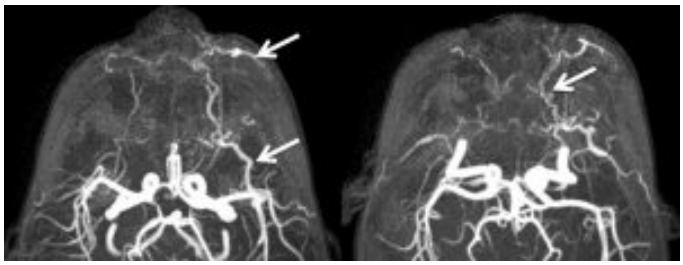
COURSE 03: IMAGING AND DIAGNOSTICS OF HEAD AND NECK VASCULAR ANOMALIES

Dr. Anton Hasso

Advanced imaging techniques have critical value for the diagnosis of vascular soft tissue anomalies.

Initial classification and categorization of vascular tumors and malformations are determined according to morphological appearances and flow dynamics, features best determined with magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA), particularly in the head and neck as lesions may show both extra- and intra-cranial components.

Ultrasound and fluoroscopic techniques are used to determine the location and treatment outcomes in patients undergoing percutaneous sclerotherapy of venous and/or lymphatic malformations.



Objectives

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

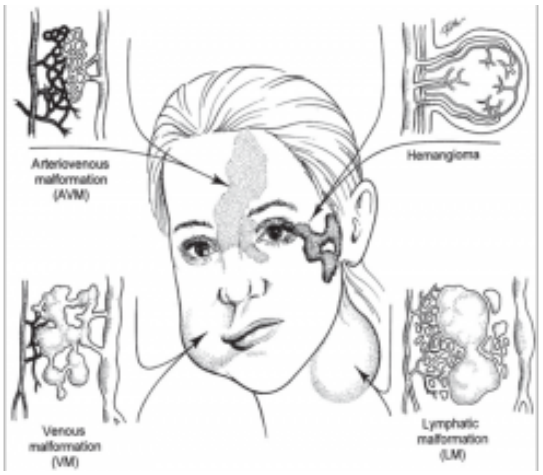
- Describe the application of various imaging modalities for the diagnosis of head and neck vascular anomalies
- Demonstrate impact of the imaging findings on critical features of various types of hemangiomas, lymphatic malformations, venous malformations, and arteriovenous malformations or fistulas
- Explain the impact of imaging on treatment options
- Choose specific treatment options for each type of lesion based on the location and flow-dynamics of each individual vascular anomaly

Imaging and diagnostics of head and neck vascular anomalies

Vascular anomalies can be divided into hemangiomas and vascular malformations based on histological features, natural history, and biological behavior. Hemangiomas are benign endothelial tumors that appear during infancy, undergo a period of rapid proliferation, and eventually regress. Accordingly, hemangiomas can be sub-classified as either proliferating or involuting. Vascular malformations, on the other hand, are developmental defects of the vasculature that are present at birth, grow in parallel with the individual, and never involute. These lesions are sub-divided based on the predominant vascular channel that is affected, namely arterial, venous, capillary, lymphatic, or combined.

MRI and MRA are critical diagnostic techniques in the characterization of vascular anomalies as they can determine hemodynamic flow properties, delineate extent of disease, and guide treatment. The hallmark imaging features of hemangiomas and vascular malformations are closely coupled to clinical decision-making strategies. In this module, we discuss the MRI and MRA findings for various vascular anomalies and discuss strategies for selecting optimal treatments for such lesions in the head and neck. Diagnostic ultrasound and catheter angiographic techniques are demonstrated for the treatment of certain lesions. Imaging and diagnostics of head and neck vascular anomalies

Common vascular soft tissue anomalies



Schematic diagram depicting the morphologic appearance and gross histology of the most common vascular anomalies and vascular tumors of the head and neck. Insets depict the aberrant angioarchitecture manifest by the various lesions.

Common vascular soft tissue anomalies

- Vascular tumors
  - Inantile hemangioma (Phase I, II, III) Congenital hemangioma
  - (RICH and NICH types)
- Superficial malformations – low flow
  - Capilary malformations (CM)
  - Lymphatic malformations (LM)
  - Venous malformations (VM)
- Superficial malformations – high flow
  - Arteriovenous malformations (AVM)
  - Arteriovenous fistula (AVF)

MRI of infantile hemangioma

Proliferative phase: high-flow lesions

- T1-weighted MR findings
  - Flow voids
  - Low signal
  - Intense enhancement post- contrast with persistent irregular flow voids of varying sizes
- T2-weighted MR findings
  - High signal

    Flow voids

Involuting phase: transform to low-flow lesions

- Hypercellularity replaced by fibrous and adipose tissue
- No flow voids
- Much less enhancement post- contrast

