VASCULAR ANOMALIES OF THE FACE

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Vascular Anomalies

are the

abnormal formation or development

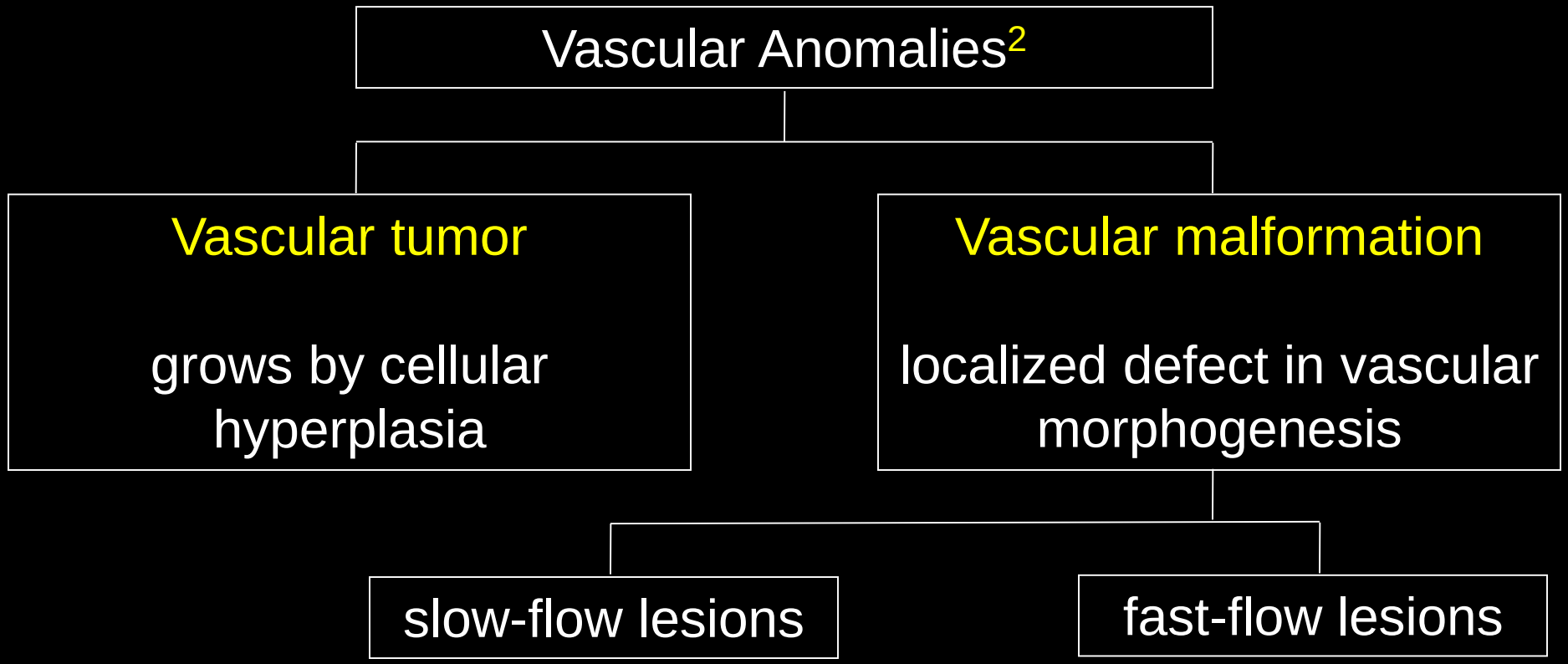
of blood vessels

affecting capillaries, arteries, veins and lymphatic channels

Vascular anomalies are localized defects of vascular development

Etiology

Vascular anomalies are histopathologically characterized by a focal increase in the number of vessels that are abnormally tortuous and enlarged[1].



1. Boon LM, Ballieux F, Vikkula M. Pathogenesis of Vascular Anomalies. Clin Plast Surg. 2011 Jan 1; 38(1): 7–19
2. Richter GT, Friedman AB. Hemangiomas and Vascular Malformations: Current Theory and Management. Int J Ped. 2012, Article ID 645678, 10 pages

Classification

ISSVA classification for vascular anomalies

Vascular Anomalies							
Vascular Tumors	**Vascular Malformations**						
	Simple	**Combined**			**Anomalies of major named vessels**		**Associated with other anomalies**
Benign Locally Aggressive or Border line Malignant	Capillary m. (CM) Lymphatic m. (LM) Venous m. (VM) Arteriovenous m. (AVM) Arteiovenous Fistula (AVF)	CM+VM	capillary-venous m.	CVM	Affect	lymphatics veins arteries	Klippel-Trenaunay syndrome Parkes Weber syndrome Servelle-Martorell syndrome Sturge-Weber syndrome Maffucci syndrome Macrocephaly Microcephaly CLOVES syndrome Proteus syndrome Bannayan-Riley-Ruvalcaba syndrome
		CM+LM	capillary-lymphatic m.	CLM			
		CM+AVM	capillary-arteriovenous m.	CAVM			
		LM+VM	lymphatic-venous m.	LVM	Anomalies of	origin course number length diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm) valves communication (AVF) persistence (of embryonal vessel)	
		CM+LM+ VM	capillary-lymphatic-venous m.	CLVM			
		CM+LM+ AVM	capillary-lymphatic-arteriovenous m.	CLAVM			
		CM+VM+ AVM	capillary-venous-arteriovenous m.	CVAVM			
		CM+LM+ VM+ AVM	capillary-lymphatic-venous-arteriovenous m.	CLVAVM			

ISSVA classification for vascular anomalies
(Approved at the 20th ISSVA Workshop, Melbourne, April 2014)

ISSVA binary classification for vascular anomalies

Vascular Anomalies		
Vascular Tumors	**Vascular Malformations**	
Hemangioma Hemangioendothelioma Angiosarcoma	Slow Flow	Capillary Lymphatic Venous
	Fast Flow	Arterial
	Combined	

Mulliken JB, Burrows PE, Fishman SJ. Vascular Anomalies. Hemangiomas and Malformations. Second Edition

