Congenital Head & Neck Lesions in Children

A Roadmap

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I am on Youtube !

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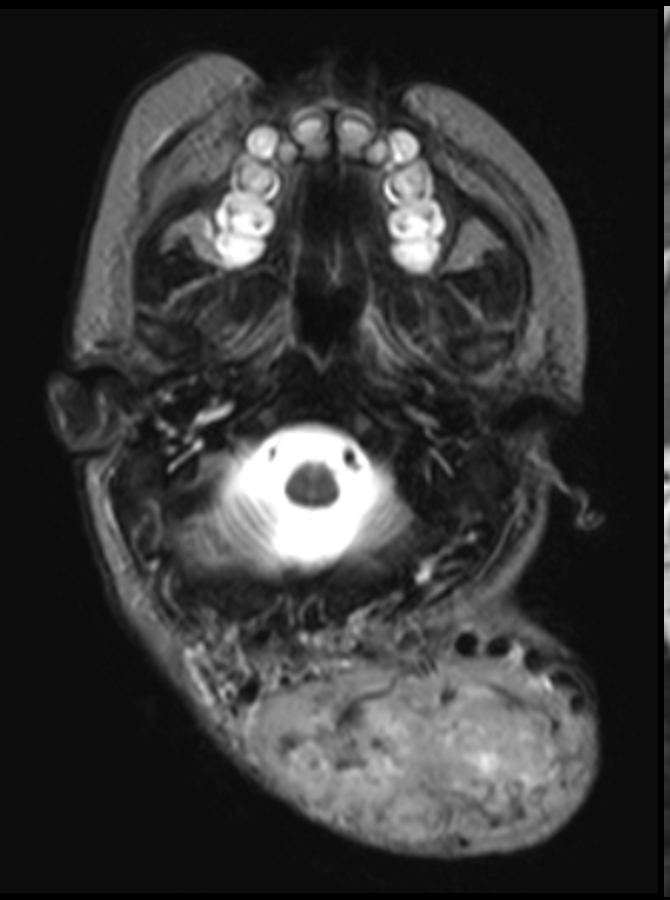
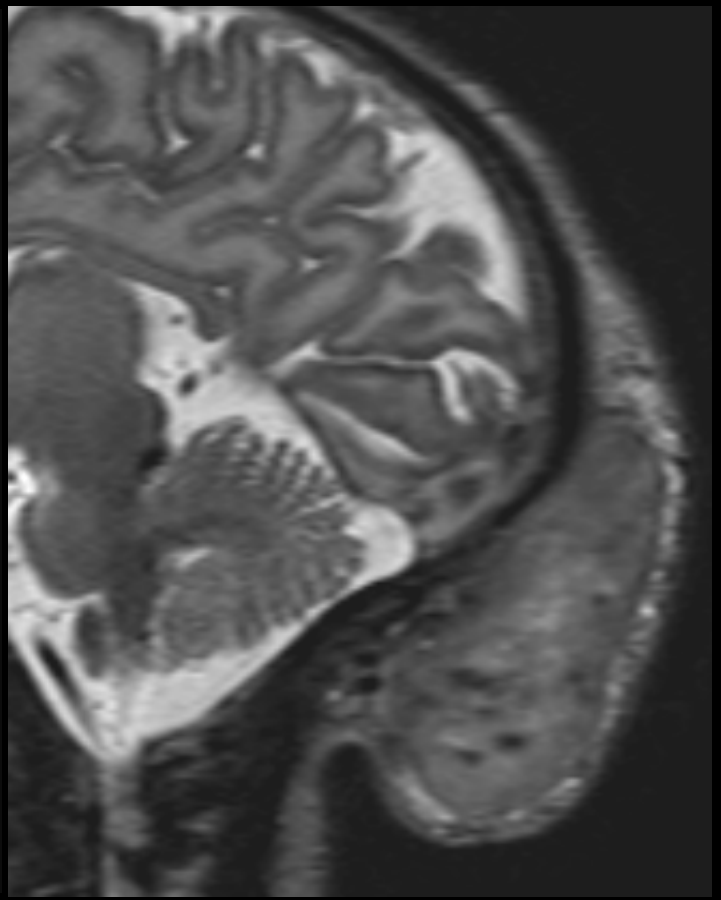
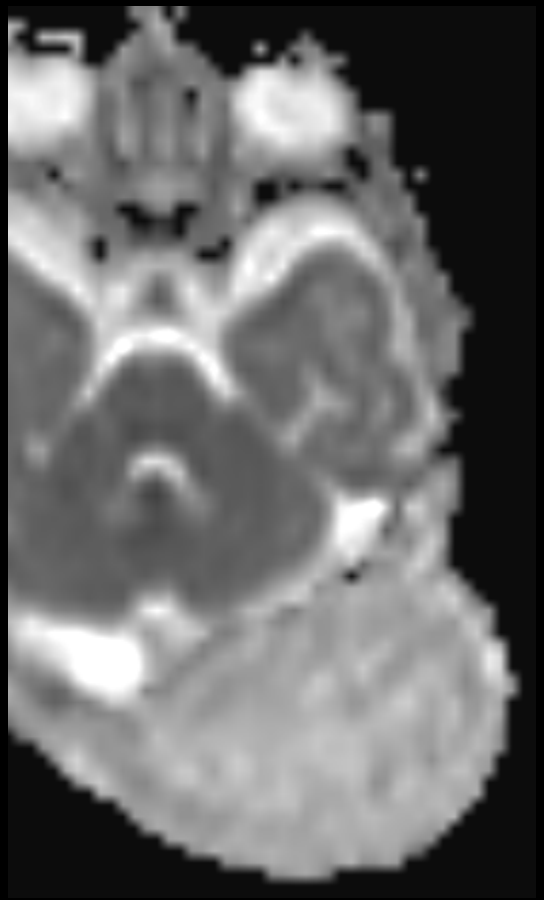
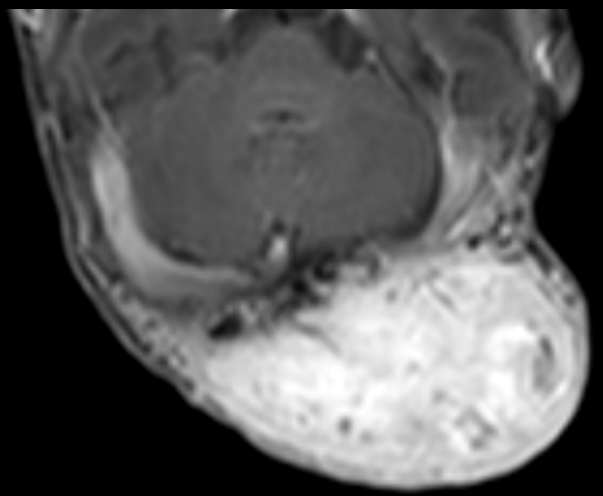
Structure of this Talk

