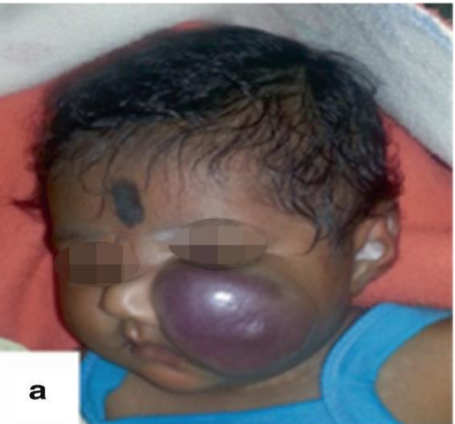
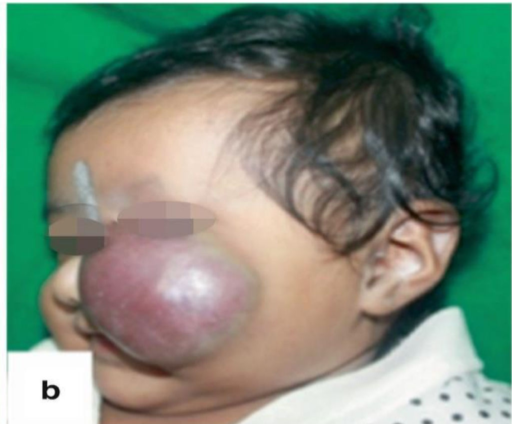
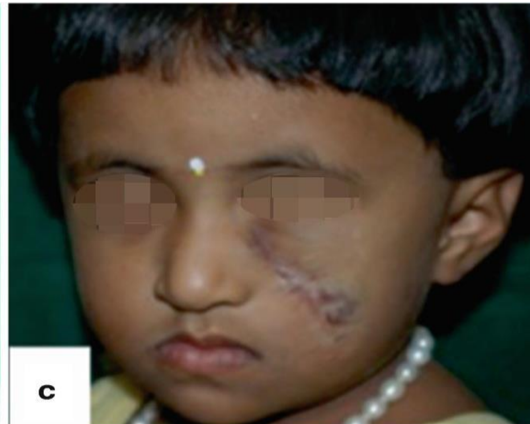
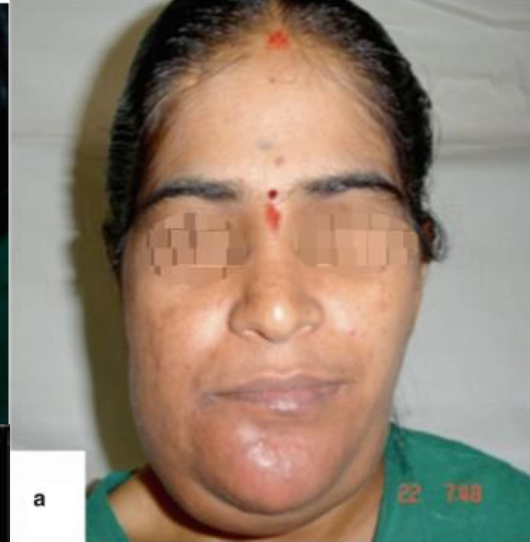
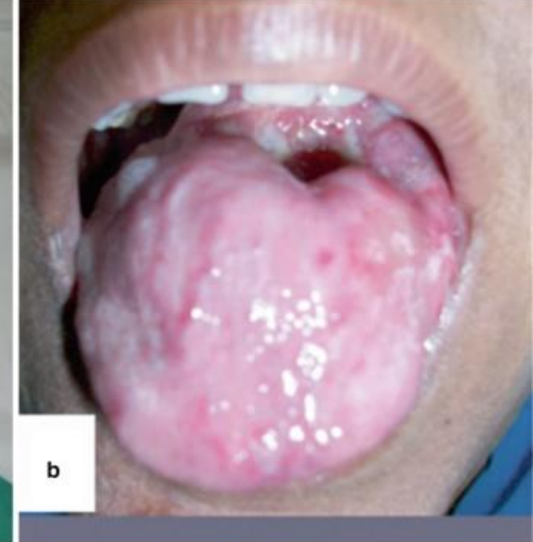
Vascular Anomalies of the Oro-Maxillofacial Region











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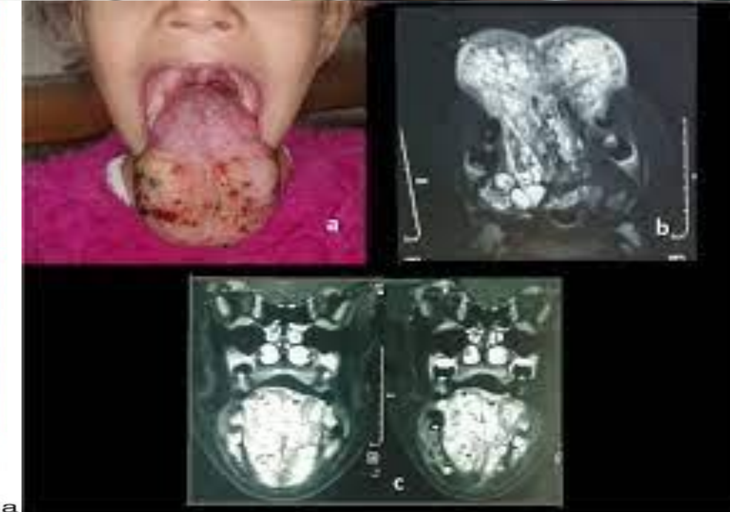