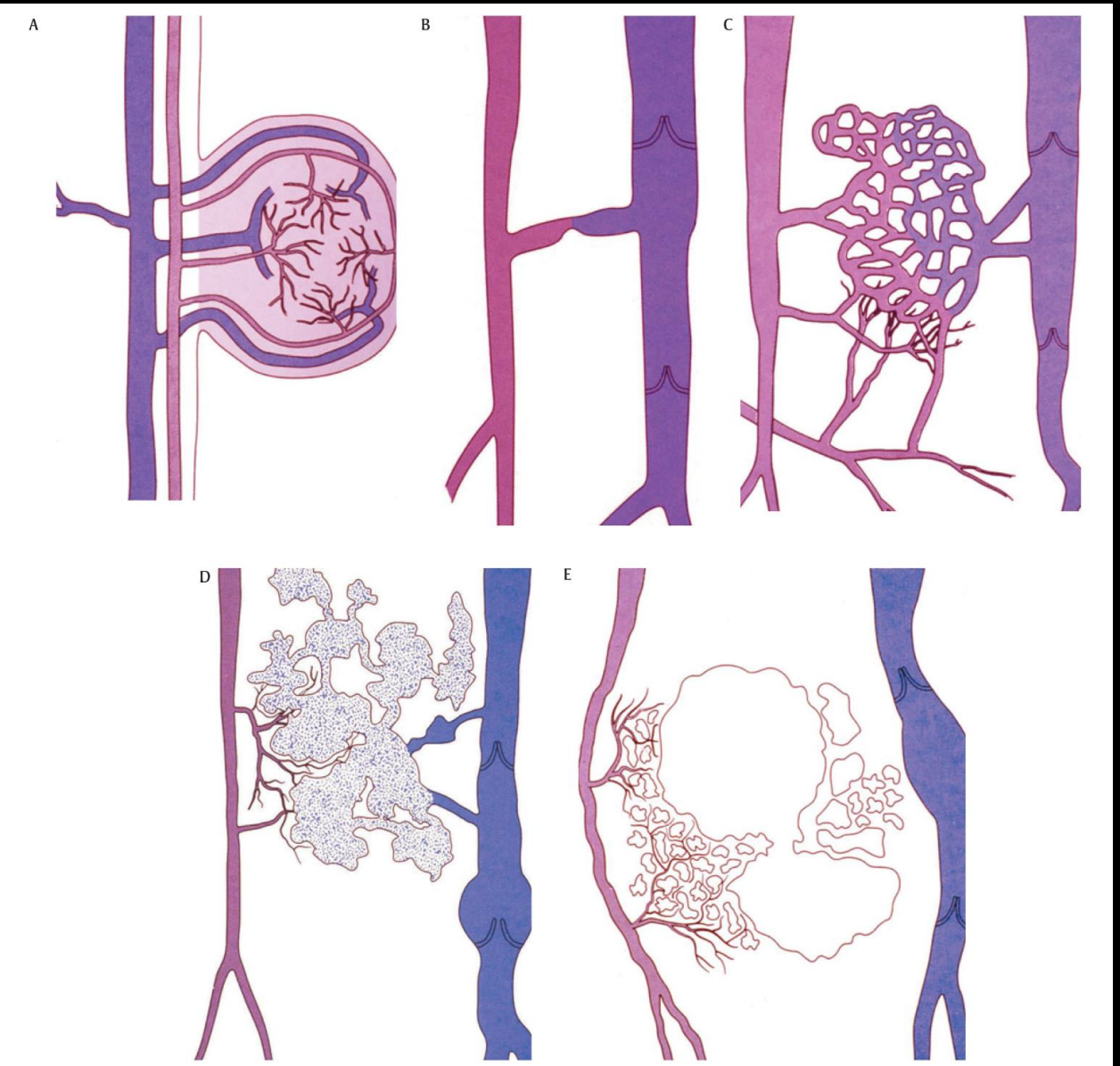


FIGURE 11-1 Diagrams illustrating the channel morphology of the most common forms of vascular anomalies. (Reprinted, with permission, Burrows and Fellows, 1995) A. Infantile hemangioma consists of a solid cellular mass with organized, acinar pattern of arterial supply and drainage into dilated regional veins. B. Arteriovenous fistula is focal macroscopic connection between artery and vein. C. Arteriovenous malformation typically consists of a nidus or network of abnormal vascular channels with feeding arteries and draining veins. D. Venous malformation is a post-capillary lesion composed of abnormally shaped, dilated venous channels. Major conducting veins can be involved. E. Lymphatic malformation composed of fluid-filled spaces or channels lined with lymphatic endothelium.

Mulliken JB, Burrows PE, Fishman SJ.
Vascular Anomalies. Hemangiomas and Malformations. Second Edition

Clinical Manifestation
Vascular Tumors

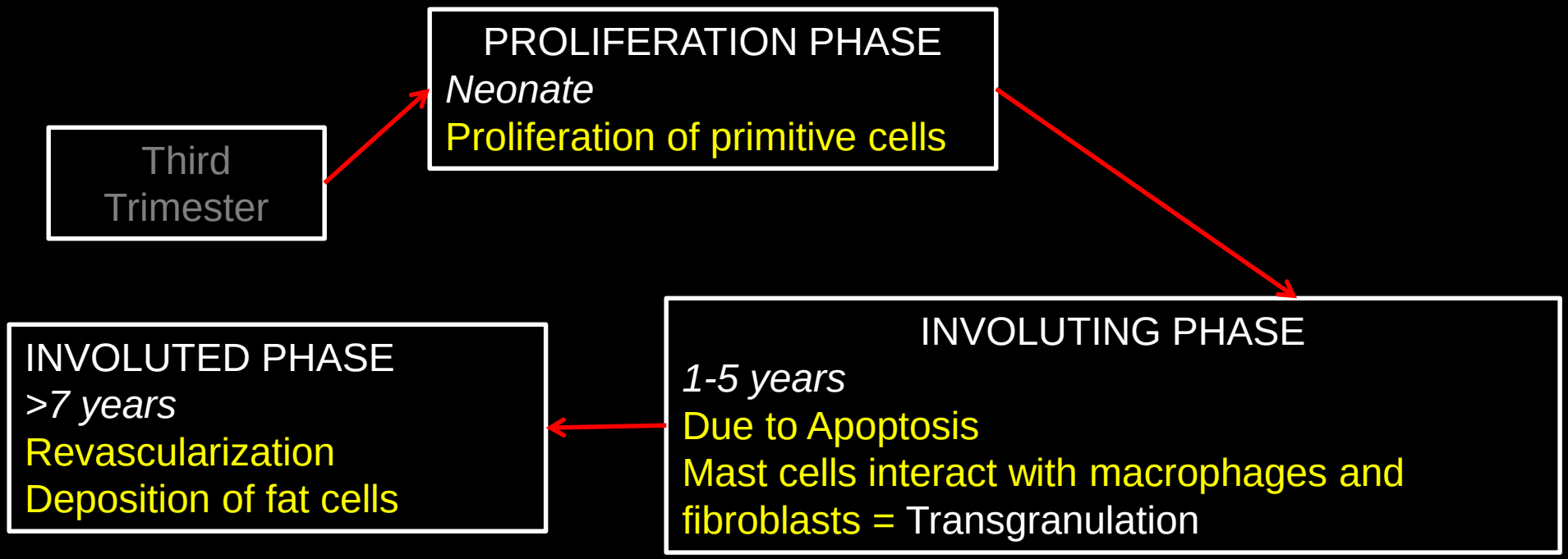
Infantile Hemangioma

Benign vascular neoplasms

Have a characteristic clinical course marked by

early proliferation and

followed by spontaneous involution.