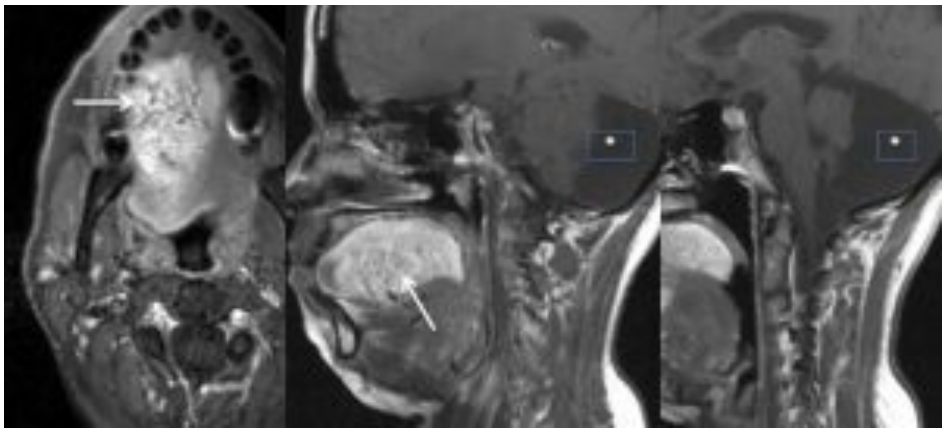
PHACE Syndrome

Upper/lower facial and maxillary/mandibular segmental hemangiomas (see photos) plus 1 or 2 of the following:

- Posterior fossa anomalies, including Dandy-Walker complex, cerebellar hypoplasia, dysgenesis, or agenesis of vermis
- Arterial anomalies, including hypoplasia, agenesis, anomalous branches, aberrant origin or course
- Cardiac defects
- Eye anomalies
- Sternal defects or supra-umbilical raphe

PHACE Syndrome can be diagnosed in adults. An astute radiologist would be the first to notice (often as an incidental finding) cerebellar hypoplasia and/or arteriopathies suggestive of PHACE Syndrome. Further questioning of the patient may elicit a history of segmental facial hemangioma in infancy.

PHACE Syndrome in an adult patient



PHACE Syndrome in an adult patient: MRI with contrast shows persistent infantile hemangioma of tongue (arrows) and cerebellar hypoplasia (*) due to a posterior fossa anomaly.

NICH (Non-Involuting Congenital Hemangioma) and RICH (Rapidly Involuting Congenital Hemangioma)

Congenital hemangiomas

- Hemangiomas that are fully formed and full size at birth, unlike infantile hemangiomas that appear a few weeks after birth.
- Lesions look and behave differently from infantile hemangiomas.

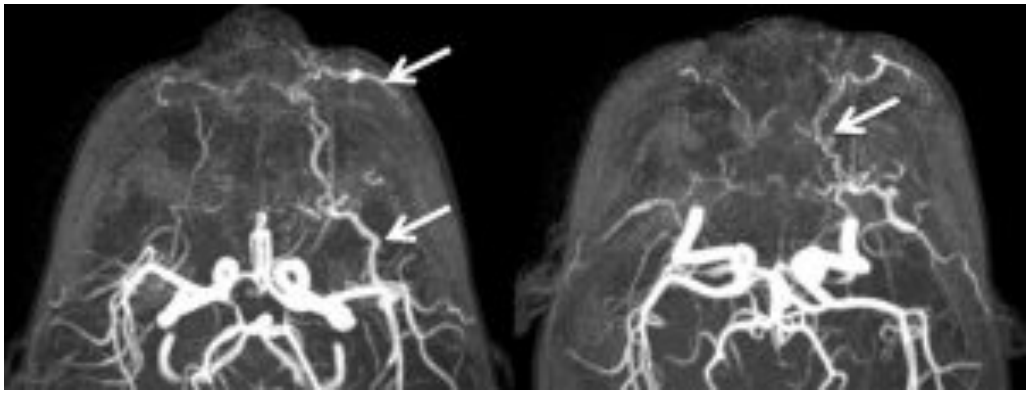
Subtypes:

- Rapidly involuting congenital hemangioma (RICH): not an issue in adults
- Non-involuting congenital hemangioma (NICH): may remain into adult life

MRI and MRA of congenital hemangiomas

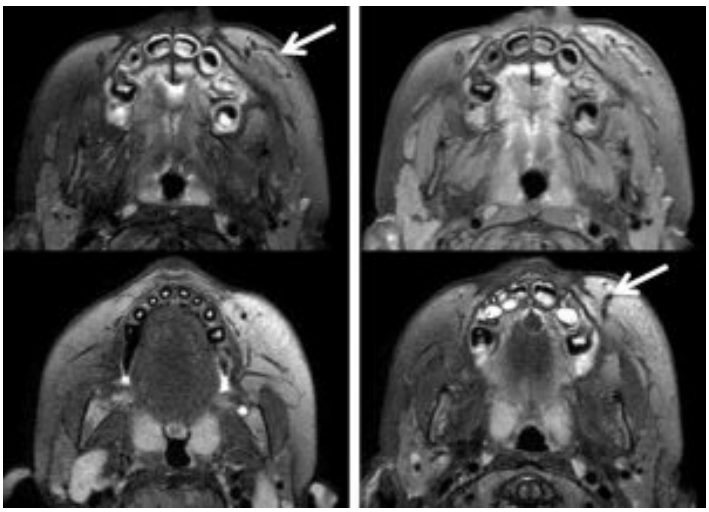
- Lesions are present at birth; subtypes include rapidly involuting congenital hemangioma (RICH) and non-involuting congenital hemangioma (NICH) lesions
- Common findings MRI/MRA:
  - Well organized, single-compartment soft tissue mass
  - Even-sized flow voids throughout the lesions
  - Moderate post-contrast enhancement vis-à-vis infantile hemangiomas that show more intense enhancement
- Lesion may be misdiagnosed as an AVM or AFV on duplex sonography due to high flow rates seen in NICH lesions

