Venular (Capillary) Vascular Malformation of Maxillofacial Region: Portwine Stain

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ABSTRACT

Background: Vascular Malformations (VMs) are localized defects of vascular morphogenesis resulting in formation of tortuous enlarged vascular channels. They become clinically visible during early childhood and persist throughout life.

Case description: Large reddish-pink macule involving left maxillary facial skin and mucosa from birth which was clinically misdiagnosed as hemangioma. The macule extended from the facial midline and involved the left forehead, bridge of the nose, and the left philtrum. The left half of upper lip was thick and prominent, sagging and redder in color compared to the right half and the lower lip. She gave a history of progressive complete loss of vision from the left eye without restriction of ocular movements.

Clinical implications: It is important that the correct nosologies for various VMs are followed, and they should be differentiated from and never wrongly termed as hemangioma. It is important that we adhere to correct clinical diagnosis as the therapeutic guidelines and follow-up of these lesions differ.

Keywords: Vascular malformations, Hemangioma, Classification, Capillary, Malformation, Portwine stain, International society for the study of vascular anomalies.


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INTRODUCTION

Vascular malformations (VMs) are developmental anomalies of the vascular plexus, whose etiopathogenesis are not properly understood.1-5 They occur in 1 to 1.5% of live births, do not have gender or race predilection, and are always present at birth.1-6 They may become clinically visible during early childhood, persist throughout life, and grow slowly with the person’s growth.1-6 The International Society for the Study of Vascular Anomalies classification differentiates vascular lesions with proliferative endothelium (hemangioma) from structural anomalies (VMs).2,7 VMs are presently categorized depending on the dynamics of vascular flow into slow or low-flow and fast or high-flow VMs.1,2,5,7

CASE REPORT

A 38-year-old woman reported with mobility and associated pain in relation to maxillary left canine and lateral incisor. She was referred to our teaching institution by a clinician who had made a diagnosis of hemangioma, and hence requiring expert management and medical care. Extraoral examination showed a large reddish-pink macule involving the left maxillary and ophthalmic dermatome, without the involvement of mandibular dermatome (Fig. 1). The macule extended from the facial midline involving the left forehead, bridge of the nose, left philtrum, to but not involving the external ear. The left half of upper lip was thick and prominent, sagging, and redder in color compared to the right half and the lower lip. The facial skin over the left maxilla had a rough texture and the macule was nontender, not warm to touch, nonpulsatile and without thrill. A deviated nasal septum and a large black nevus near the ala of nose were identified. She had complete loss of vision from the left eye, with change in pupil coloration, and without restriction of movement of eyeball or upper eyelid. She gave a history of gradual loss of vision in this eye from late childhood. The facial macule was present from birth and has become prominent, darkened in color with loss of normal skin texture, and grown commensurately with her development. She never took any medical advice for the discoloration or for the loss of vision due to economical constraints. She gave no relevant history of any systemic disease, convulsive disorders, or other lesions on any other part of her body, and had adequate mental ability. Intraoral examination revealed very poor oral hygiene with chronic generalized periodontitis, halitosis, generalized grade I-II mobility of teeth, and missing maxillary left first premolar. The left maxillary labial and buccal mucosa, vestibule, gingiva, and hard palate from the midline were redder in color in comparison to the other quadrants (Fig. 2). The maxillary left canine and lateral incisor had grade II mobility and was tender to percussion. She was not willing due to poor financial status for further investigations or therapeutic intervention of the facial discoloration and the ophthalmic manifestation. Based on the clinical history, appearance, and physical examination, a clinical diagnosis

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squamous surface epithelium with underlying associated fibrovascular connective tissue having moderate collagen fibers, numerous large and small blood vascular channels some congested with RBCs, and chronic inflammatory cells (Fig. 3). A histopathological diagnosis of capillary vascular malformation was made after correlation with clinical history and features. Clinical photographs were taken prior to treatment and a written patient consent was obtained to reproduce or publish them along with her clinical details for academic advancement without revealing her personal identity.