Cutaneous Hemangioma
small/large/extensive
Ulcerative/non-ulcerative

Deep Hemangioma

Involuting Hemangioma

Involuted Hemangioma

Clinical Manifestation
Vascular Malformations

Capillary Malformations

- Caused by a defect of autonomic nervous system supplying capillaries
- The number of blood vessels are normal, but the diameter of the affected vessels is much larger.
- This enlargement results in increased blood flow.
- Since the vessels are close to the surface, this increased flow gives the skin its pink to purple appearance.
- The affected blood vessels will continue to enlarge and thicken with age, causing the color of the lesion to darken.

Venous Malformations

- Made up of malformed veins
- Vary in color from blue to dark purple, depending on how deep the malformation extends.
- Tend to swell with activity/exercise
- The mass is usually soft and compressible and then refills when released.
- There may be small hard masses palpable in the lesion, called phleboliths, which are small collections of calcium that have resulted from slow blood flow and blood clots.

Lymphatic Malformations

- Exact cause is unknown. ? Errors in the formation and development of lymphatics during fetal development.
- Made up of abnormal, dilated lymph channels that can be focal or diffuse.
- Increase in size with infection such as upper respiratory infections
- Difficult to treat if they are diffuse (affecting more than one small area).
- Three types. Micro cystic, Macro cystic and Mixed

Arterio-Venous Malformations (AVM)

- Involve an abnormal connection between arteries and veins
- Consist of a blood vessel “nidus” (nest) through which arteries connect directly to veins, instead of through capillaries.
- Symptoms include throbbing pain and growth/thickness of the area involved.
- Palpation over lesion will reveal a pulsation or thrill.
- If bleeding occurs it can be quite brisk and may require medical attention.

Combined Malformations
Sturge Weber Syndrome

Sturge-Weber syndrome consists of

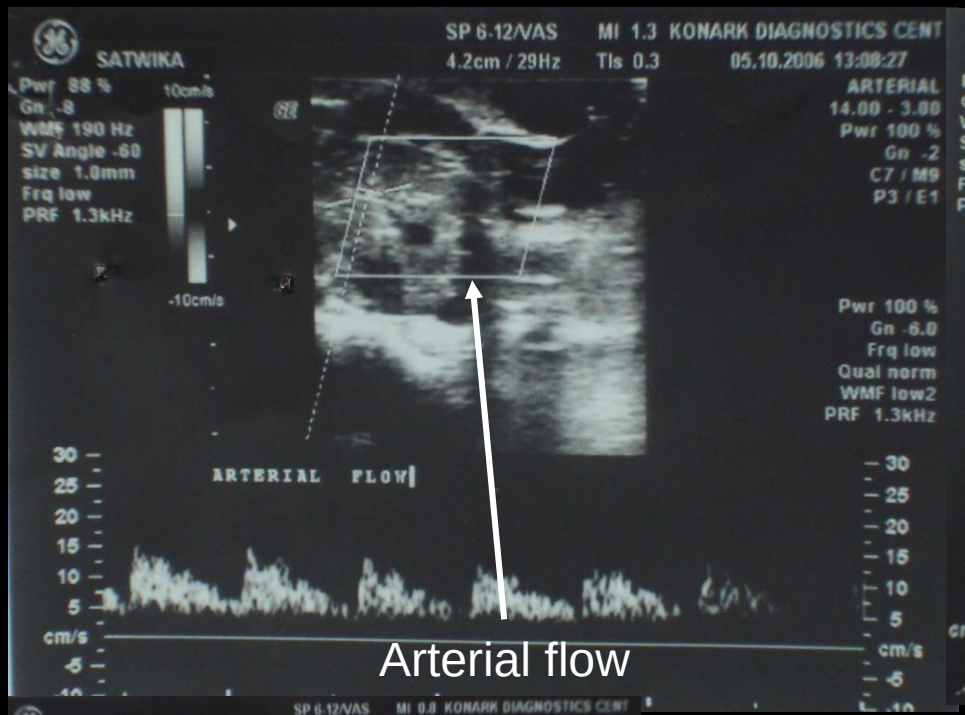
Tortuous slow-flow vessels
involving the conjunctiva, episclera, retina or choroid.

Glaucoma is the most common and serious ophthalmological complication; the prevalence is 60% (Sujansky and Conradi, 1995a).

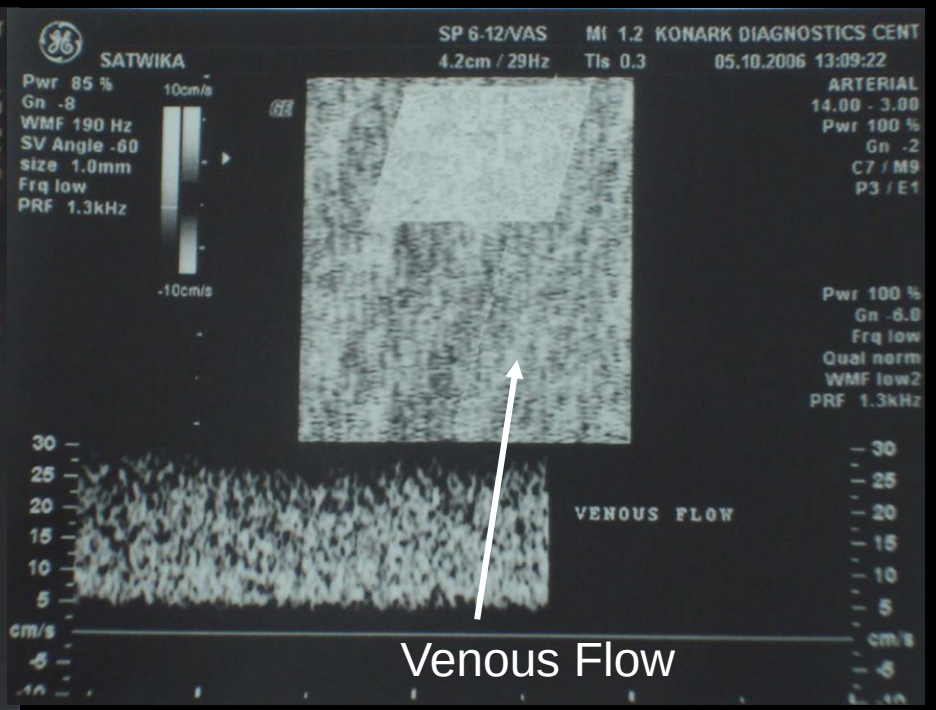
Sudden corneal clouding is the pathognomonic sign of acute glaucoma; this is an emergency.