Congenital hemangioma of left cheek MRA



Congenital hemangioma of left cheek (NICH lesion): MRA shows anomalous branches of facial artery to lesion (arrows) indicating persistent high-flow vascularity.

Congenital hemangioma of left cheek MRI

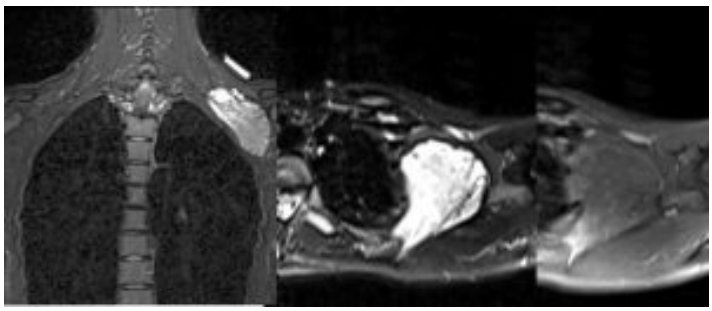


Congenital hemangioma of left cheek (NICH lesion): MRI shows discrete soft tissue mass with few organized flow voids (arrows).

MRI of lymphatic malformations

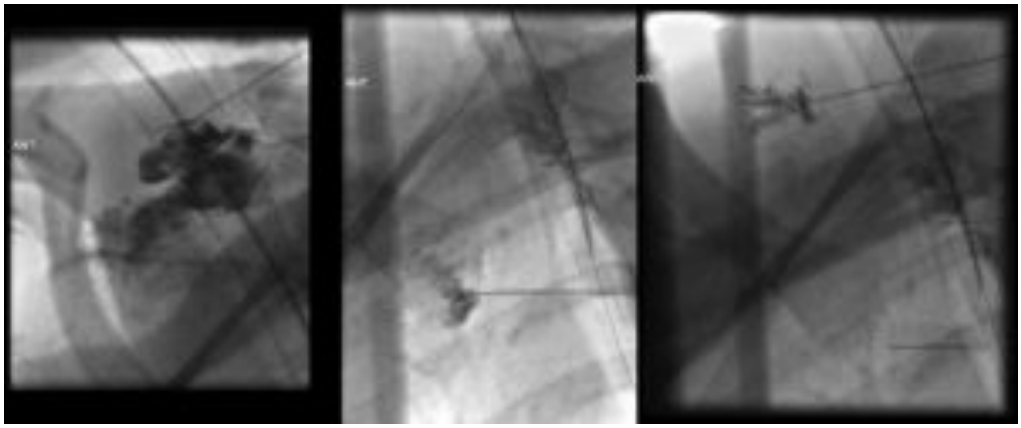
- Description:
  - Multi-loculated
  - Insinuating cross into multiple spatial compartments
  - Cystic: microcystic and macrocystic varieties
  - May show hemorrhage, often with fluid levels
- T1W MR findings: low signal; low flow
- T2W MR findings: low signal; hemorrhage
- Post- contrast T1W MR findings; none typically
  - Possible slight enhancement of rim and/or of internal septations, especially if infected

Lymphatic malformation of the left shoulder MRI



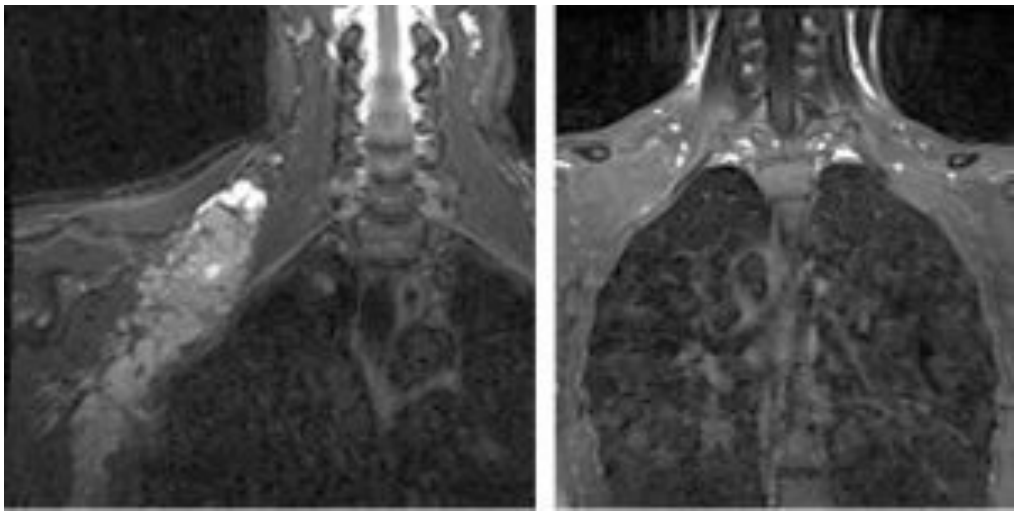
Lymphatic malformation of the left shoulder: MRI without (left 2 images) and with (right image) contrast—show high T2 signal and no enhancement