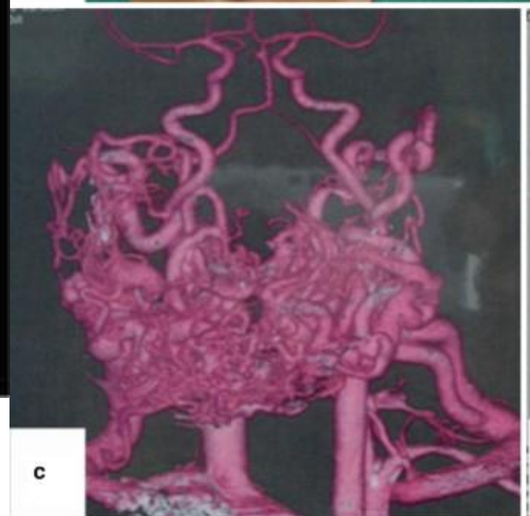

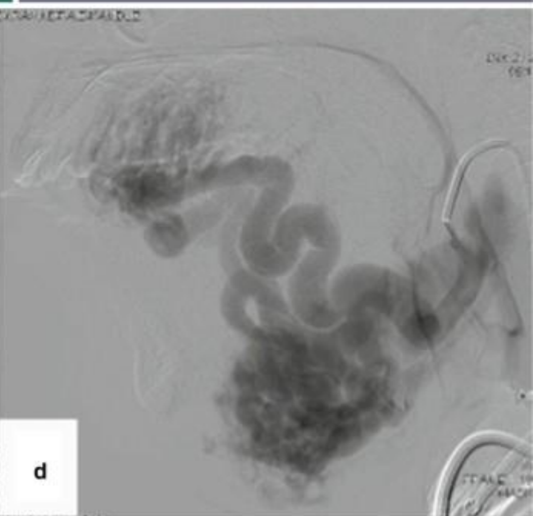

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PROFESSOR ABBAS AY TAHER

Vascular anomalies :

Vascular anomalies are heterogeneous group of congenital lesions of abnormal vascular development and may occur **anywhere on the body**.

Vascular lesions can be **broadly divided into two group :**

A-vascular tumors and B-**vascular malformations**

Vascular tumor are true neoplasm that arise from **endothelial hyperplasia**

Vascular malformation are congenital lesions of vascular **dysmorphogenesis** that arise because of erroes of embryonic development , these lesion exhibit normal endothelial cell turnover.

There is a primary **distinction** between a vascular tumor, which grows by **cellular hyperplasia**, and **a vascular malformation**, which represents a **localized defect in vascular morphogenesis**.

Due to the differences in **biologic behavior** and **radiographic features**, **malformations** are further subdivided into **low-flow** and **high-flow lesions** .

The common characteristic feature of all vascular anomalies is **extreme bleeding during surgical excision**.

Differences

Vascular tumor

- Abnormal endothelial cell proliferation
- Rapid growth
- No associated thrill and bruit
- Usually undergoes spontaneous involution
- Recurrence uncommon
- More commonly affect female

Vascular malformation

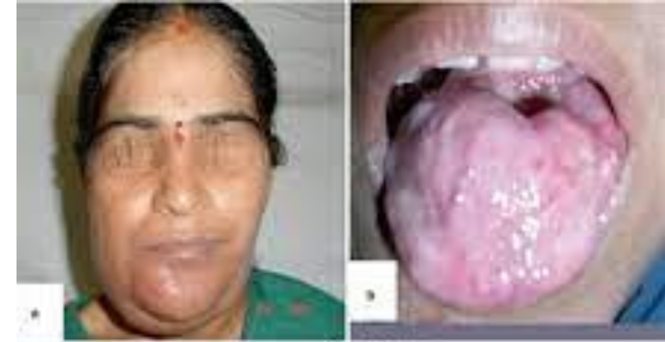
- Normal endothelial cell turnover
- Grows with patient
- May produce thrill and bruit
- Does not involute
- Recurrence common

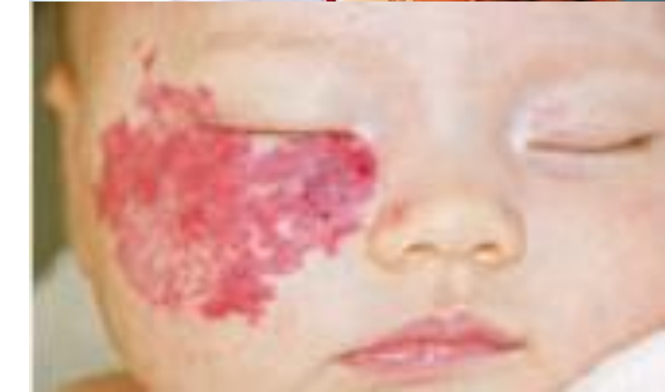
Clinicians throughout the world use the **classification by Mulliken and Glowacki (1982)** to classify these lesions. This classification is based on the **vascular lesion's histology**, **biological behavior** and **clinical findings**.
Some of the lesions **cause esthetic problems**, while some of them are malignant; thus, the **therapeutic approach is variable.**

Vascular tumors	Vascular malformations
	Slow-flow
Infantile hemangioma	Capillary malformations
Congenital hemangioma	Venous malformations
Tufted angioma	Lymphatic malformations
Kaposiform hemangioendothelioma	Fast-flow
	Arteriovenous malformations

Classification of vascular anomalies by Mulliken and Glowacki.

Vascular lesions within the head and neck **have a broad pathological spectrum**. These include a variety of tumors and malformations ranging from **simple capillary irregularities** to **complex structures involving arteries, veins and lymphatics**. One of the most well-known is that **of Mulliken and Glowacki**, who published the **first histology-based scheme**, which described lesions in the **pediatric population**, dividing them into two **major groups, tumors, and malformations**
Vascular anomalies, consistent of **vascular tumors** and **malformations**, frequently arise in the head and neck and often occur in the pediatric patient.
Infantile hemangiomas are the most common tumors in infancy; 10% are **found by 1 year of age**, with white premature babies and girls affected more often. weighing less than **1000 g**, with **girls affected 3 to 5 more often than boys**. **Whites** are more prone to hemangioma development, whereas the incidence in African-Americans and Asians is **low (1.4% and 0.8%, respectively**).
Other risk factors include multiparity(Parity is the number of pregnancies carried to viable gestational age. A woman who has given birth two or more times is multiparous and is called a multip. Grand multipara describes the condition of having given birth three or more times. Classifying grand multiparous women as a high-risk group without clear evidence of a consistent association with adverse outcomes can lead to **socioeconomic burdens to the mother**, **family and health systems**, **advanced maternal age**, **placental abnormalities** (ie, placenta previa).