Diagnosis

Ultrasound…



Arterial flow



Venous Flow

Nidus

Ultrasonography of AV malformation of upper lip
Note the arterial flow, venous flow and nidus of capillaries

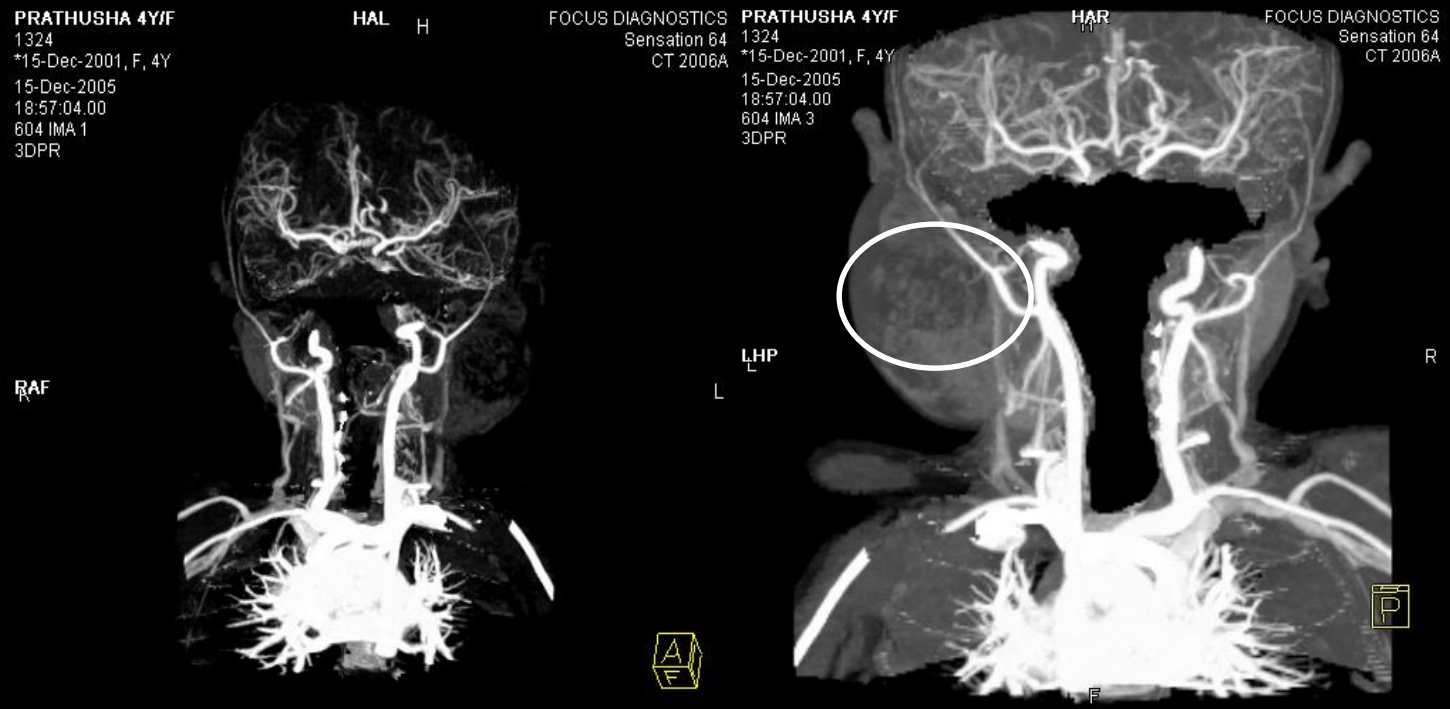
CT Scan

CT scan helps locate the position and extent of the lesion
They also help in identifying bony structures adjacent to the lesion

Coronal CT Scan of A-V malformation of the cheek

Arteriogram

Arteriogram is a CT scan with contrast that offers a clear view of the vessels in the vascular malformation



