### COURSE 06: DENTAL AND ORAL ISSUES IN PATIENTS WITH VASCULAR ANOMALIES- SURGERY

### Dr. David Darrow

# Introduction: Dental and oral issues in patients with vascular anomalies

There is considerable inaccuracy among both patients and clinicians regarding the diagnosis of vascular malformations. Even when vascular malformations are identified correctly, misinformation regarding their natural history and potential morbidity often leads to mismanagement. While not all vascular malformations of the oral cavity require intervention, the primary care practitioner should be familiar with symptoms and complications that suggest a need for active management.

This module serves to guide the primary care clinician in correctly identifying and managing vascular malformations of the oral cavity, and appropriately referring patients with such disorders when necessary. In this module, we will review for the primary care provider:



- > Types and locations of vascular malformations in the oral cavity;
- > Symptoms and complications of vascular malformations of the oral cavity; and
- > Treatment options for vascular malformations of the oral cavity.

### Objectives

Upon successful completion of this activity, participants should be able to:

- > Identify vascular malformations of the oral cavity based on appearance and distribution
- > Recognize symptoms and complications of vascular complications of the oral cavity
- > Choose interventions appropriate for specific oral cavity vascular malformations
- > Identify patients appropriate for referral to a specialist in vascular malformations

### Identifying vascular anomalies of the oral cavity

Most oral cavity malformations fall into one of the following categories:

- > Capillary malformations (CMs, or "port wine stains" [PWSs])
- > Venous malformations (VMs)
- > Lymphatic malformations (LMs)
- > Arteriovenous malformations (AVMs)

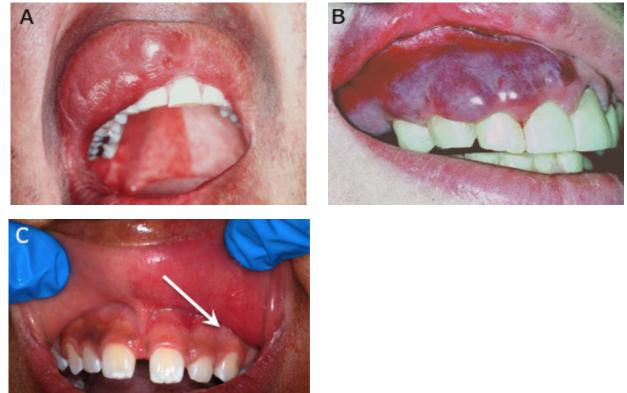
Hemangiomas are lesions of infancy and are not seen in adults.

#### **Capillary malformations (CMs)**



Early-stage CMs demonstrate reddish blush of the oral mucosa and gingiva without significant hyperplasia of the soft tissue or bone in the affected area.

Similar to other body locations, CMs of the oral cavity have a dermatomal distribution. As a result, unilateral lesions will have a sharp demarcation at the midline (A).



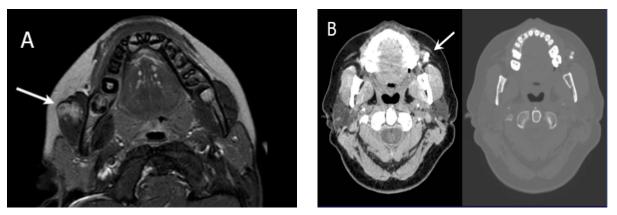
Late-stage lesions demonstrate increased blush of the oral tissues, as well as hyperplasia of the lips (A), gingiva (B), or bone (C [arrow]) in the affected area.

Venous malformations



Photos: VMs of the oral cavity

Pools of venous blood impart a blue-purple hue to the overlying tissues. Darker color implies a lesion closer to the surface. Oral cavity VMs transiently increase in size with increased venous pressure (head dependent, Valsalva).



VMs of the oral cavity that involve deeper tissues have a predilection for the masseteric space (A) and the buccal fat space (B). Characteristic phleboliths are seen in buccal space.