Lymphatic malformation of the left shoulder sclerotherapy



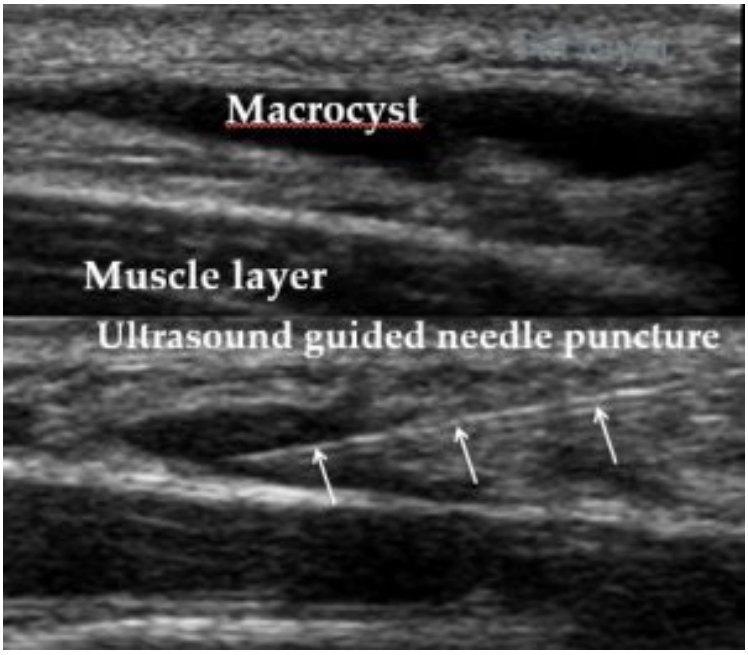
Lymphatic malformation of the left shoulder: sclerotherapy with Sotradecol (sodium tetradecyl sulfate) under fluoroscopic guidance

Macrocystic lymphatic malformation: right neck and shoulder



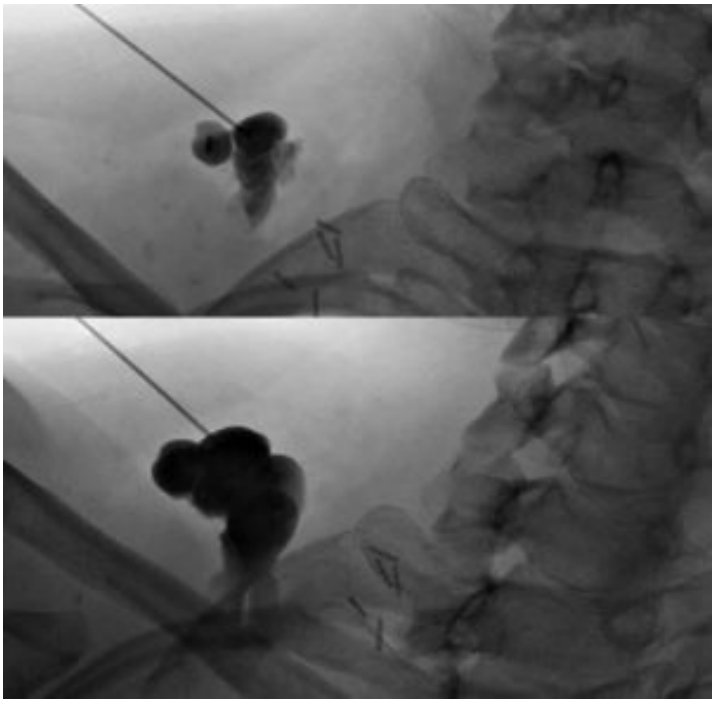
Macrocystic lymphatic malformation: right neck and shoulder: T2 W (left) and post-contrast T1 W (right) MR images