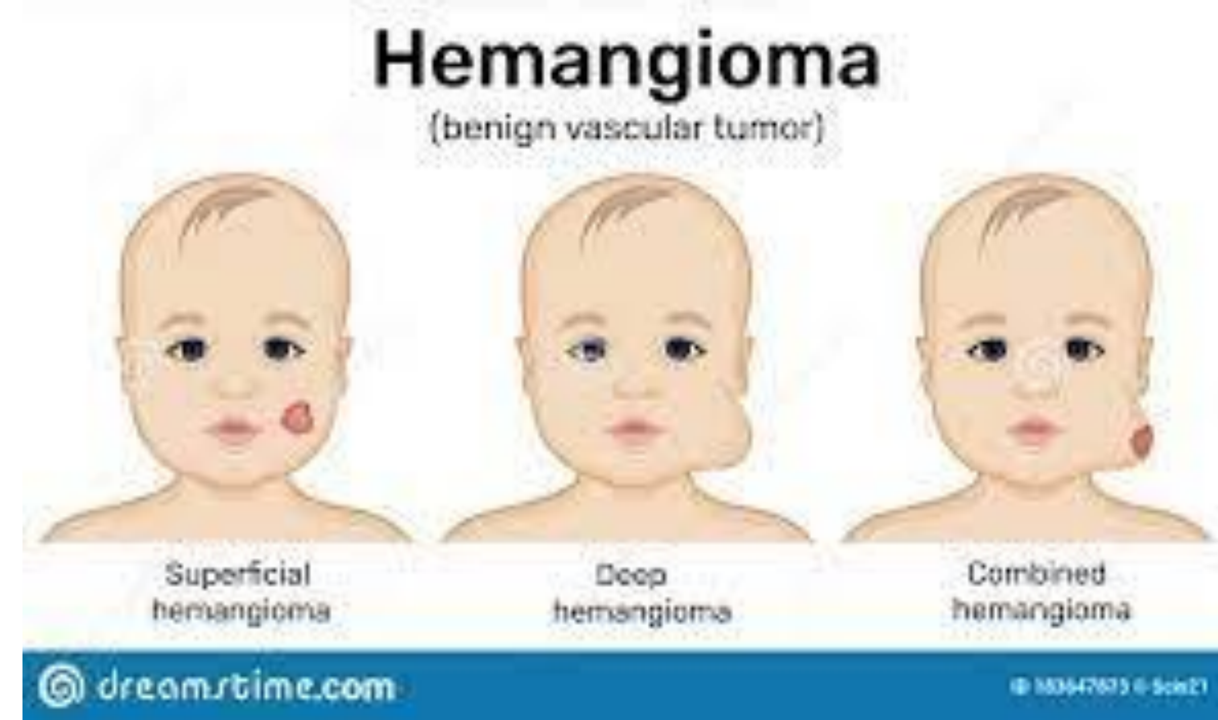


Hemangiomas are proliferative lesions characterized by increased turnover of endothelial cells.

Histologic landmarks are the **hyperplasia of endothelial cells,** which will stain positive for glucose transporter (in infertile hemangiomas), and the large number of mast cells.

The lesions appear as solitary lesions in approximately 80% of children and 60% of the tumors are found in the cervicofacial region.

The lesions occur sporadically, although there are some tumors that may follow an autosomal dominant inheritance pattern in familial cases.



Hemangiomas may be superficial, deep, or visceral, and, although the location does not alter the biologic behavior, it does affect the clinical manifestations.

The lesions typically appear during the **first 2 years of life,** if they are not present or evident at birth, as an erythematous patch, telangiectasia, or blanched area when they are located superficially.

Deep lesions may appear bluish or **a deep purple color**, whereas the visceral ones will generally not be evident on clinical examination.

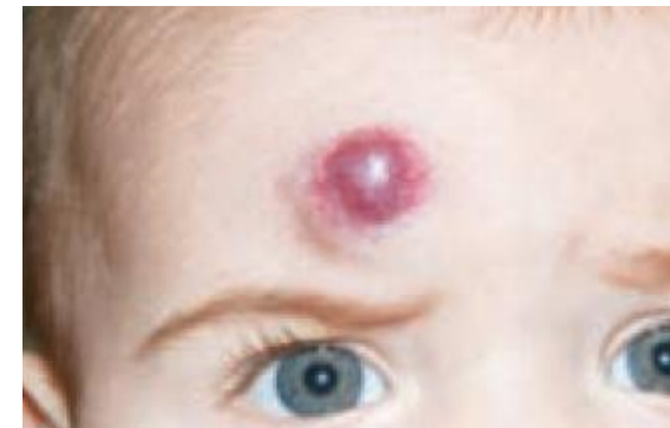
Infantile hemangiomas follow a triphasic pattern of evolution during which there is classically a proliferative phase of rapid enlargement that follows and outpaces the child's body growth, which may last for 4 to 8 months before a plateau phase that is consistent with body habitus growth followed by involution.

Vascular malformations represent an **abnormal proliferation of mature vascular elements**, are present **at birth,** and **show equal distribution between the sexes**.

Vascular malformations grow with the child and are usually affected by **hormonal changes with occasional accelerated growth during puberty.**

Capillary-venular and **venous malformations represent the most common vascular malformations, approaching 1 in 10,000**.

Lymphangiomas or lymphatic malformations are areas of abnormal development of the lymphatic system of unknown etiology, commonly found in the head and neck region. Surgery, sclerotherapy, and laser treatments have all been used with various success rates in the management of lymphatic malformations.



Introduction

In 1982, Mulliken and Glowacki proposed a biological classification for these lesions based on their **clinical** and **histologic findings**. Based on this classification scheme, which has been supported by **subsequent radiographic** and **biochemical studies**,

Vascular anomalies are **classified** as hemangiomas (**now known as infantile hemangioma**) and **vascular malformations (VMs**).

This classification was widely adopted and became the official classification of the **International Society for the Study of Vascular Anomalies in 1996** with some updates to include **infantile hemangioma variants**, combined lesions, and other benign vascular origin tumors.