#### Lymphatic malformations (LMs)



Left photo: Diffuse microcystic lymphatic malformation of the tongue Right photo: Focal lymphatic malformation of the tongue

- > Most LMs affecting the oral cavity are microcystic (consisting of cysts <1 cm in diameter)
- > Most oral cavity LMs present superficially with clear or hemorrhagic vesicles
- > Lesions may be focal or diffuse
- > Most common subsite is the tongue



Left photo: Microcystic lymphatic malformation left oral vestibule Right photo: Lymphatic malformation of oral mucosa with extension into buccal space

- > Other oral cavity LM subsites include lips, buccal mucosa, soft palate, and floor of the mouth.
- > LMs may extend from oral mucosa into deeper tissue spaces (buccal, masseteric) or infiltrate soft tissues of lips or face.

#### Arteriovenous malformations



AVMs are the rarest of oral cavity vascular malformations, but approximately 50% of AVMs involve the oral and maxillofacial region.

AVMs develop from a "nidus"- a bed of ectatic capillaries across which there is shunting of blood from high to low pressure. AVMs can be distinguished based on:

- > Later age at presentation
- > Hypertrophied arteries, dilated veins
- > Firmness and delay in emptying on palpation
- > Characteristics of high-flow lesion: palpable thrill, audible bruit
- > High output cardiac failure or distal ischemia at presentation

# Complications of vascular anomalies of the oral cavity

#### Bleeding

- > Uncommon in CMs, even in dental and oral surgical procedures
- > Uncommon in VMs, unless traumatized or bitten
- > Common in LMs with surface involvement when lesions are inflamed or traumatized; vesicles become hemorrhagic
- > Bleeding usually mild, more a nuisance than medical issue
- > Common in AVMs, especially those in which surface tissues have become ulcerated or necrotic
- > Bleeding occasionally severe due to involvement of arterial feeders

#### **Tissue distortion**

All vascular malformations have the potential to distort underlying soft tissue and bone due to pressure on these tissues.

#### Tissue overgrowth

CMs, VMs, and LMs cause tissue overgrowth due to:

- > Vascular expansion with the affected tissue
- > Effects of hypothesized growth factors on underlying soft tissue and bone
- > In oral cavity, overgrowth more common in CMs and LMs

#### Tissue destruction

AVMs cause destruction of soft tissue and bone due to:

- > Tissue ischemia from shunting of blood
- > Tissue replacement by the mass

### Oral tissue changes with CMs



May result in:

- > Periodontal disease associated with gingival overgrowth (A)
- > Bony overgrowth (occasionally resulting in malocclusion of the teeth)



May result in:

> Hyperplasia of the lips (B)

### Oral tissue changes with LMs, VMs, and AVMs

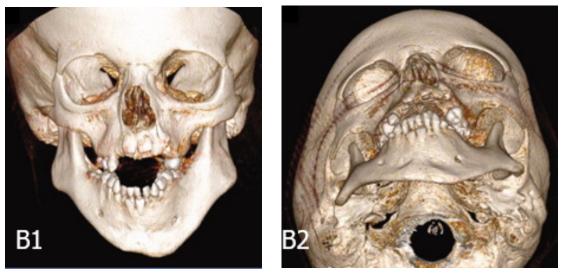
May result in difficulties with airway obstruction, speech, mastication, swallowing, and oral hygiene due to:

- > Tongue enlargement
- > Elevation of the floor of mouth
- > Open bite deformity
- > Distortion of the mandible and/or maxilla Malocclusion of the teeth
- > Severe bleeding (mainly AVMs)

### Imaging of vascular anomalies of the oral cavity



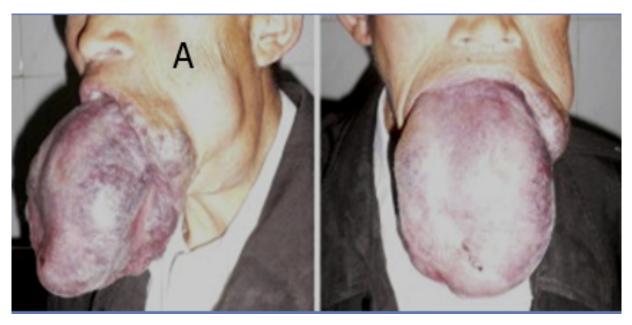
LM of the tongue and oral vestibule. Note anterior open-bite deformity due to tongue swelling and vesicles of oral mucosa.