Macrocystic lymphatic malformation ultrasound



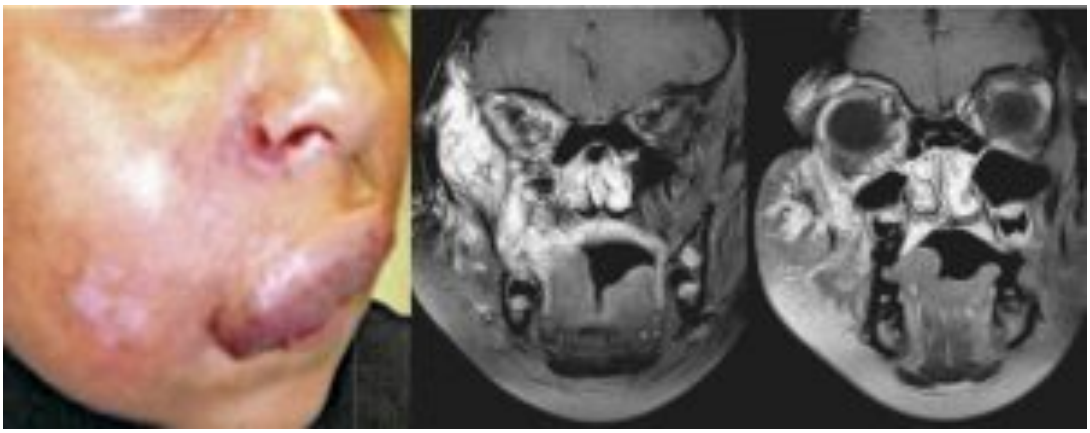
Macrocystic lymphatic malformation: ultrasound-guided needle puncture (arrows) would be followed by schlerotherapy

Macrocystic lymphatic malformation sclerotherapy



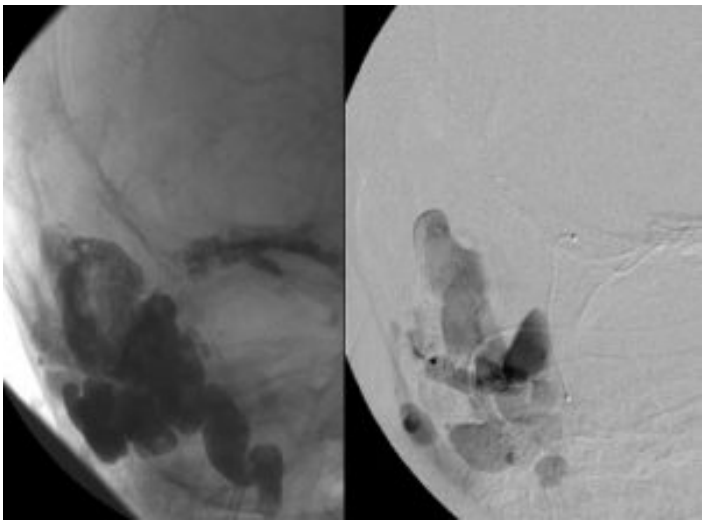
Macrocystic lymphatic malformation sclerotherapy with Sotradecol (sodium tetradecyl sulfate) under fluoroscopic guidance

Segmental venous malformation: pre-treatment



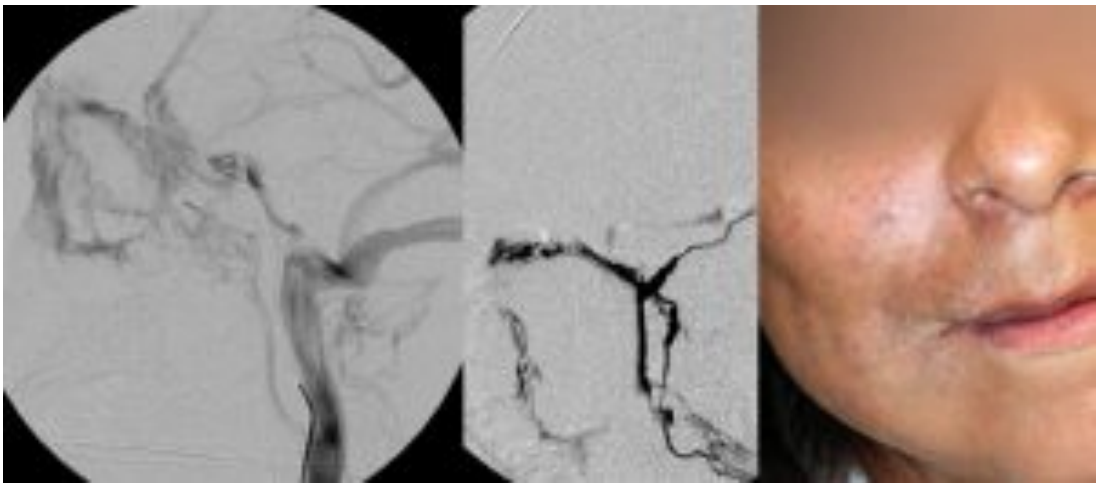
Segmental venous malformation: pre-treatment appearance (left image). MRI T1W fat- saturated, post-contrast showing right facial/periorbital multi-compartmental venous malformation (right images)

Segmental venous malformation: Initial partial treatment



Segmental venous malformation: initial partial treatment under fluoroscopic guidance; images prior (left) and following sclerotherapy (right) with Sotradecol (sodium tetradecyl sulfate) of facial veins