International Society for the Study of Vascular Anomalies(ISSVA)

VASCULAR ANOMALIES

- **VASCULAR TUMORS**
  - BENIGN
    - Infantile hemangioma/Hemangioma of infancy
    - Congenital hemangioma
      - Rapidly involuting
      - Non-involuting
      - Partially involuting
    - Epithelioid hemangioma
    - Spindle-cell hemangioma
    - Tufted angioma
    - Pyogenic granuloma (aka lobular capillary hemangioma)
    - Others
  - LOCALLY AGGRESIVE OR BORDERLINE
    - Kaposiform hemangioendothelioma
    - Retiform hemangioendothelioma
    - Papillary intralymphatic angioendothelioma
    - Composite hemangioendothelioma
    - Kaposi sarcoma
    - Others
  - MALGINANT
    - Angiosarcoma
    - Epithelioid hamngioendothelioma

- **VASCULAR MALFORMATIONS**
  - SIMPLE
    - Capillary malformations
    - Lymphatic malformations
    - Venous malformations
    - Arteriovenous malformations
    - Arteriovenous fistula
  - COMBINED
    - CVM, CLM
    - LVM, CLVM
    - CAVM
    - Others
  - OF MAJOR NAMED VESSELS
  - ASSOCIATES WITH OTHER ANOMALIES:
    - Klippel-Trenaunay syndrome
    - Parkes Weber syndrome
    - Servelle-Martorell syndrome
    - Sturge-Weber syndrome
    - Maffucci syndrome
    - Proteus syndrome

This classification was widely adopted and became the **official classification of the International Society for the Study of Vascular Anomalies in 1996 with some updates to include infantile hemangioma variants, combined lesions, and other benign vascular origin tumors**.

Infantile hemangiomas are the most common tumors in infancy, found in as many as 10% of infants by 1 year of age, especially in white and premature babies

Diagnosis:

- History
- Clinical examination
- MRI with IV contrast
- Doppler ultrasound
- CT angiography

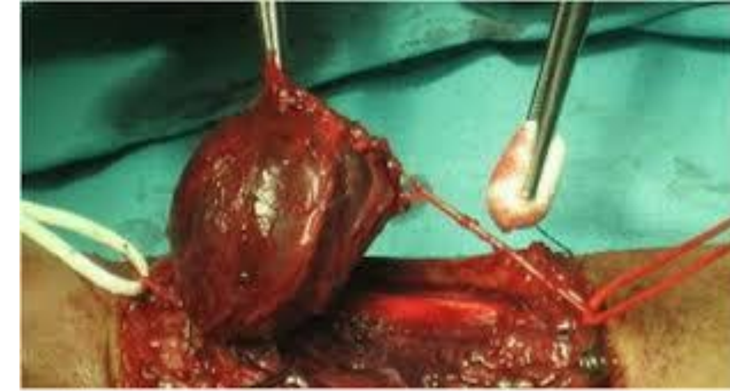
Infantile hemangioma

- The most common benign tumor in children
- Present shortly after birth (during 1st and 4th week of birth)
- Affect female more than male
- Three developmental phases
  - proliferation 2 months – 1 year
  - involuting 1 – 5 years
  - involuted 5 – 12 years

Congenital hemangioma

- Compeletly developed at the time of birth
- Do not pass the pathognomonic phase of IH
- M and F equally affected
- Types Rapidly involuting(RICH) Non-involuting (NICH) Partially involuting(PICH)

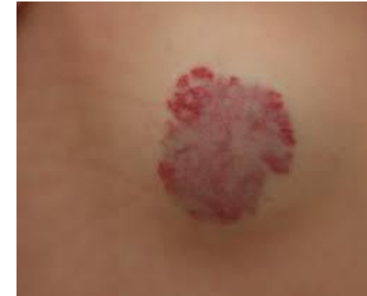
Treatment options



1-Medical therapy, 2-Surgical therapy

Before management you should know the followings :

A-The first and most important step in management of vascular lesions is to determine whether the **vascular lesion** is **tumor** or **malformation**



B-If **tumor** , determine whether it is **proliferating** or **involuting**
If **malformation** , determine **slow** or **high flow**

Indications for treatment

-Well localized tumor
-Ulceration
-Bleeding
-Airway obstruction
-Interfering with vision
-Facial features distortion

Medical therapy

Figure 3. Needle oriented toward forward flow of supraorbital

1-Intralesional steroids

-Block vascular endothelial growth factor

-Most commonly used for **upper eyelid hemangioma ,using 3 mg\kg once a week for 6 weeks**

2-Systemic corticosteroid

-Used for destructive, **function impairing** or **life-threatening lesions**

-Blocks vascular endothelial growth factor ,using 3mg\kg once a day for 2 weeks)

3 - Interferon alfa

-In cases of unresponsiveness to corticosteroid

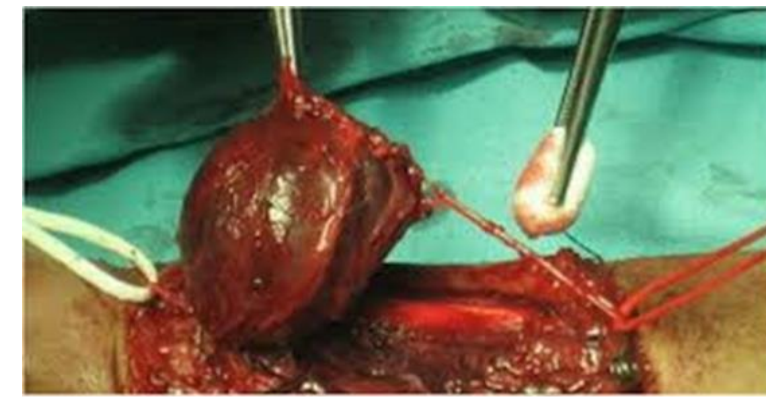
-Inhibit endothelial cell proliferation

-Inhibit angiogenesis,   using dose : 3 millions units\day subcutaneously , up tp 5 months

4-B-blocker (propranolol)

