogen (-196 ^o C) |
| Dermatoscopy - capillaroscopy | Sclerotherapy: ethionamide |
| | Injection of antiangiogenic agents: sirolimus, retinoids, corticosteroids, propanolol |
| | Photosensitizers: artificial RBCs –haemoglobin vesicles in laser therapy |
| | Photodynamic therapy using hematoporphyrin (HMME-PDT) |

TABLE 1: Various Investigation and management modalities for port-wine stain

The various research studies on the concurrent occurrence of lobular capillary hemangioma and port-wine stain are listed in Table 2. Swerlick et al. stated that the clinical appearance of nodular growth of lobular capillary hemangioma can be mistaken for Kaposi sarcoma, hence termed Pseudo-Kaposi sarcoma [16]. Sheehan et al. stated that the presence of pyogenic granuloma in areas of port-wine stain can mimic malignancy [17]. Rodins et al. reported the occurrence of pyogenic granuloma and port-wine stain in pregnant patients [18]. Madi et al. stated that port-wine stains are distributed along the branches of the trigeminal nerve [19]. Luna-Ceron et al. stated that lobular capillary hemangiomas are so called because they are characterized histologically by the lobular arrangement of capillaries in a fibrous stroma. They occur clinically as a bright red, exophytic, nodular growth on the gingiva, which are friable on slight provocation and are susceptible to bleeding. Port-wine stains also occur as bright red macules and characteristically do not cross the midline. Both lobular capillary haemangioma and port-wine stain have mutations in the G protein subunit alpha q (GNAQ), implying that lobular capillary haemangioma emerges from port-wine stain cells [20].

| Author | year | Age/Gender | No. of cases |
|------------------------|------|------------------------------------|--------------|
| Swerlick et al. [16] | 1983 | 22-year-old female, 8-year-old boy | 2 |
| Sheehan et al. [17] | 2004 | 35-year-old male | 1 |
| Rodins et al. [18] | 2011 | 23-year-old female | 1 |
| Madi et al [19] | 2017 | 33-year-old female | 1 |
| Luna-Ceron et al. [20] | 2021 | 21-year-old-male | 1 |

TABLE 2: Research studies on cases of concurrent occurrence of lobular capillary haemangioma with port-wine stain

Conclusions

Haemangioma affecting the capillaries can occur as growth on the gingiva. such growth on the gingiva is friable easily and patients often complain of bleeding from the gingiva while brushing their teeth and may fear cancer and visit a dentist in doubt. Haemangiomas occurring in the gingiva are friable, have an increased tendency to bleed even on slight provocation during brushing. Bleeding, ulcer, airway compromise are some of the main complications that can be caused by a hemangioma if left untreated. Hence, it is always better to treat hemangioma occurring on the gingiva. Any growth involving the gingiva present from birth with a pinkish surface hue showing a positive modified diascopy test must be suspected for the vascular origin. Surgical procedures, which involve excision of such growth involving the gingiva, must be performed with precautions to avoid postoperative complications such as excessive bleeding, which can lead to hypovolemic shock.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

Acknowledgements

We would like to thank the Government Dental College, Salem, Tamil Nadu, India for its support