Segmental venous malformation: Percutaneous antegrade venous embolization

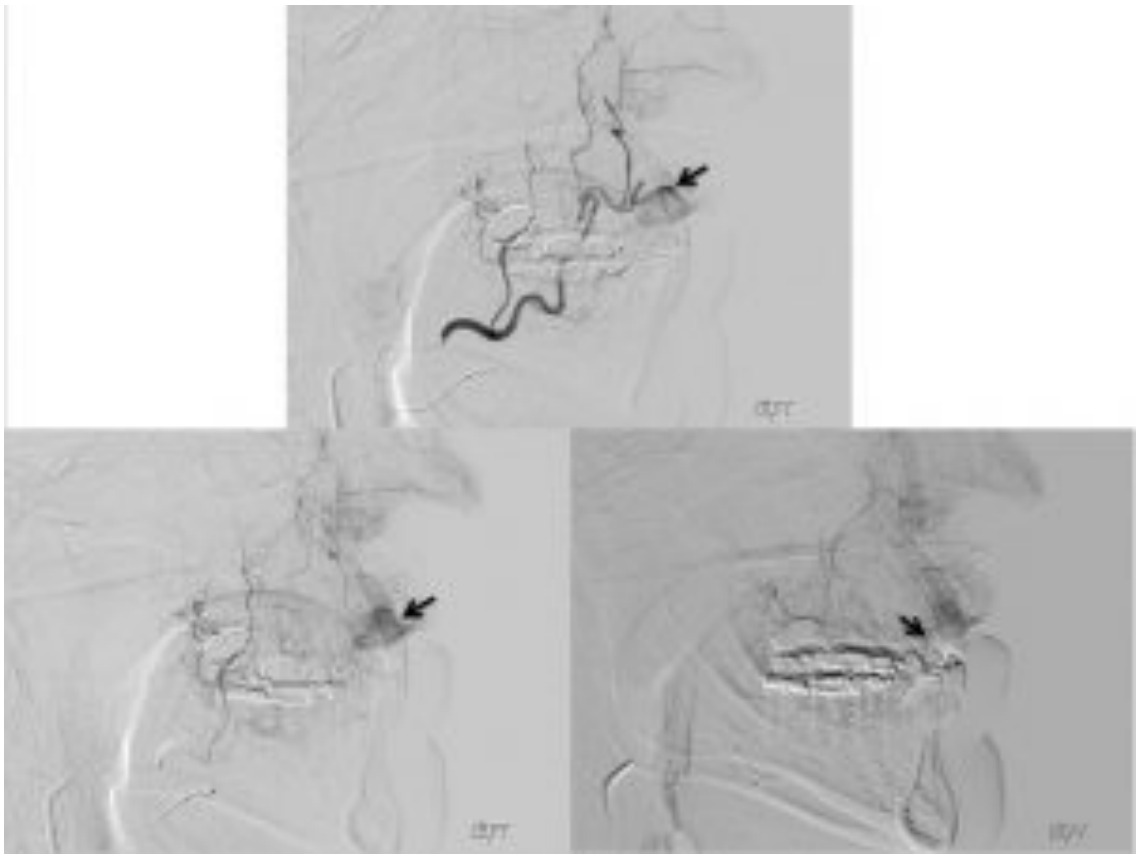


Segmental venous malformation: percutaneous antegrade venous embolization of facial/ periorbital veins (left images; improved appearance 12 months post multiple treatment modalities (right image)

AV malformation of left upper lip

AV malformation of the left upper lip: MRA shows nidus of arteriovenous shunting, drainage in the ranine veins of the tongue go to both sides

AV malformation of left upper lip facial angiogram

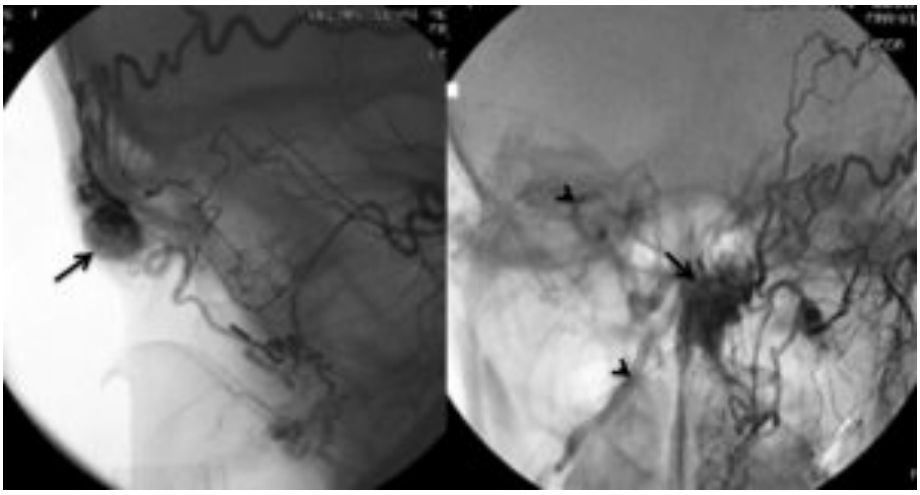


(Same patient from above) AV malformation of the left upper lip demonstrated on selective facial angiogram: superior labial branch of the facial artery (arrow right upper) to the facial vein (arrow right lower) and nidus (arrow below) are well seen