-Inducing apoptosis in endothelial cells using 1 mg\kg once a day for 6 months

1) Surgical therapy

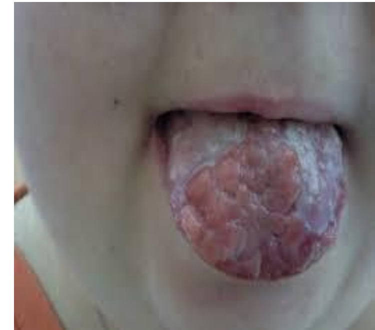


Surgical excision :For lesions that **not respond** to **medical therapy**

Staged resection

- Used for labial hemangiomas because they cause distortion in three dimentions
- The mass debulked by excision of mucosal and submucosal tissues , later excision at the vermilion cutaneous junction

Vascular malforamtions:

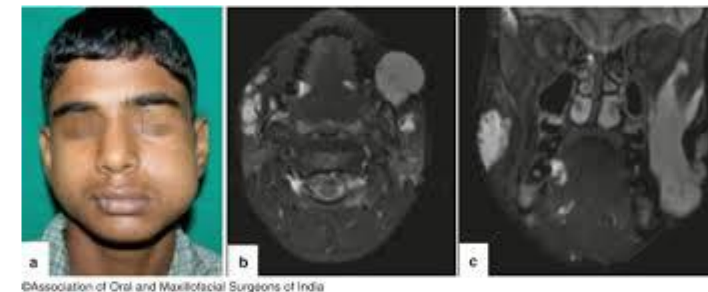


The first step in the care of the patient with VM is to determine whether it is slow or high flow lesion

Complete resection is not possible in most cases

Slow flow lesion without intrabody involvement **cause secondary bony distortion** (bony overgrowth)

Fast flow lesions usually produce bony destruction



Indications for treatment

- Increasing in size and swelling, Pain, Bleeding, Infection
- Macroglossia, Feeding difficulties, Bone destruction, Airway obstruction

Capillary malformation

They appear small pink spot formerly termed port wine stain or nevus flammeus

They especially spread along the trigeminal nerve

Associated syndromes

sturge-weber syndrome

klippel-trenaunay syndrome

parkes-Weber syndrome (proteus syndrome).

Treatment options

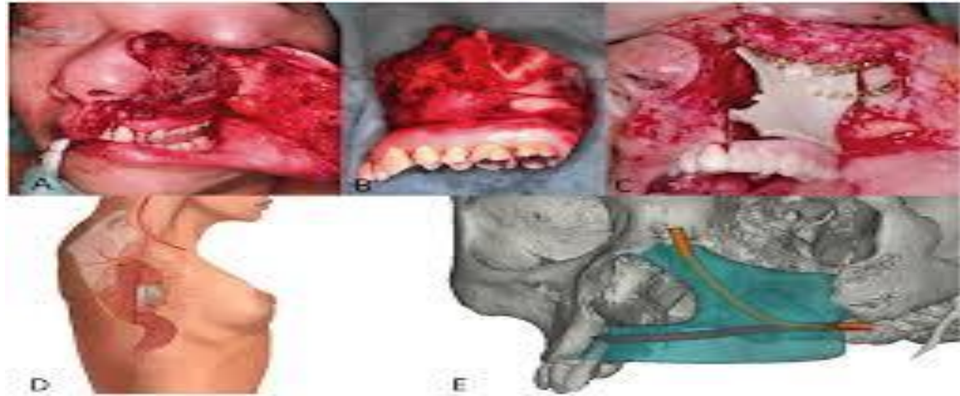
1-Pulsed dye laser

Uses a concentrated beam light that destroying the blood vessels without damaging the skin

2-Excisoin and skin graft or flap

Uses in cases when the skin is very thick and the laser unsuccessful

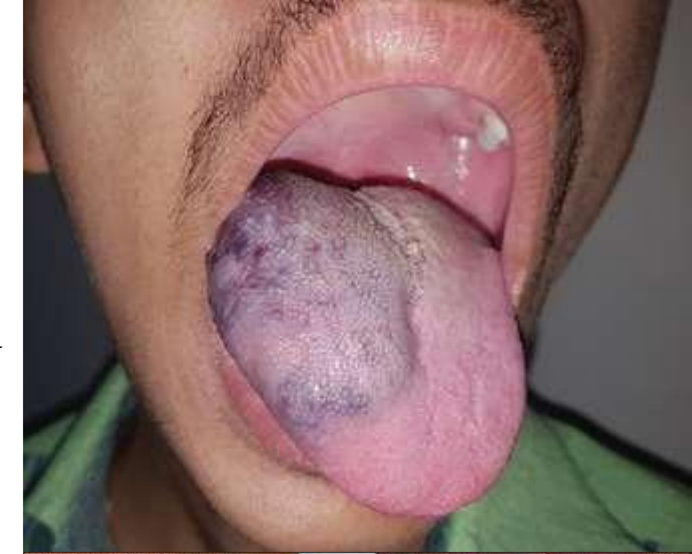
3-Orthognathic surgery -In patients with maxillary or mandibular overgrowth



Venous malformation

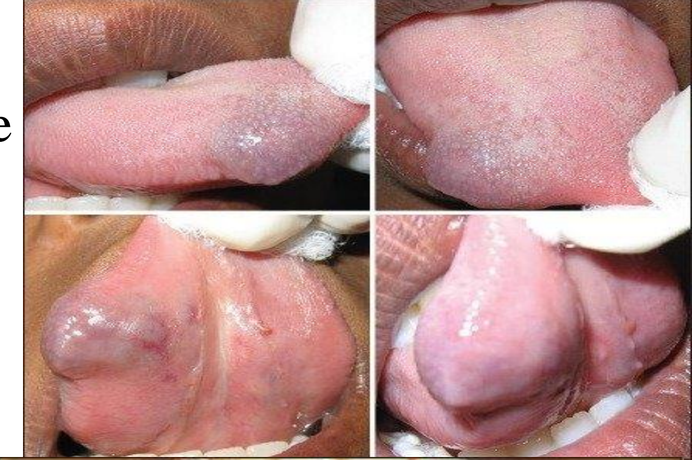
The most common type of vascular malformation