References

1. Sabeti S, Ball KL, Burkhart C, et al.: Consensus statement for the management and treatment of port-wine birthmarks in Sturge-Weber syndrome. *JAMA Dermatol.* 2021, 157:98-104. [10.1001/jamadermatol.2020.4226](https://doi.org/10.1001/jamadermatol.2020.4226)
2. Shajil C, Das JM: Nevus flammeus. StatPearls Publishing, Treasure Island, FL; 2021.
3. Sarwal P, Lapumnuaypol K: Pyogenic granuloma. StatPearls Publishing, Treasure Island (FL); 2021.
4. Singh AK, Keenaghan M: Sturge-Weber syndrome. StatPearls Publishing, Treasure Island (FL); 2021.
5. Bhasin M, Bhasin V, Bhasin A: Peripheral ossifying fibroma. *Case Rep Dent.* 2013, 2013:497234. [10.1155/2013/497234](https://doi.org/10.1155/2013/497234)
6. Artzi O, Mehrabi JN, Heyman L, Friedman O, Mashiah J: Treatment of port wine stain with Tixel-induced rapamycin delivery following pulsed dye laser application. *Dermatol Ther.* 2020, 33:e13172. [10.1111/dth.13172](https://doi.org/10.1111/dth.13172)
7. Zhang M, Wu Q, Lin T, et al.: Hematoporphyrin monomethyl ether photodynamic therapy for the treatment of facial port-wine stains resistant to pulsed dye laser. *Photodiagnosis Photodyn Ther.* 2020, 31:101820. [10.1016/j.pdpdt.2020.101820](https://doi.org/10.1016/j.pdpdt.2020.101820)
8. Sun B, Han T, Wang Y, Gao Q, Cui J, Shen W: Sirolimus as a potential treatment for Sturge-Weber syndrome. *J Craniofac Surg.* 2021, 32:257-60. [10.1097/SCS.00000000000007034](https://doi.org/10.1097/SCS.00000000000007034)
9. Fölster-Holst R, Shukla R, Kassir M, et al.: Treatment update of port-wine stain: a narrative review. *J Drugs Dermatol.* 2021, 20:515-8.
10. Nguyen V, Hochman M, Mihm MC Jr, Nelson JS, Tan W: The pathogenesis of port-wine stain and Sturge Weber syndrome: complex interactions between genetic alterations and aberrant MAPK and PI3K activation. *Int J Mol Sci.* 2019, 20:2243. [10.3390/ijms20092243](https://doi.org/10.3390/ijms20092243)
11. Almeida FT, Caldas R, Duarte MD, Brito C: Port-wine stain as a clue for two rare coexisting entities. *BMJ Case Rep.* 2018, 2018:bcr-2018-225721. [10.1136/bcr-2018-225721](https://doi.org/10.1136/bcr-2018-225721)
12. Sarac G, Kapicioglu Y: Efficacy of 577-nm Pro-Yellow laser in port wine stain treatment. *Dermatol Ther.* 2019, 32:e13078. [10.1111/dth.13078](https://doi.org/10.1111/dth.13078)
13. Flors L, Hagspiel KD, Park AW, Norton PT, Leiva-Salinas C: Soft-tissue vascular malformations and tumors. Part 2: low-flow lesions. *Radiologia (Engl Ed).* 2019, 61:124-33. [10.1016/j.rx.2018.02.012](https://doi.org/10.1016/j.rx.2018.02.012)
14. Fu Z, Huang J, Xiang Y, et al.: Characterization of laser-resistant port wine stain blood vessels using in vivo reflectance confocal microscopy. *Lasers Surg Med.* 2019, 51:841-9. [10.1002/lsm.23134](https://doi.org/10.1002/lsm.23134)
15. Fusano M, Bencini PL: Capillaroscopy and reflectance confocal microscopy characterization of refractory port-wine stains. *Lasers Med Sci.* 2021, 36:407-12. [10.1007/s10103-020-03084-1](https://doi.org/10.1007/s10103-020-03084-1)
16. Swerlick RA, Cooper PH: Pyogenic granuloma (lobular capillary hemangioma) within port-wine stains. *J Am Acad Dermatol.* 1983, 8:627-30. [10.1016/s0190-9622\(83\)70068-x](https://doi.org/10.1016/s0190-9622(83)70068-x)
17. Sheehan DJ, Leshner JL Jr: Pyogenic granuloma arising within a port-wine stain. *Cutis.* 2004, 73:175-80.
18. Rodins K, Gramp D, James D, Kumar S: Pyogenic granuloma, port-wine stain and pregnancy. *Australas J Dermatol.* 2011, 52:e8-e10. [10.1111/j.1440-0960.2010.00680.x](https://doi.org/10.1111/j.1440-0960.2010.00680.x)
19. Madi M, Babu SG, Achalli S, et al.: Oral pyogenic granuloma in a port-wine stain: a rare co-occurrence. *SRM J Res Dent Sci.* 2017, 28:37-40.
20. Luna-Ceron E, Gómez-Gutiérrez AK, Gonzalez-Hernandez C, Gatica-Torres M: An uncommon presentation of pyogenic granuloma. *Cureus.* 2021, 13:e12509. [10.7759/cureus.12509](https://doi.org/10.7759/cureus.12509)