- **Case-based presentation**
- **Case → teaching points → radiological differential diagnoses**
- Few pools: try to answer

3 month-old baby girl, redish cutaneous mass lesion in the occipital region

1) Well-defined mass
2) Homogeneous, intense enhancement
3) Internal flow voids
4) Free diffusion

Question: what's the diagnosis?

A. Infantile Hameangioma
B. Venous malformation
C. Veno-lymphatic malformation
D. AVM

Question: what's the diagnosis?

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B. Venous malformation
C. Veno-lymphatic malformation
D. AVM

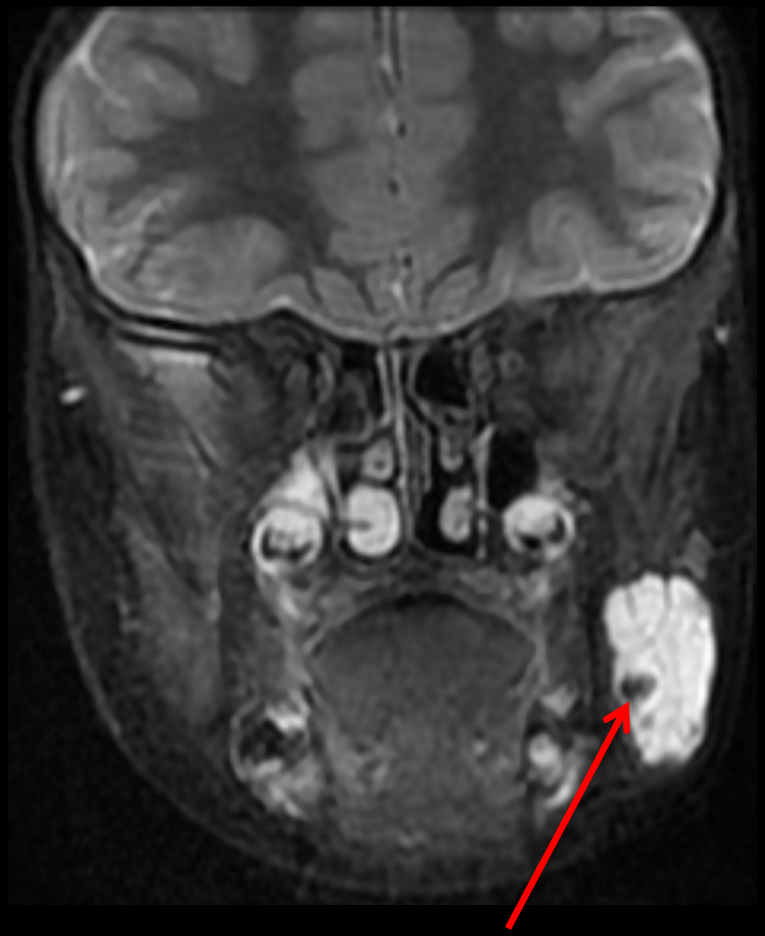
- Why is not any of the others??