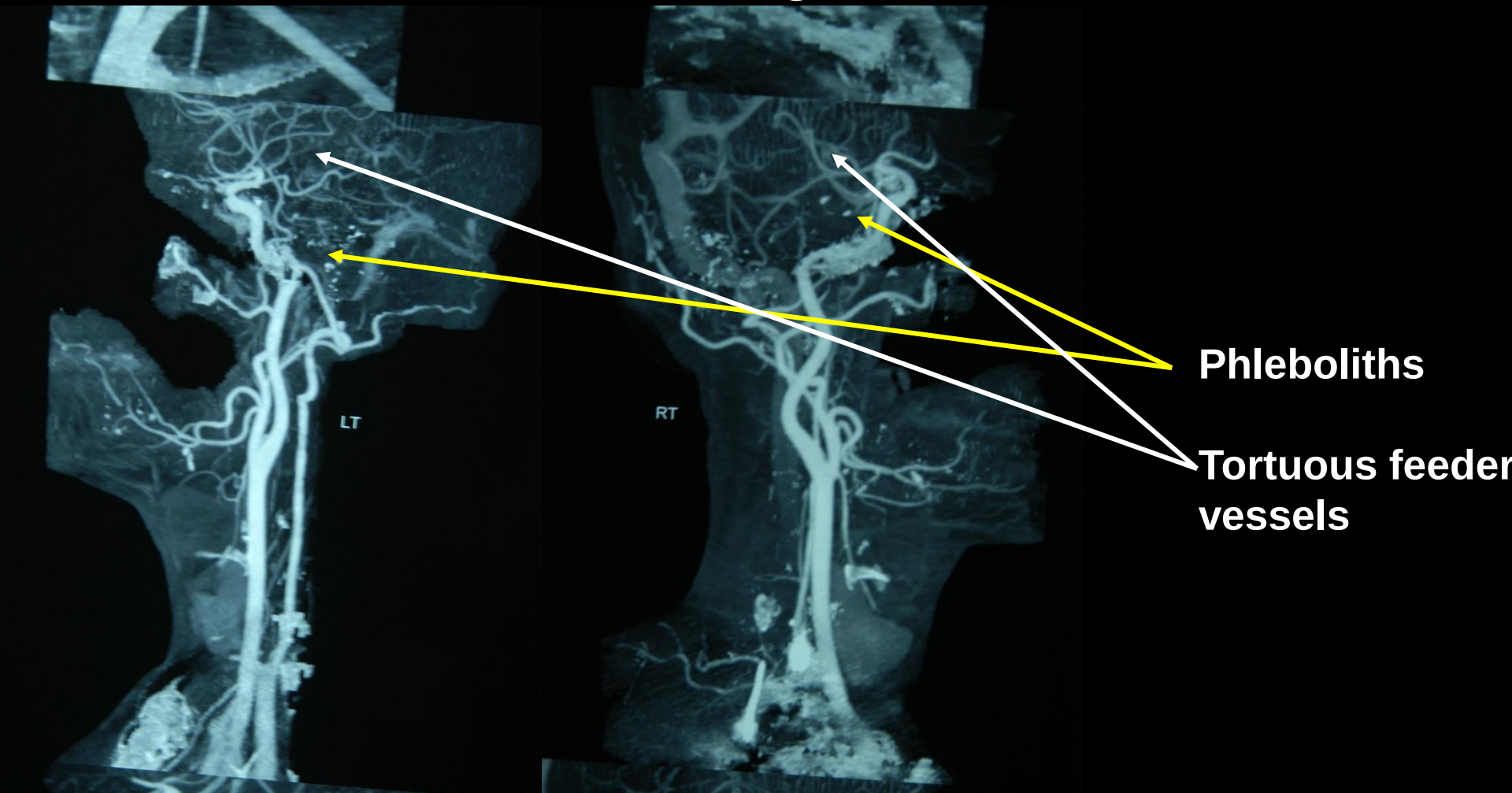
Arteriogram of Lymphangioma of Cheek

Note the absence of feeder vessels

Note the size of the cystic lesion.

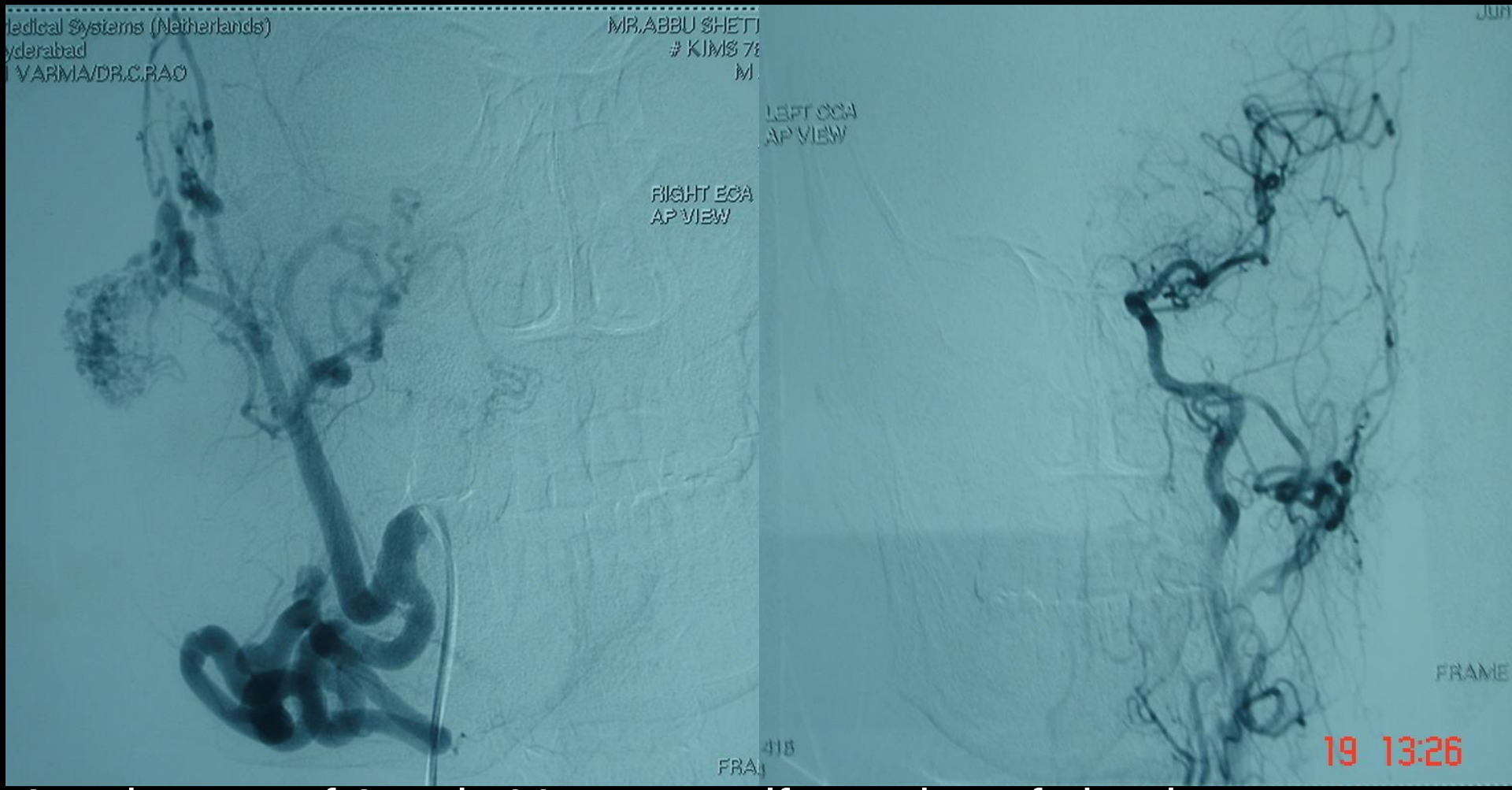
If it is more than 2 cm it is a macrocystic lesion otherwise it is microcystic

Arteriogram



Arteriogram of Venous malformation of cheek
Note the presence of feeder vessels and phleboliths