DISCUSSION

The Greek suffix ‘oma’ means cellular tumoral proliferation and thus the term hemangioma should never be used for describing a VMs which are developmental anomalies and are not true tumors. Capillary malformations (CMs) can be separated into the following:

1. Port-wine stain (other names: telangiectatic nevus, nevus flammeus, traditional CMs).
2. Salmon stain (other names: stork bite, angels kiss, mid-line CMs).
3. Hereditary hemorrhagic telangiectasia (HTT).

Port-wine stain occurs in 0.4 to 1% of newborns and appear as reddish-pink macules without any thrill or bruit over facial dermatomes supplied by the branches of the trigeminal nerve. In 1999, Waner and Suen based on their identification of the anomalies of these lesions in the postcapillary venules (rather than in the capillaries) recategorized them as venular VMs. A total of 90% of the port-wine stain affect more than one dermatome with the combined affliction of maxillary and ophthalmic dermatomes the most common. With age the lesions darken in color and thicken to produce a violaceous colored cobblestone surfaced vascular lesion. The affected area usually show hypertrophy of the associated jaw bones, gingiva, oral mucosa and lips, leading to interdental spacing, malocclusion and unesthetic thick sagging lip. A total of 10% of port-wine stains are associated with Sturge-Weber syndrome which is characterized by CMs mainly involving the maxillary and ophthalmic dermatomes, hemifacial hyperplasia, and ipsilateral leptomeningeal VMs of the cerebral cortex. The leptomeningeal VMs are diagnosed by the presence of gyriform ‘tramline’ calcification on skull imaging studies, and are usually associated with ophthalmic manifestations, convulsive disorder, mental retardation, and contralateral hemiplegia. Salmon stain are present from birth as pink-red macule commonly on the midline of the forehead along the anatomic areas innervated by the supra-s
trochlear and supraorbital nerves, and sometimes involving the glabella, supra-alar region and philtrum. They never progress or show hypertrophy, and involute to disappear by the age of 1 year in 55 to 65% of the newborns. HTT is an inherited autosomal dominant mucocutaneous disorder affecting 1 in 5 to 8000. The Curacao criteria helps in arriving at a definite diagnosis of HHT if atleast three of the following clinical manifestations are present:

1. Spontaneous recurrent epistaxis.
2. Multiple mucocutaneous telangiectasia involving finger-tip, lips, oral mucosa, or tongue.
3. Gastrointestinal, pulmonary, hepatic, cerebral, or spinal arteriovenous malformations (AVMs).
4. A first-degree relative with these clinical presentations.

Our case report had all the features of traditional capillary (vennular) malformation, which is commonly referred to as port-wine stain. As the patient was not willing for imaging or other investigatory studies we were unable to rule out Sturge-Weber syndrome, but she gave no clinical history of convulsive disorder, mental retardation, or hemiplegia.

Diagnosis of vascular anomalies can be made by the clinical history and the physical findings, with a minority requiring imaging studies and even smaller number requiring biopsy and histopathological examination. Doppler-ultrasound, magnetic resonance imaging, computerized topography, and angiography are necessary to identify the size, location, extent, and collateral feeders before any therapeutic or surgical interventions are attempted. The management of VMs depends on the type and flow characteristics of the malformation, the functional impairment, disfigurement, the threat to life, and on the expertise of the surgeon or the interventional radiologist. The therapeutic modalities include sclerotherapy, embolization, laser photocoagulation and surgical excision.

CONCLUSION

It is important that the correct nosology for various VMs is followed, and they should be differentiated from and never wrongly termed as hemangioma. VMs are vascular anomalies that are usually present from birth, persist throughout life, and grow slowly with the person. Hemangiomas are true tumors caused by endothelial cell proliferation, are not present at birth, and 90% involute by 9 years of age. In addition to overcoming obstacles in communication and research, it is important that we adhere to correct clinical diagnosis as the therapeutic guidelines for management and follow-up of these lesions differ. While describing VMs, discarded and sometimes erroneous terms like angioma, birthmarks, capillary hemangioma, lymphangioma, and inappropriate use of the term hemangioma should be avoided.

REFERENCES