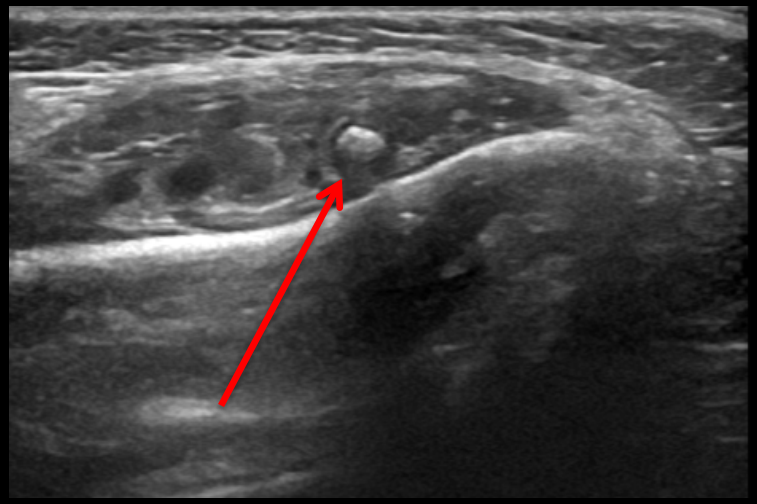
Congenital and Infantile Hemangiomas

- IMAGING Key Features:

- ✓ Well-defined, strongly enhancing mass, mildly hyper T2 to muscle
- ✓ Internal Vessels (Serpiginous Flow Voids)
- ✓ No Calcifications! (*DD Venous Malformation*)
- ✓ US: mean venous peaks not elevated *(DD AVM)*
- ✓ Involuting Phase: fatty replacement

Differential Diagnosis





Question: What's the arrow pointing at?

A. Flow void (vessel)
B. Calcium (Phlebolith)
C. Blood (clot)
D. Nidus (AVM)

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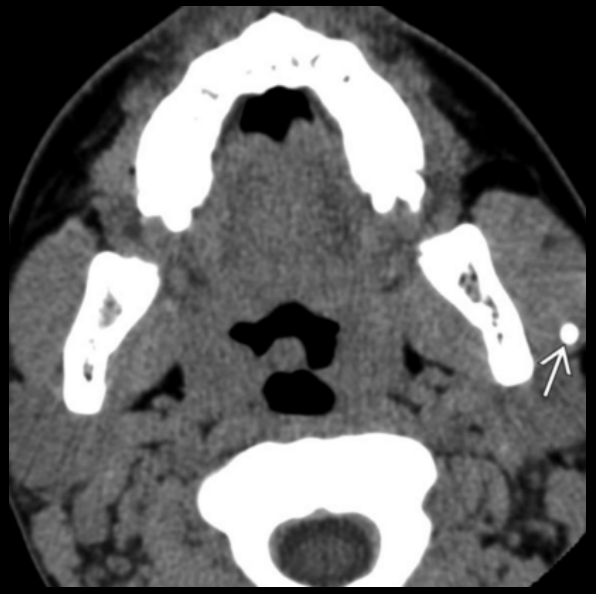
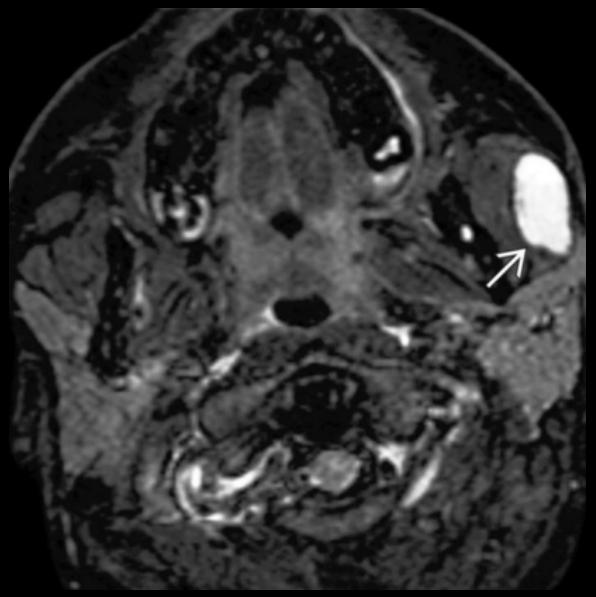
Differential Diagnosis