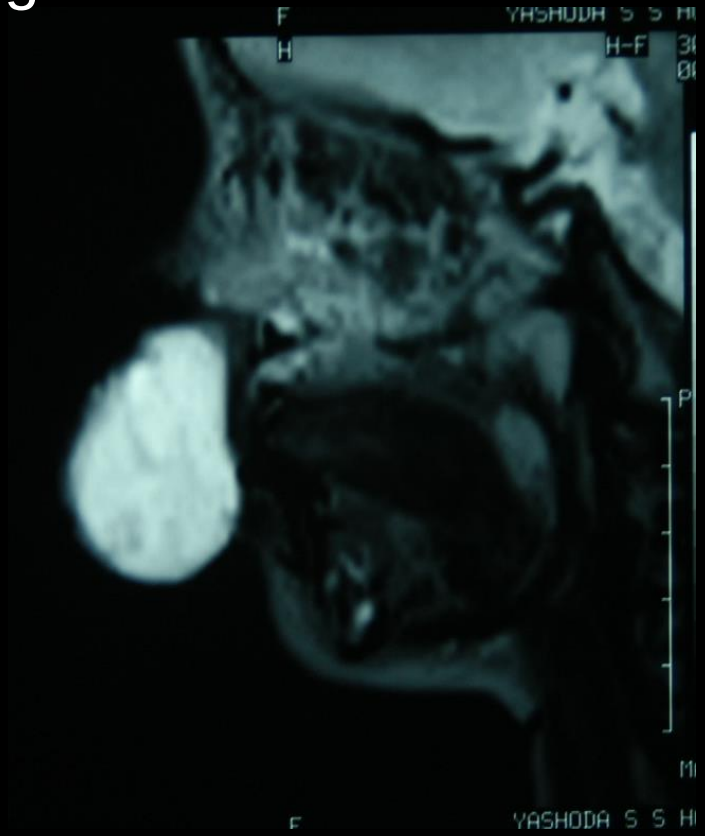
Arteriogram



Arteriogram of Arterio-Venous malformation of cheek
Note the difference between right and left External Carotid Artery

MRI

The MRI helps the surgeon visualize the extent of the lesion in relation to surrounding soft tissue structures



MRI Scan of AV malformation of upper lip

Note the clarity of the extent of the lesion

Treatment

Treatment

Treatment for Vascular Malformations and Hemangiomas is usually as follows

For Low Flow Superficial Lesions
Sclerotherapy followed by
Conventional surgery

For High Flow Lesions
Subtraction angiography with embolization with gel foam or stents followed by
Conventional surgery within 72 hours

In India Angiography is beyond the capacity of most patients. Therefore angiography is considered only if any great vessel is involved.

Cutaneous Hemangiomas

Sclerotherapy

- Syrup or Tablet Propranolol: 0.5 -1 mg/kg of body weight in two divided doses for 6 months under strict pediatric supervision

  (***Propranolol***, β-blocker, vascoconstrictor, regulating angiogenic pathways inducing apoptosis of vascularized endothelial cells)
- Injection Triamcinolone (Kenocort): One 20 mg /ml vial diluted in 2 ml saline and 1ml lignocaine injected intralesionally, once a month for six months.

  (***Triamcinolone***, corticosteroid suppresses vasculogenic capability of multipotent stem cells)
- Contractubex (10% aqueous onion extract, 50 U heparin per gram of gel,1% allantoin) gel and olive oil: massage on the lesion twice daily till the regression of the lesion.

All Vascular Malformations and Hemangiomas

Bleomycin Treatment

- Pingyangmycin (Bleomycin A5) : 2-6 ml (0.5 -4 mg/ml concentration) given intralesionally and repeated every 4 weeks for a maximum of 12 sessions. **OR**
- Bleomycin: 0.5 – 1.0mg/kg body weight up to a maximum of 6mg (0.5-105 mg/ml concentration) given intralesionally and repeated every 4 weeks for a maximum of 12 sessions.
- Bleomycin acts by producing a sclerosing effect due to its direct action on the endothelial cells of the lesion producing non-specific inflammatory reaction
- Can be given in Capillary, Venous, Arterio-venous and Lymphatic malformation and Hemagiomas.

Surgical Protocol

- Key is Accessibility

  Accessible = Surgery

  Inaccessible = Embolisation and surgery

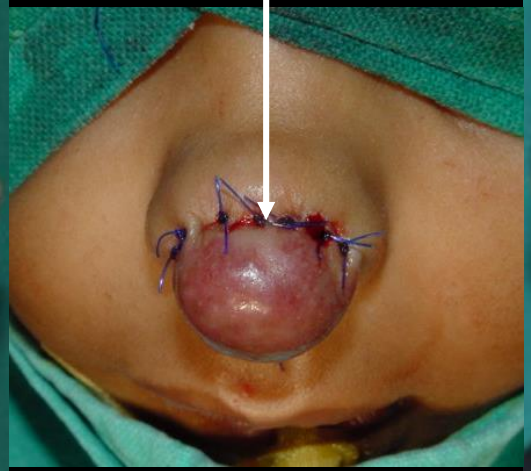
- Ligation of all possible blood vessels in the vicinity of the lesion
- Aim of surgery
  - HARMONIC SCALPEL is used to radically excise all affected tissue as remnants of necrotic tissue can form a focus of a granuloma or further infection.
  - Reconstruct what ever possible
  - Post operative maintenance with steroidal injections intra-lesionally

Harmonic Scalpel

- Cutting instrument that can cut and coagulate tissue simultaneously
- Can cut through thicker tissue and create less toxic surgical smoke than a Bovie
- Offers greater precision than a Bovie
- Cuts via vibration. Bovie cuts via an electrical current (and production of heat)