AV malformation mid-forehead lesion

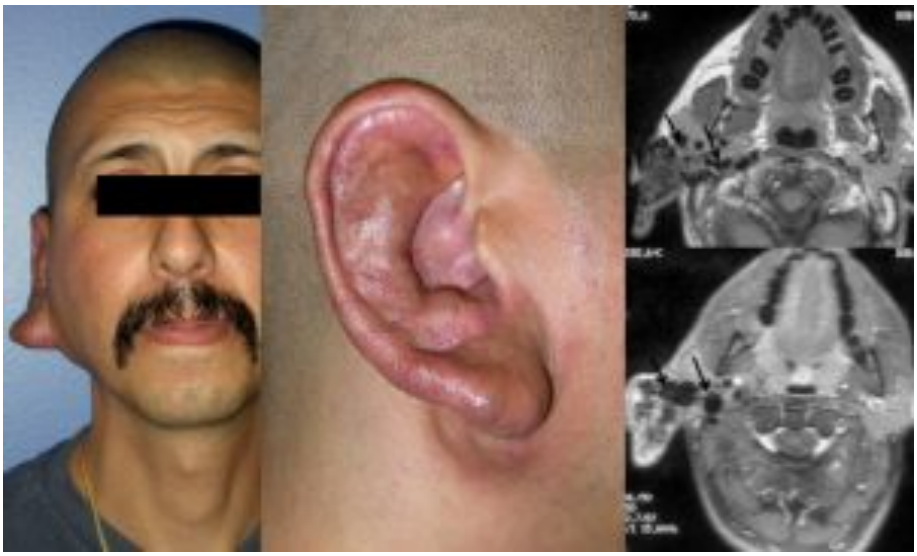
Arteriovenous malformation (AVM): pulsatile mid-forehead lesion

…VM of mid-forehead angiogram



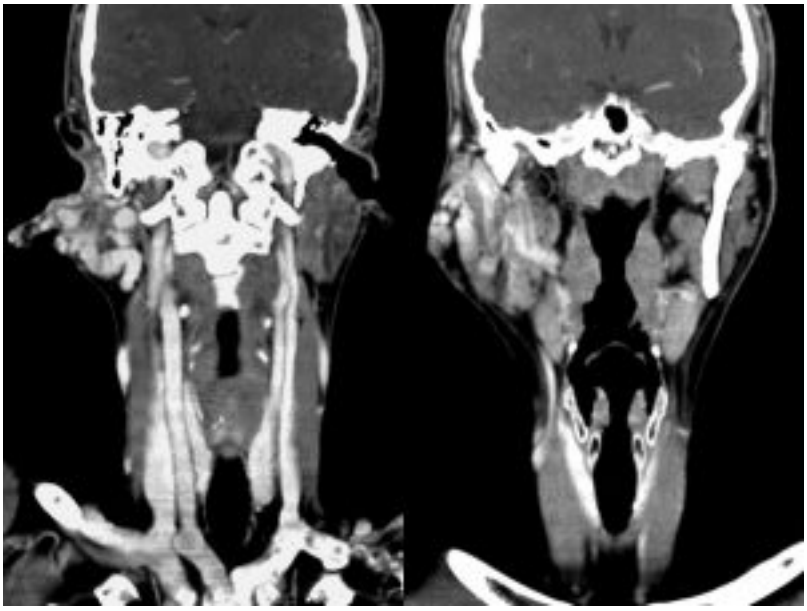
Arteriovenous malformation of mid-forehead: Selective left maxillary artery angiogram shows feeding superficial temporal and facial arteries, central nidus (arrows) with early venous filling to contralateral ophthalmic and facial veins (arrowheads)

AV fistula



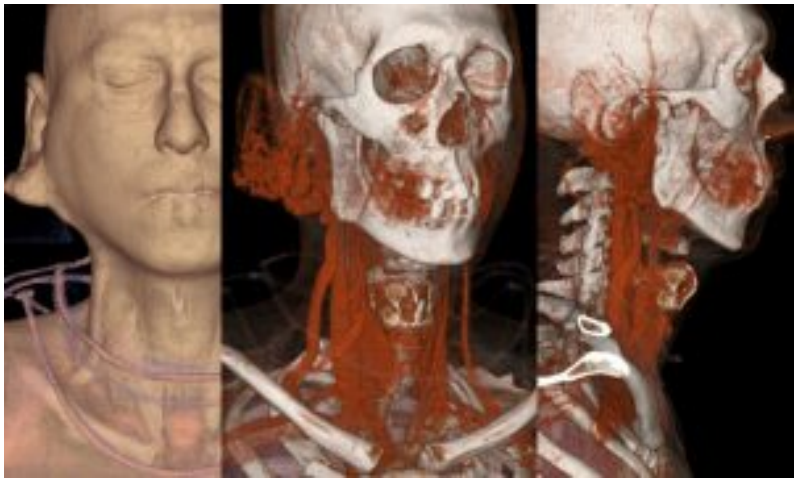
Arteriovenous fistula (CT without contrast (upper right, arrows) and with contrast (lower right, arrows) showing multiple maxillary artery arteries and veins within the right parotid gland and pinna

AV fistula right maxillary artery



Arteriovenous fistula right maxillary artery (CT angiogram, early and late coronal images): large arteries and veins entangled within the right parotid gland and pinna