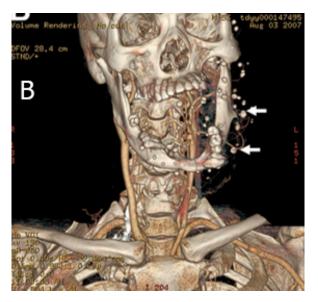
3D computerized tomography of patient with oral cavity LM. Note anterior open-bite deformity (B1).

3D computerized tomography of patient with oral cavity LM. Note deformation of the mandible and leftsided crossbite (B2).

# Imaging of vascular anomalies of the oral cavity



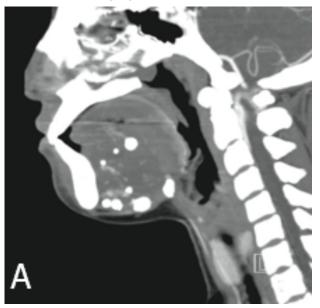
(A): Massive VM of the tongue, with risk of airway obstruction.



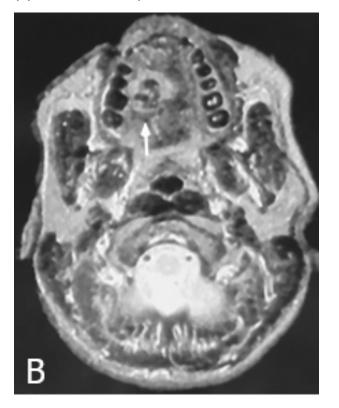
B): 3D computerized tomography of above patient demonstrating open-bite deformity and severe maxillomandibular distortion. Arrows indicate phleboliths characteristic of VMs.

# Imaging of vascular malformations of the oral cavity

As a rule, imaging of oral cavity CMs is not necessary. Conversely, imaging of oral cavity VMs, LMs, and AVMs is critical to establish the diagnosis and assess the extent of the lesion. For soft tissue lesions, MRI imaging is best; for bone lesions, CT is needed.



(A): Phleboliths are present on CT scan of VMs



(B): Signal voids due to flow are characteristic of VMs and AVMs

#### Management of oral cavity CMs





Goals include:

- > Prevention of periodontal disease by:
  - > Meticulous oral hygiene
  - > Excision of overgrown tissue (A)
  - > Periodontal surgery (B)
- > Maintenance of dental occlusion includes:
  - > Orthodontic intervention
  - > Osteotomy in severe cases of bony overgrowth

#### Management of oral cavity VMs





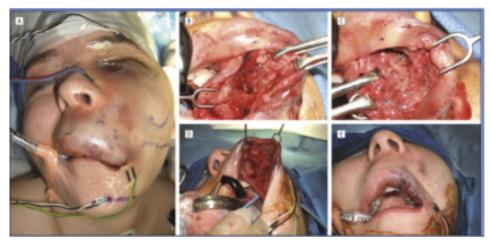
Goals include reduction of bleeding

- > Laser treatment of the mucosa
  - > Long wavelength lasers for deeper penetration
  - > (Nd:YAG, diode) Also helps preserve mucosa

-Nd:YAG (1064 nm) -Diode (980 nm)

- > Sclerotherapy and embolization
  - > Sodium tetradecyl sulfate (Sotradecol®)
  - > Ethylene vinyl alcohol copolymer (Onyx®)
  - > N-butyl-2-cyanoacrylate (nBCA)
  - > 100% ethanol

Management of oral cavity VMs (2)



Surgical excision of buccal VM with buccal space extension. Electrodes are for monitoring of facial nerve during procedure. Goals include:

- > Reduction of bulk affecting oral function and hygiene by
  - > Surgical resection
  - > Sclerotherapy
- > Preparation for surgery
  - > Laser
  - > Sclerotherapy

### Management of vascular anomalies of the oral cavity

Management of acute swelling and pain due to inflammation in oral cavity LMs



- > Oral steroids are the most effective intervention (taper from initial dose of 50-60 mg of prednisone).
- > Antibiotics may be added when infection suspected as source of inflammation.
- > Inflammation may continue to improve for 2-4 weeks after medical intervention.
- > mTor inhibitors are used as well (sirolimus, everolimus).
- > Tracheotomy may be necessary if airway is obstructed.