  Therefore Harmonic Scalpel causes less lateral thermal damage

Treatment…



Capillary Hemangioma

Treatment…

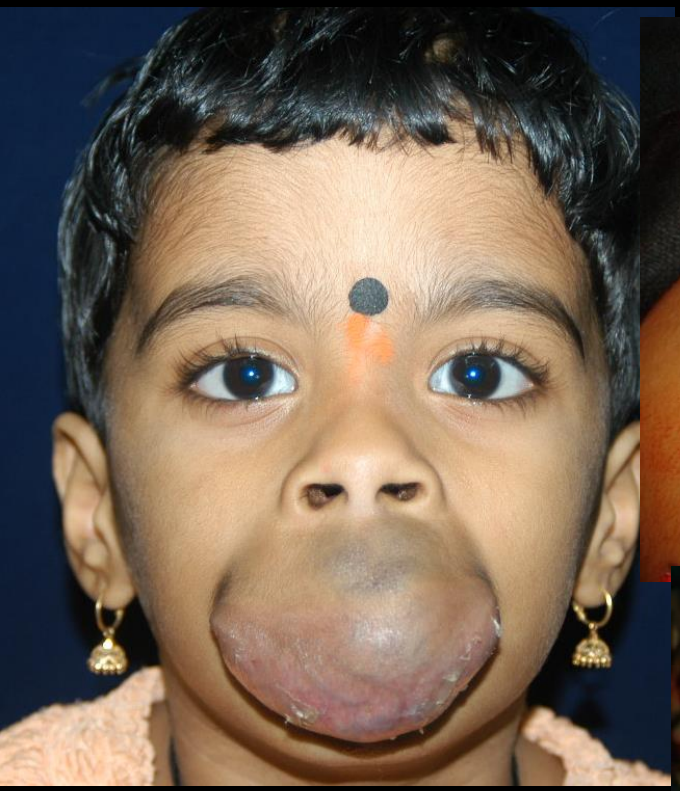
Hypertrophied Capillary Malformation

Treatment with full thickness skin graft harvested from right groin

Treatment…

Low Flow Venous Malformation

Treatment…



High Flow Venous Malformation

Treatment…

Lymphatic Malformation

Surgery is only performed as a cosmetic adjuvant to other therapies.
Macrocystic lymphatic malformations are treated with drainage and ethanol injections as a sclerosing agent.
Microcystic lymphatic malformations are treated with doxycycline injections as sclerosing agent

Treatment…

High Flow A-V Malformation

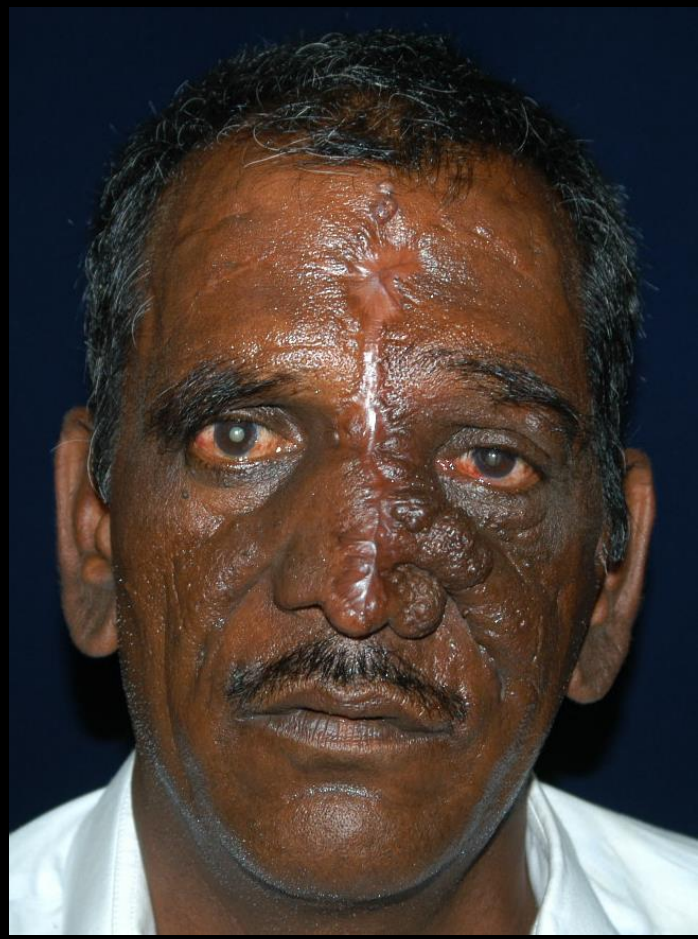
Treatment…

High Flow A-V Malformation

Treatment…

Sturge Weber Syndrome

Treatment…



Sturge Weber Syndrome

Complications

Hemangioma

- Very problematic, interfering with eating, breathing, seeing, hearing, and speaking.

Vascular malformations: capillary, venous and arterio-venous

- Patients with port-wine stains should be evaluated and monitored for a larger syndromic entity.
- Malformations that are part of the Klippel-Trenaunay-Weber syndrome can be located on the lungs, spleen, liver, bladder, or colon. Visceral involvement can often lead to substantial morbidity in the form of internal hemorrhage.

Complications…

Vascular malformations - Lymphatic malformations

- Diffuse cervicofacial disease can result in mandibulomaxillary hypertrophy because of direct invasion of the bone and growth of the malformation within the bone..
- Lymphangiomas often swell with the onset of general viral infection or remote bacterial infection. This typically resolves with the resolution of the infection.
- Lymphangiomas can become infected

Do not confuse a Vascular malformation with…

Treatment…

Carotid Body Tumor

Slowly enlarging(~5mm per year), non-tender neck masses located just anterior to the sternocleidomastoid muscle at the level of the hyoid.

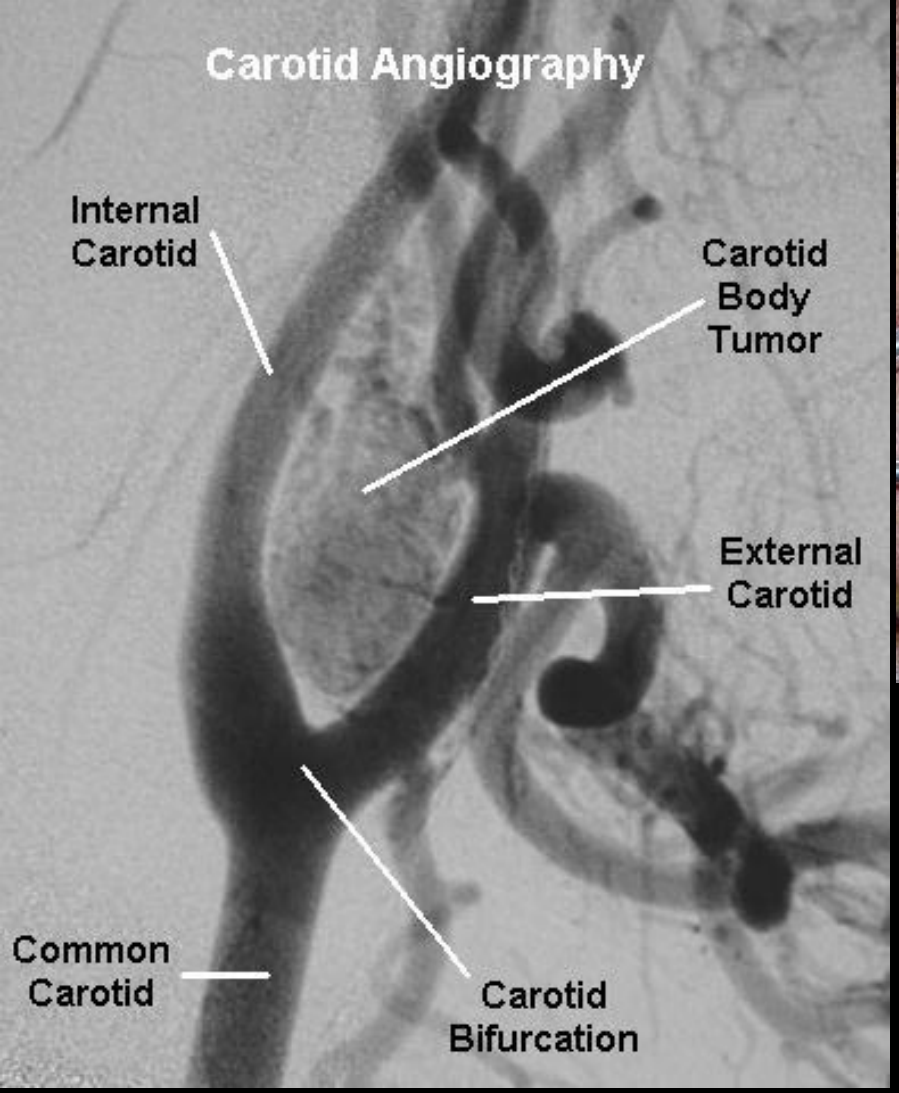
The mass may transmit the carotid pulse or demonstrate a bruit or thrill, which might confuse the clinician to think it is a vascular malformation.