1) Venous Malformation

- Large venous lakes

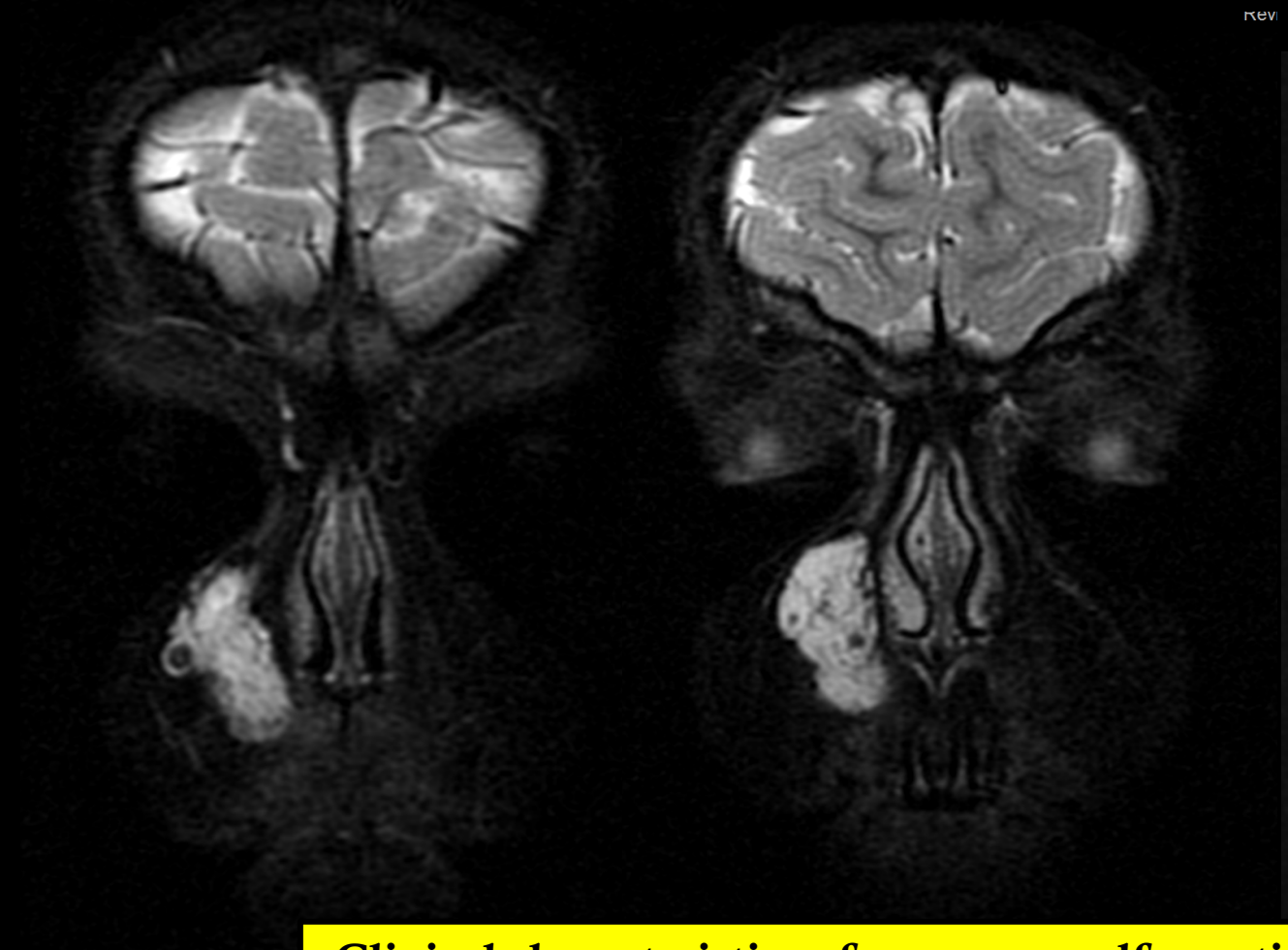
- **T2 signal more hyperintense**

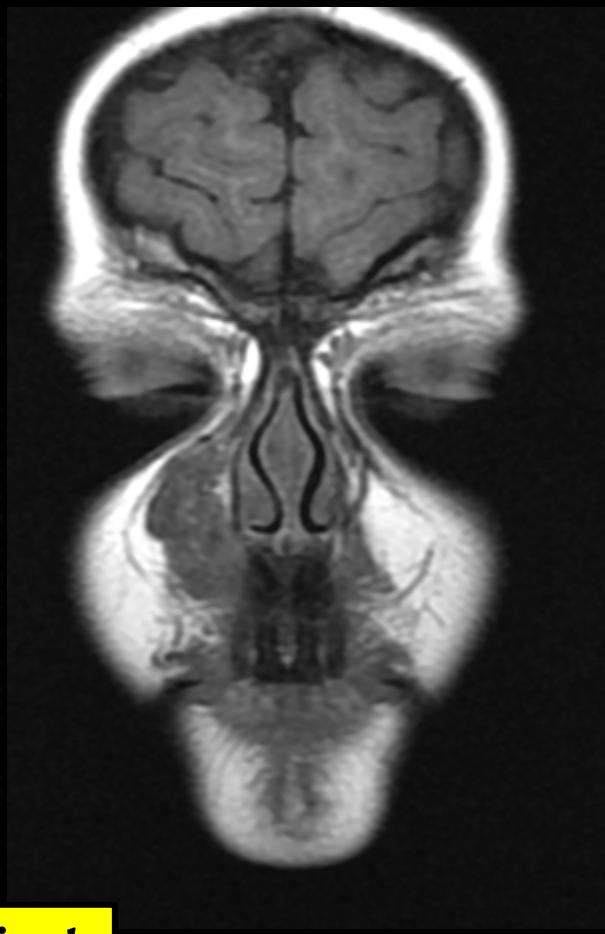
- Variable enhancement (patchy, heterogeneous), changes over time