#### Long-term management of oral cavity LMs



(A) Lymphatic malformation causing airway obstruction necessitating tracheotomy.



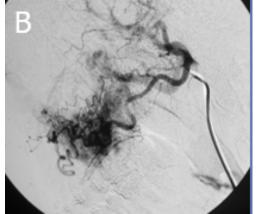
(B) Tongue after two surgical reductions.

Goals include:

- > Restoration of tongue and/or mucosa to normal size and normal surface characteristics using:
  - > Surgery
  - > CO2 laser
  - > Bleomycin injection
  - > mTor inhibitors
  - > Tracheotomy
- > Correction of dental and skeletal deformities by means of:
  - > Orthodontia
  - > Surgery

Management of oral cavity AVMs





(A) CT demonstrating lytic lesion of left maxilla

(B) pre-embolization angiography demonstrating characteristic blush of AVM in left maxilla



(C) surgical window in cortical for access to embolized AVM

Goals include:

- > Reduction of bleeding by
  - > Ligation of offending vessels
  - > Embolization
  - > Surgical resection
- > Reduction of bulk, affecting oral function
  - > Surgical resection (excision of the "nidus" is key to avoiding recurrence)
  - > Resection usually follows embolization

## Oral tissue changes with CMs



May result in:

- > Periodontal disease associated with gingival overgrowth (A)
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May result in:

> Hyperplasia of the lips (B)

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### Course 06: Lesson Quiz

- 1. The most important step in surgery for arteriovenous malformations is:
- A. Ligation of the draining vein
- B. Embolization
- C. Excision of the nidus
- D. Ligation of the feeding artery
- 2. Superficial venous malformations limited to the oral mucosa of limited depth are best treated using:
- A. Improved oral hygiene
- B. Sclerotherapy
- C. Laser
- D. Surgical resection
- 3. Acute swelling and pain associated with a traumatized lymphatic malformation are best managed with:
- A. Corticosteroids
- B. Antibiotics
- C. Narcotics
- D. Ice packs
- 4. Overgrowth of oral tissues due to a capillary malformation may result in:
- A. Bleeding
- B. Pain
- C. Periodontal disease
- D. Bleeding and pain
- 5. The most important complication of venous and lymphatic malformations of the oral cavity is:
- A. Bleeding
- B. Airway obstruction
- C. Mandibular deformity
- D. Malocclusion
- 6. Bleeding from the tongue associated with microcystic lymphatic malformations is:
- A. Brisk, because lymphatic malformations are high-flow lesions
- B. Impossible, because lymphatic malformations are filled with lymph, not blood
- C. Slow, because it is due to oozing from inflamed vesicles
- D. Slow, because lymphatic malformations are low-flow lesions

7. A patient who has had no prior history of oral lesions or swelling suddenly develops pain and swelling in the tongue and floor of mouth after an upper respiratory infection. Two red vesicles are seen on the ventral surface of the tongue. This patient most likely has a(n):

- A. Capillary malformation
- B. Lymphatic malformation
- C. Venous malformation
- D. Arteriovenous malformation

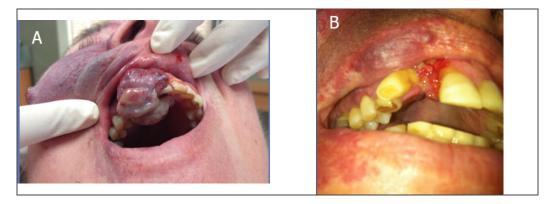
9. With which type of vascular anomaly might one see ulceration of oral tissues due to ischemia from shunting of blood?