AV fistula right maxillary artery CT angiogram



Arteriovenous fistula right maxillary artery (CT angiogram, 3D surface images): large arteries and veins entangled within the right parotid gland and pinna

Lessons learned

- Correct diagnosis of vascular anomalies of the head and neck hinges on our understanding of their natural history, clinical presentation, and imaging features. High resolution, 3-dimensional capabilities, and non-invasiveness make MR imaging and MR angiography the gold standard in the diagnosis and treatment planning of craniofacial vascular anomalies.
- The impetus for treatment of head and neck lesions is to improve cosmetic appearance, eliminate functional impairment, and preclude life-threatening complications. A number of effective treatments are currently employed in the management of vascular anomalies, including pharmacotherapy, laser photocoagulation, sclerotherapy, cryosurgery, embolization, and surgical resection.
- Understanding treatment decision making is necessary to achieve optimal outcomes.

Course 03: Lesson Quiz

1. Which of the following is true for an MRI/MRA finding for a NICH and RICH?
   A. Irregular flow voids throughout the lesion
   B. Well organized, single-compartment soft tissue mass with even flow voids and moderate enhancement
   C. Well organized, multi-focal firm nodules
   D. Extremely intense enhancement with irregular flow void
2. Which of the following is true for clinical findings of an arteriovenous malformation (AVM)?
   A. Pulsitile to the touch
   B. Are always pulsitile and cold when compressed
   C. Cold and blush when compressed
   D. Compresses easily when patient is lying down, with no pulsation
3. Ultrasound and fluoroscopic techniques are used to determine the location and treatment outcomes in patients with which of the following conditions?
   A. Infantile hemangiomas and NICH lesions
   B. Venous and/or lymphatic malformations
   C. Lymphatic malformations only
   D. Arteriovenous malformations and fistulas
4. On TW2 imaging for an infantile hemangioma, which findings are atypical?
   A. Low signal and intense enhancement with irregular flow voids
   B. Intense enhancement with persistent irregular flow voids only
   C. High signal and flow voids
   D. Flow voids with low signal
5. The hallmark imaging features of hemangiomas and vascular malformations are closely coupled to
   A. Hematologic findings
   B. Clinical decision-making strategies
   C. Growth cycle of the malformation
   D. Evidence-based outcome measures
6. Post-treatment of venous malformations with sclerotherapy may typically
   A. Enhance on imaging for only 24 to 48 hours post-treatment
   B. Enhance on imaging for up to 6 to 8 weeks post-treatment
   C. Will continue to persist with enhancement on imaging despite resolution of lesion
   D. Show no signs of enhancement immediately following treatment
7. Which one of the following imaging options may lead to a surgical failure?
   A. MRI with contrast done 1 to 2 days before surgery
   B. Imaging done 5 to 7 days prior to surgery
   C. Lack of any imaging study
   D. Ultrasound with MRI done 24 hours before surgery

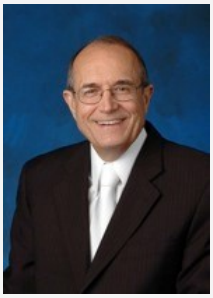
8. With an arteriovenous fistula, you will typically see the following upon image review

A. Large veins only
B. Small arteries and capillaries
C. Large capillaries and lymphatics
D. Larger arteries and veins

9. Classification and categorization of vascular anomalies of the head and neck is best determined with which of the following?

A. MRI and ultrasound
B. MRI without contrast
C. MRA with contrast and ultrasound
D. MRI and MRA

AUTHOR PROFILES



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Dr. Hasso is board certified in Diagnostic Radiology and sub-certified in diagnostic neuroradiology. He is an expert in clinical imaging of the brain and spine, and an investigator on applications of CT and MRI in disorders of the head and neck. His clinical expertise is held in the highest regard not only within neuroradiology, but also among specialists in neurology, neurosurgery, otolaryngology/head and neck surgery, ophthalmology, and by radiologists worldwide.

Dr. Hasso is a superb teacher and has received “Teacher-of-the-Year” awards from both the Department of Radiology and the Department of Neurology at University of California Irvine. His knowledge of all aspects of neuroradiology and head and neck radiology is extensive and he has a wonderful ability to communicate to students his knowledge of, and enthusiasm for, the subject. The popularity of his teaching is reflected in the large number of both national and international lectures that he has given in the last 35 years at various society meetings and seminars. He has been an invited professor at 12 major universities, and a Gold Medalist of both the American Society of Neuroradiology and the American Roentgen Ray Society.

Dr. Hasso is the author or co-author of six textbooks and over 140 articles in refereed scientific journals. He has contributed over 70 chapters for textbooks in the fields of neuroradiology, head and neck radiology and MRI. His most recent textbook on Diagnostic Imaging of the Head and Neck has a chapter dedicated to imaging of vascular anomalies and vascular tumors in children and adults. Dr. Hasso is a reviewer and member of the editorial boards of seven scientific journals in his specialty.

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Course 03: Lesson Quiz Answer Key

1. B
2. A
3. B
4. A
5. B
6. B
7. C
8. D
9. D