Increasing in size when venous pressure is elevated or during crying
Characterized by presence of phleboliths (result from gradual calcification in of clotted blood in venous vessles



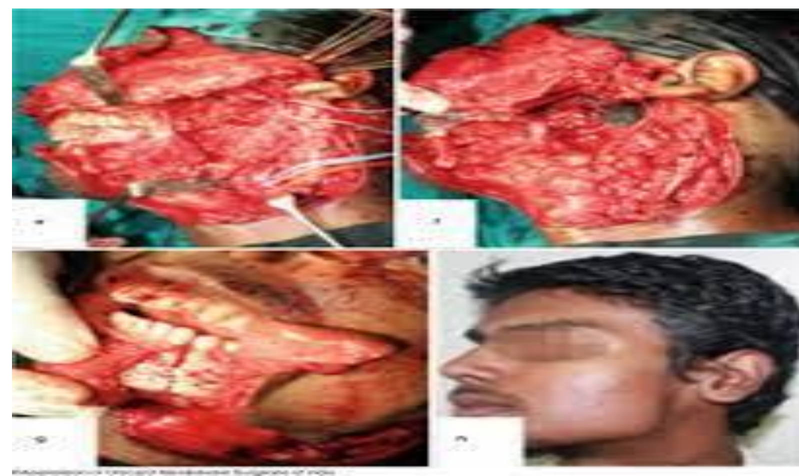
Treatment options

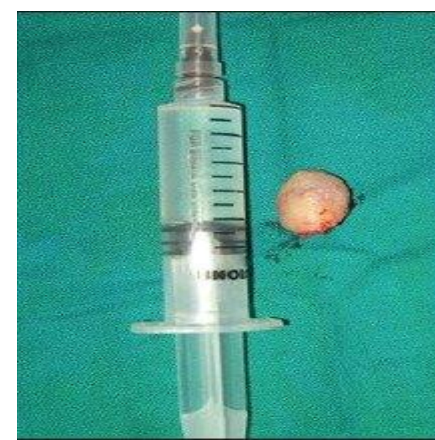
A-Sclerotherapy using Ethanol 100% (scelrozing agent of choice)
Produce endothelial damage that result in permanent endofibrosis and obliteration of the vessels

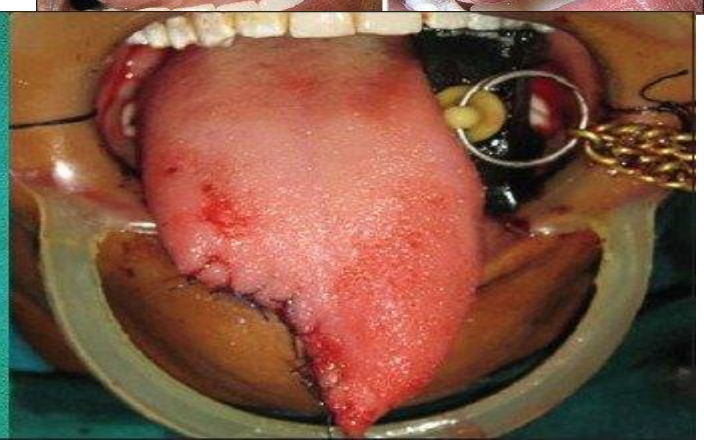


B-Excision for –well-localized cutaneous lesion

C-Resection for marginal or segmental resection for well localized bones lesions







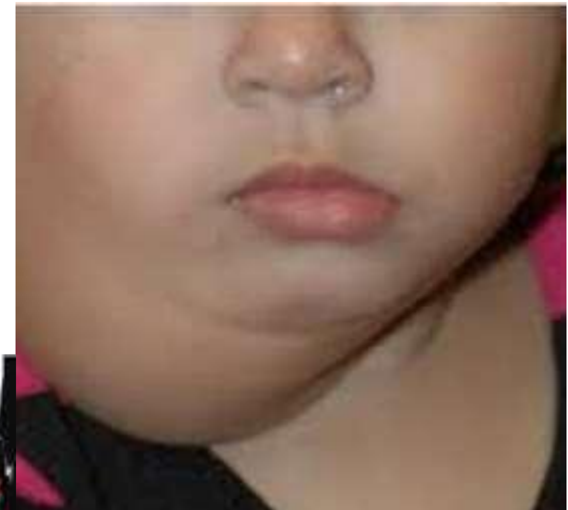
Lymphatic malformations

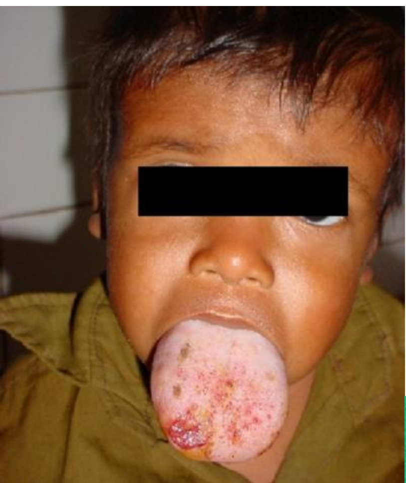
Lymphangiomas, or lymphatic malformations (LM), are benign malformations of the lymphatic system characterized by abnormal proliferation of lymphatic vessels. It was first described by Virchow in 1854. They occur rarely in the oral cavity and involve the tongue dorsum more often.Classified as **macro cystic** and **microcystic**

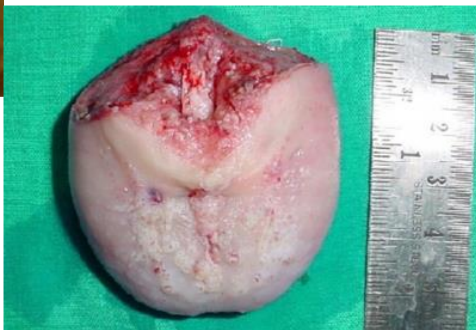
Macrocystic called **cystic hygroma** and occur **below the mylohyoid muscle** in the neck most freguently in front of