- **Phleboliths:** Calcium within the lesion

- **No Flow voids**

4 yo, Female, right cheek vascular malformation (*external scan… that's all we got*)

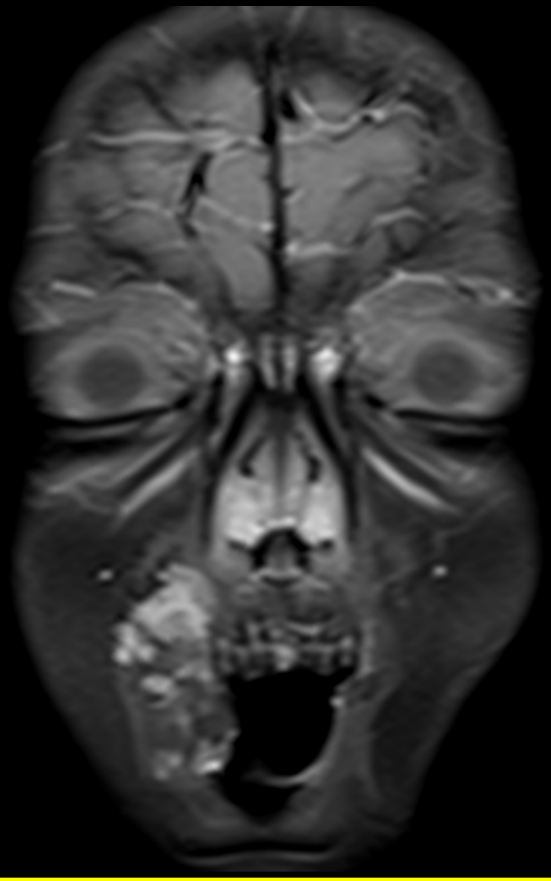


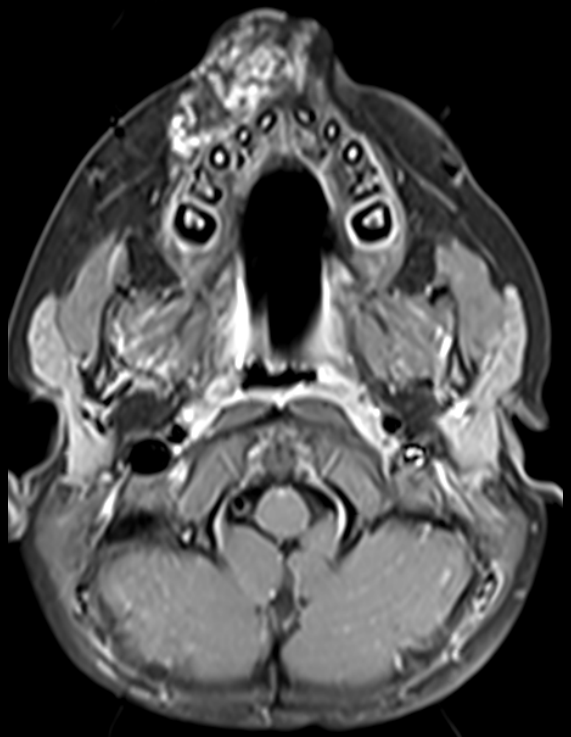


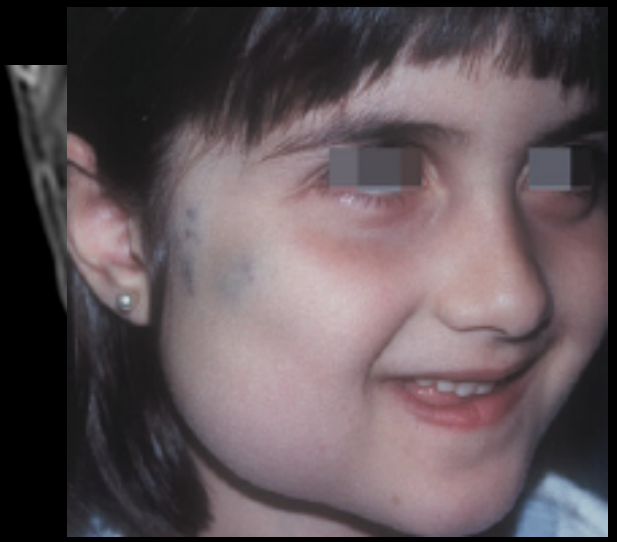
Clinical characteristics of a venous malformation!

"present at birth, flesh colored bluish lesion that expands after the Valsalva maneuver and may be flattened with applied pressure"

2 yo, Female, upper lip lesion







Pearl: trust the contrast more than the T2 signal!

Pearl: ask your clinician what the lesion looks like!