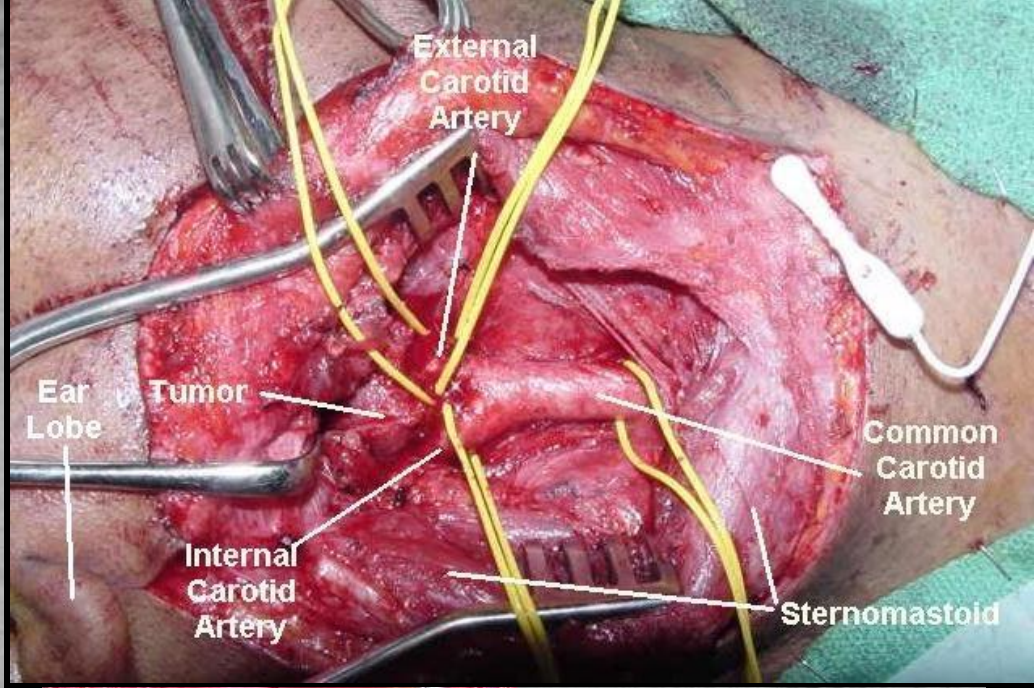
As these tumors enlarge, progressive symptoms of dysphagia, odynophagia, hoarseness and other cranial nerve(IX-XII) deficits appear.

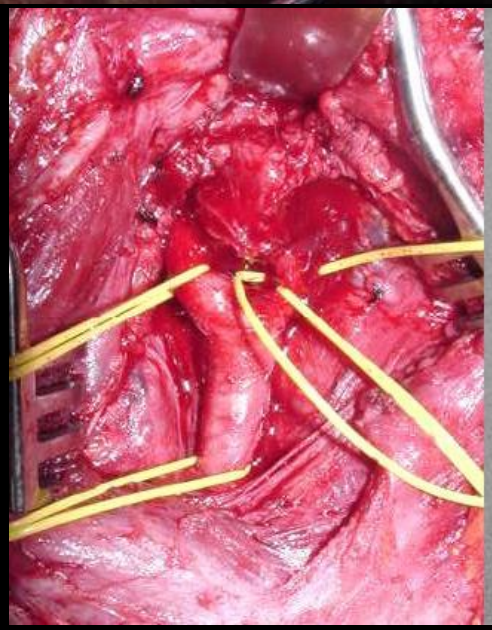
Carotid angiography is by far the most useful diagnostic test for paragangliomas.

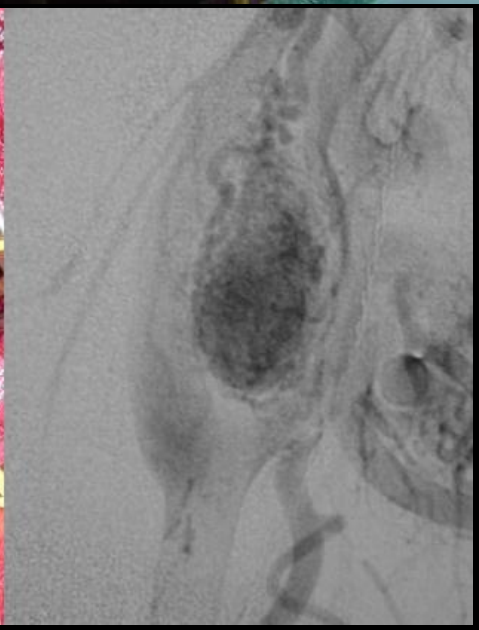
Treatment…

Carotid Body Tumor









Carotid Body Tumor

Vascular Malformations of the head and neck region are something that are treatable in most situations

Care must be taken, however, to do a work up for the patient

Most lesions that you will find in your practice will be low flow lesions

To diagnose the flow of vascular malformation lesions requires nothing more than an ultra sound.

The key to treatment is Accessibility

Bring the Smile Back

Thank You