- A. Capillary malformation
- B. Arteriovenous malformation
- C. Venous malformation
- D. Lymphatic malformation
- 9 The purple-blue color of a venous malformation of the oral cavity is due to which of the following?
- A. Proximity of lakes of venous blood to the surface mucosa
- B. Infection
- C. High flow of blood through the lesion
- D. Trauma

10 Which type of vascular anomaly can be recognized in the mouth by blush and overgrowth of the affected tissue?

- A. Capillary malformation
- B. Venous malformation
- C. Lymphatic malformation
- D. Arteriovenous malformation

### Case Study 1

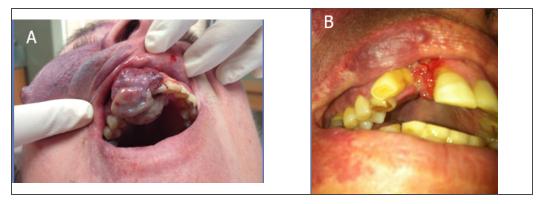


11. A 35-year old female with a facial port wine stain (capillary malformation) with oral cavity involvement presented with a mass on the upper jaw (A). The lesion had been present for 6 months and bled intermittently. It was resected, returning a diagnosis of "pyogenic granuloma," and now is recurring (B).

The likely cause of the initial mass is:

- A. Use of toothpicks
- B. Tissue overgrowth due to the malformation
- C. Irritation due to cigarette smoking
- D. Malignant transformation of the malformation

### Case Study 1.2

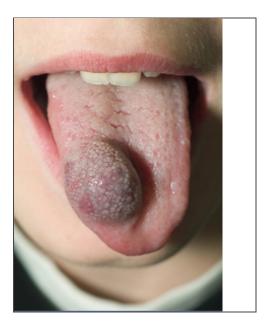


12. A 35-year old female with a facial port wine stain (capillary malformation) with oral cavity involvement presented with a mass on the upper jaw (A). The lesion had been present for 6 months and bled intermittently. It was resected, returning a diagnosis of "pyogenic granuloma," and now is recurring (B).

The most important factor in preventing recurrence of this mass after re-excision is:

- A. Laser treatment of the involved tissue
- B. Injecting steroids into involved tissue
- C. Using electrosurgery for the excision rather than a scalpel
- D. Meticulous oral hygiene and periodontal care

### Case Study 2



13. An adolescent male presents with a vascular lesion of the tongue. The lesion has been slowly increasing in size but has not bled. It occasionally interferes with speech but not with swallowing. It has not been painful. The patient does note that it increases in size when his head is dependent.

This vascular lesion should be classified as a:

- A. Arteriovenous malformation
- B. Hemangioma
- C. Venous malformation
- D. Lymphatic malformation

# Case Study 2.1



An adolescent male presents with a vascular lesion of the tongue. The lesion has been slowly increasing in size but has not bled. It occasionally interferes with speech but not with swallowing. It has not been painful. The patient does note that it increases in size when his head is dependent.

14. Laser treatment for the surface tissue:

- A. Is insufficient and superfluous
- B. Should never be used on the tongue
- C. Is insufficient to treat the deeper portions of the lesion
- D. Is superfluous since the lesion is exophytic and the mucosa can be sacrificed

### AUTHOR PROFILES



#### David H. Darrow, MD, DDS

David H. Darrow, MD, DDS, is Professor of Otolaryngology-Head & Neck Surgery at Eastern Virginia Medical School in Norfolk, VA. He is the founder and co-director of the EVMS Center for Hemangiomas and Vascular Birthmarks, and his clinical practice includes vascular lesions of the head and neck in children and adults.

Dr. Darrow has served as a member and chair of the executive committee of the American Academy of Pediatrics Section on Otolaryngology-Head &

Neck Surgery, and as a member of the Board of Directors of the American Society of Pediatric Otolaryngology. Dr. Darrow has authored a study of orodental manifestations of facial port wine stains, and was lead author of the AAP Clinical Report on Infantile Hemangiomas. He has also authored several book chapters onvascular anomalies in neonates and children.

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