Bonifazi E. – Differential diagnosis in Pediatric Dermatology

1. Hemangioma

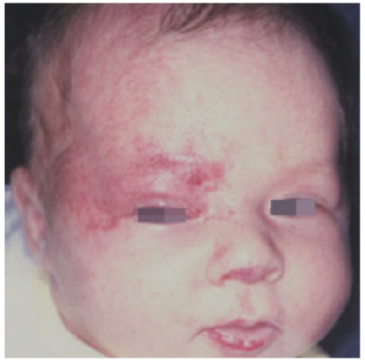


Fig. 1a

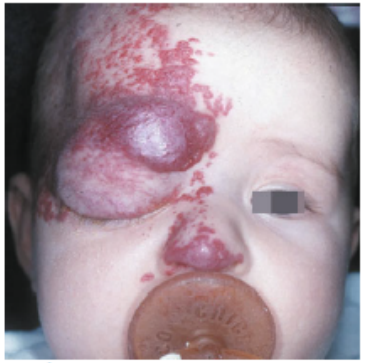


Fig. 1b Same patient as in Fig. 1a, aged 3 months

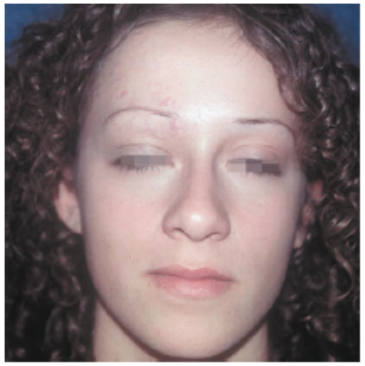


Fig. 1c Same patient as in Fig. 1a and 1b, aged 16 years

2. Venous Malformation

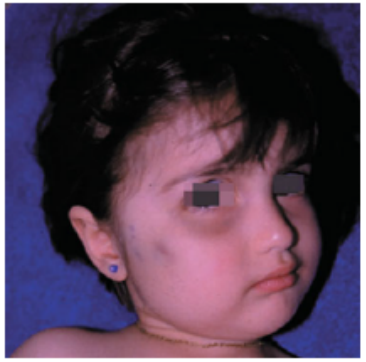


Fig. 2a

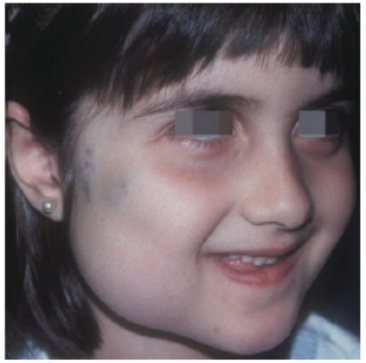


Fig. 2b Same patient as in Fig. 2a, aged 5 years

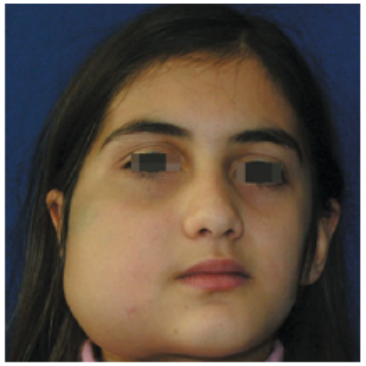


Fig. 2c Same patient as in Fig. 2a and 2b, aged 16 years

Pearl: ask your clinician what the lesion looks like!