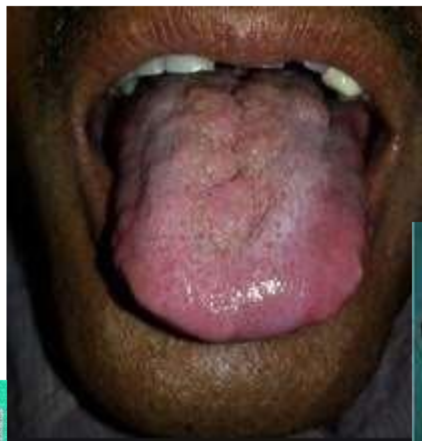
Sternocleidomastoid is the most superficial and largest muscle in the front portion of the neck(**SCM muscle)**

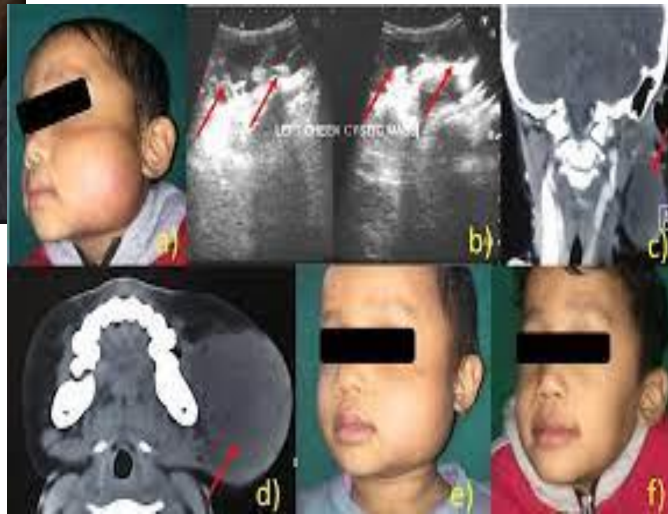
Microcystic commonly involves the tongue , cheek and lips.





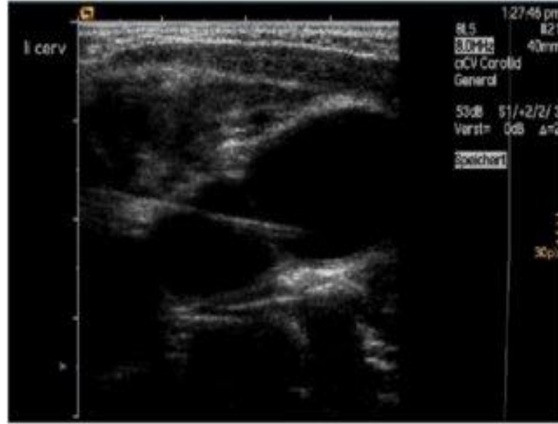




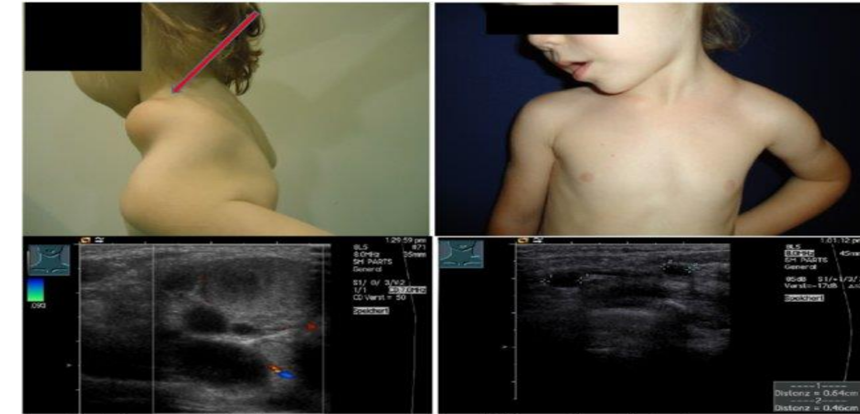


Treatment options:

1-Sclerotherapy -Cause obliteration of the lymphatic vessels associated with a local fibrosis using **Picinable (OK-432 most commonly used**

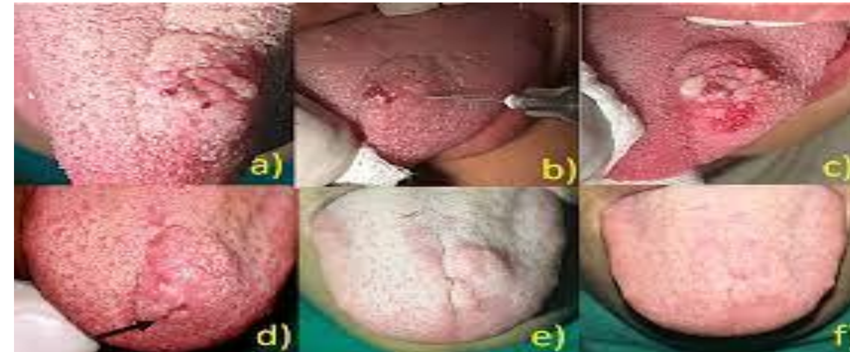

(a)

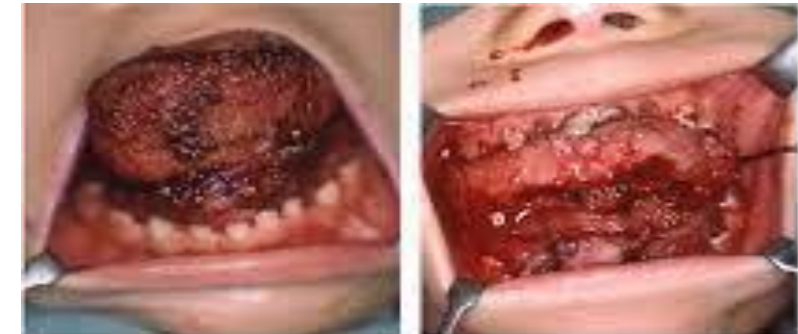
(b)