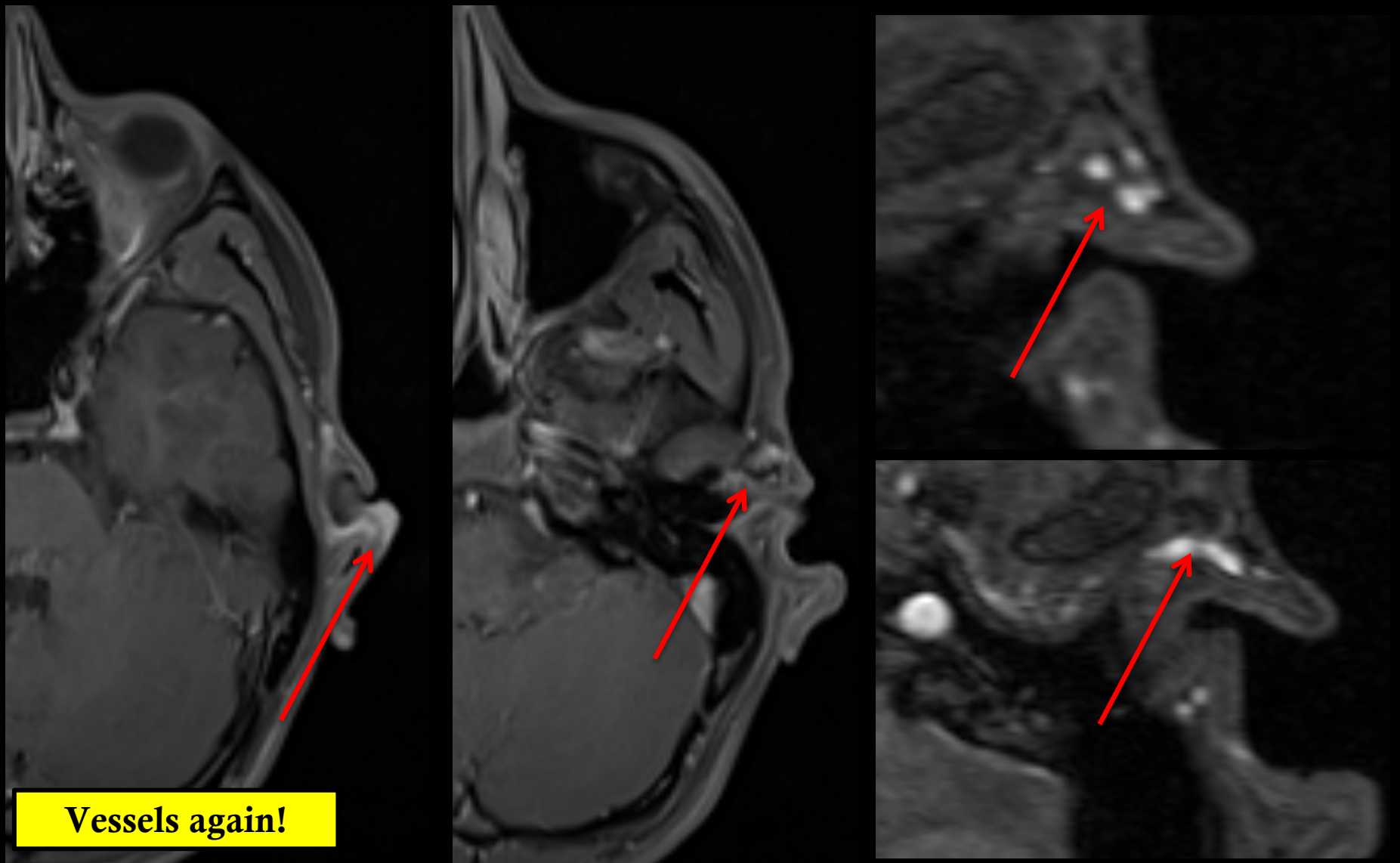
1. Hemangioma
2. Venous Malformation

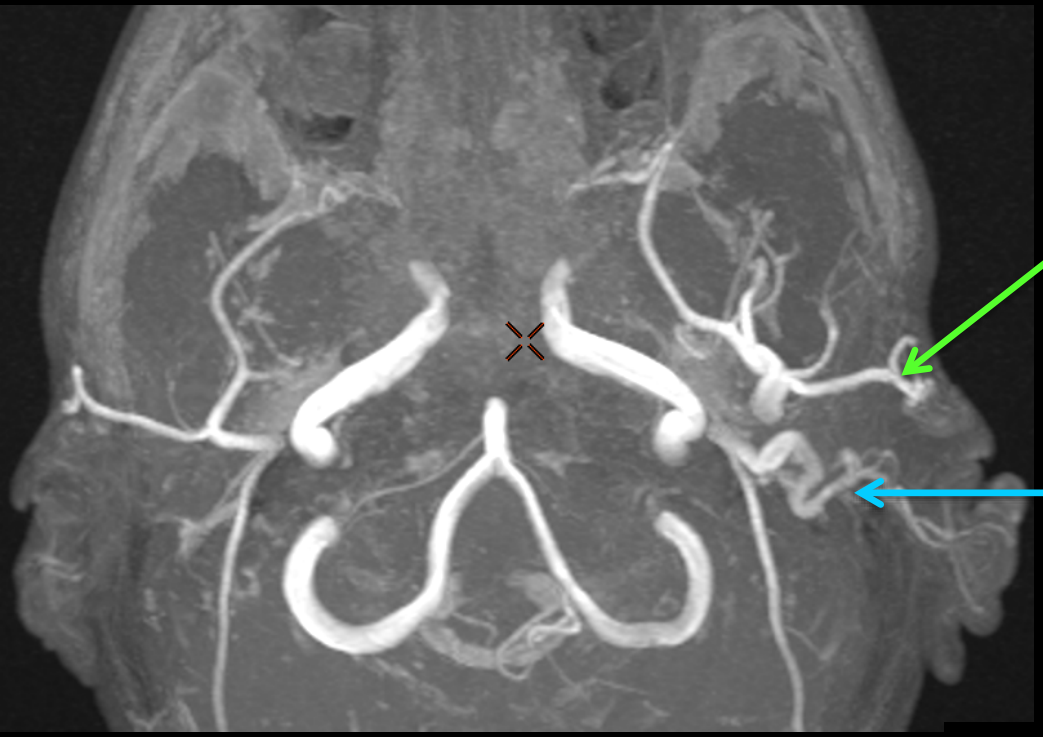
Although their natural history is completely different, hemangioma and venous malformation may present with comparable clinical features, making the differential diagnosis difficult.