(a) 4 days post-injection *(b)* 2 month post-injection

2) Excision

- For well localized lesion





3) Staged surgical excision for large lesion

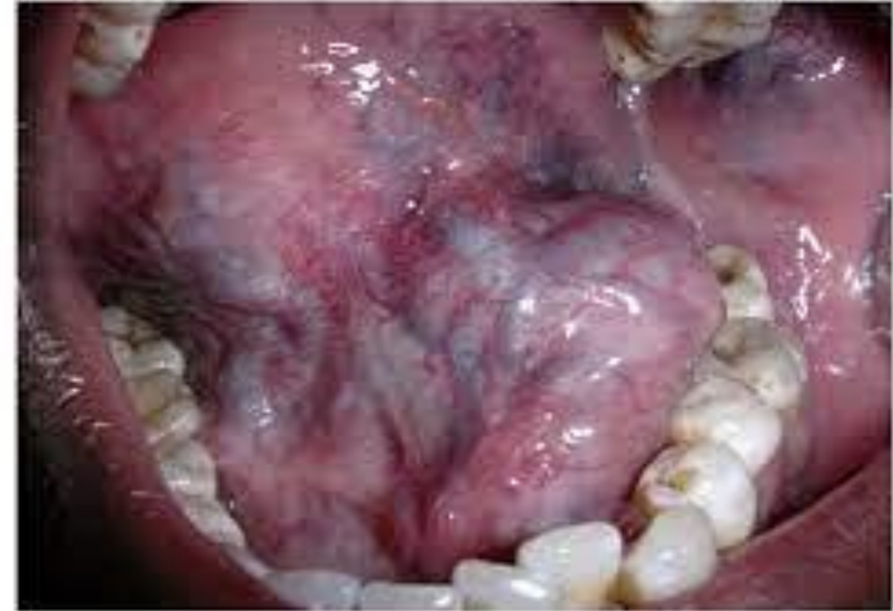
4) Orthognathic surgey

-After soft tissue excision to minimize recurrence and manage malocclusoin

Arteriovenous malformation:

Arteriovenous malformations (AVMs) are rare congenital vascular malformations accounting only 1.5% of all vascular anomalies with 50% occurrence in the oral and maxillofacial region. It usually results from birth defects of the vasculatureThe most dengerous subtypes of vascular maloformations

- Consist of nidus that is composed of abnormal capillary system
- Appears as a red tumor with elevated temperature and typical palpable pulsation
- Associated with arterial steal syndrome
- Intraosseous lesions mostly malformation not hemangioma

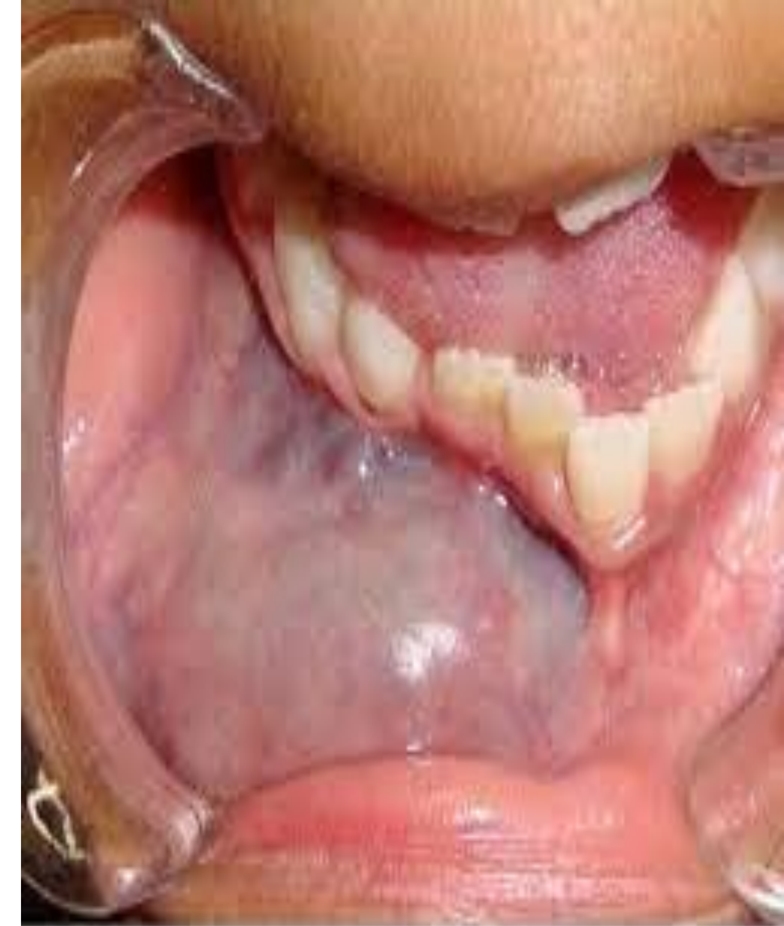


Staging of arterio-venous malformations Schobinger Staging of AVM

- Stage 1
- Stage2
- Stage 3
- Stage 4
- Clinical examination and imaging are crucial in the diagnosis of AVMs. Imaging,
- either contrastenhanced computed tomography (CT) or MRI will distinguish

- Arteriovenous Malformations

- Clinical examination and imaging are crucial in the diagnosis of AVMs. Imaging,
- either contrastenhanced computed tomography (CT) or MRI will distinguish
- between VMs and vascular tumors. The VM will lack a discrete soft tissue mass
- and will have enlarged feeding and draining vessels. Areas of thrombosis or
- intralesional hemorrhage may be found within the VM as well. Magnetic
- resonance angiogram and/or CT angiogram are essential for evaluation of the flow
- dynamics, examination of the detailed vascular anatomy, determination of extent
- of the lesion, and treatment planning. Classically, one or most commonly multiple
- hypertrophied arterial feeders will be identified shunting rapidly into the enlarged
- draining venous system across a nidus without a normal intervening capillary
- network.
- Management of Arteriovenous Malformations
- Management of AVMs consists of surgical extirpation, embolization alone, and
- embolization followed by surgical resectionpensation) :lesions producing heart failure



THANK YOU