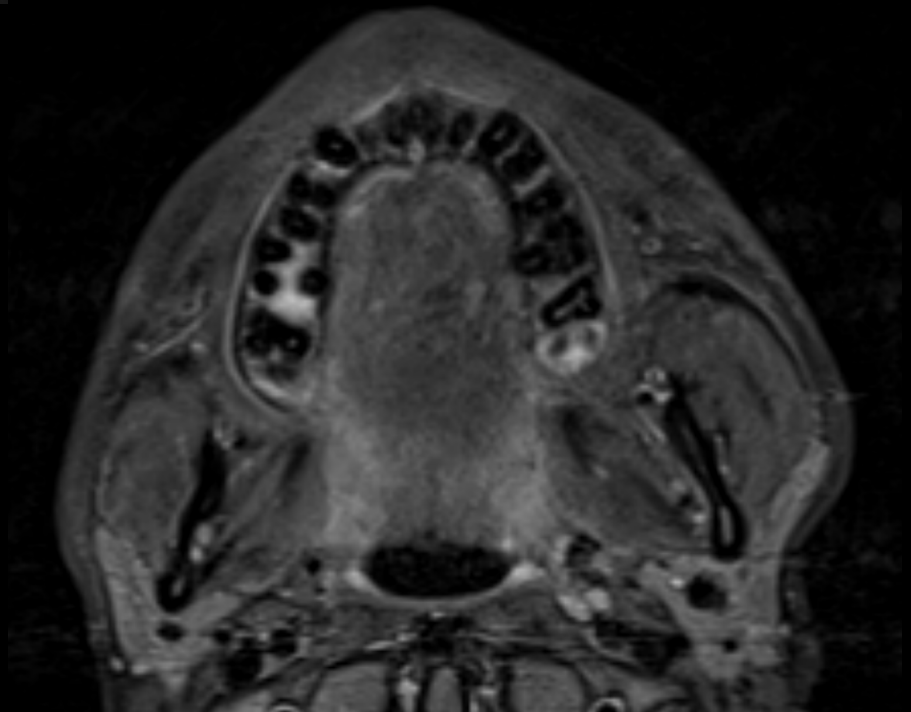
	1. Hemangioma	2. Venous Malformation
Definition	Benign tumor of vasoformative cells.	Malformation of venous vessels.
Frequency	4% of all pediatric skin disorders [5].	Less than 0.1% of all pediatric skin disorders [5].
Time of onset	Present at birth in 50% of cases.	Present at birth.
Initial clinical features	Usually, flat pink patch, sometimes ischemic with telangiectasias, livedoid. Rarely raised lesion.	Flesh-colored or red-bluish, compressible tumor, sometimes with coarse vessels.
Changes in the first few months	It grows, sometimes significantly.	It does not grow.
Changes in subsequent years	Slow, though significant, regression.	It becomes significantly more evident within decades.

Bonifazi E. – Differential diagnosis in Pediatric Dermatology

Differential Diagnosis







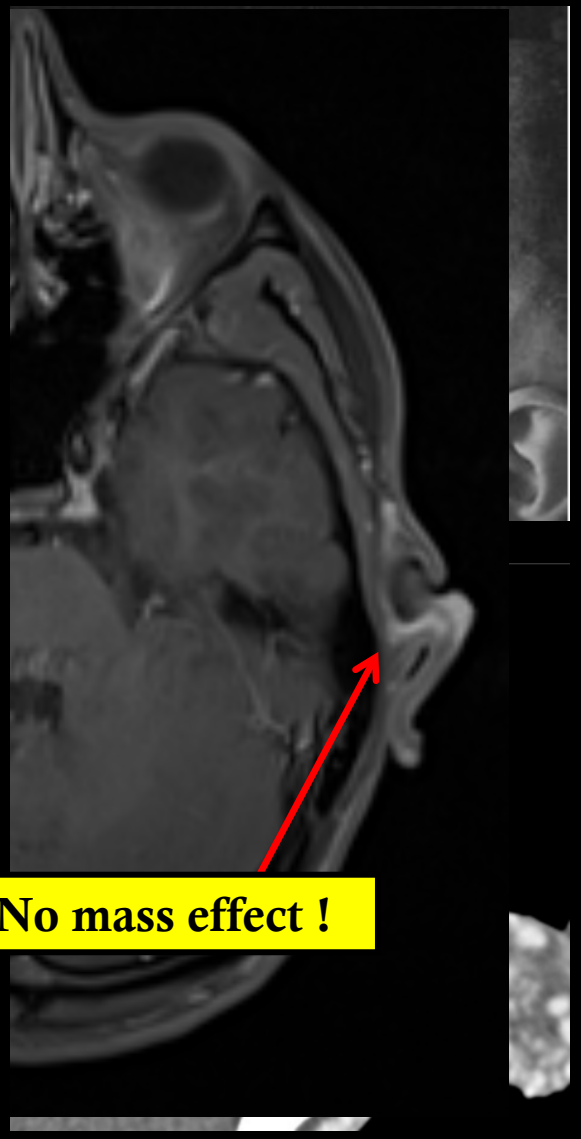
RASA1 mutation (CM-AVM syndrome)

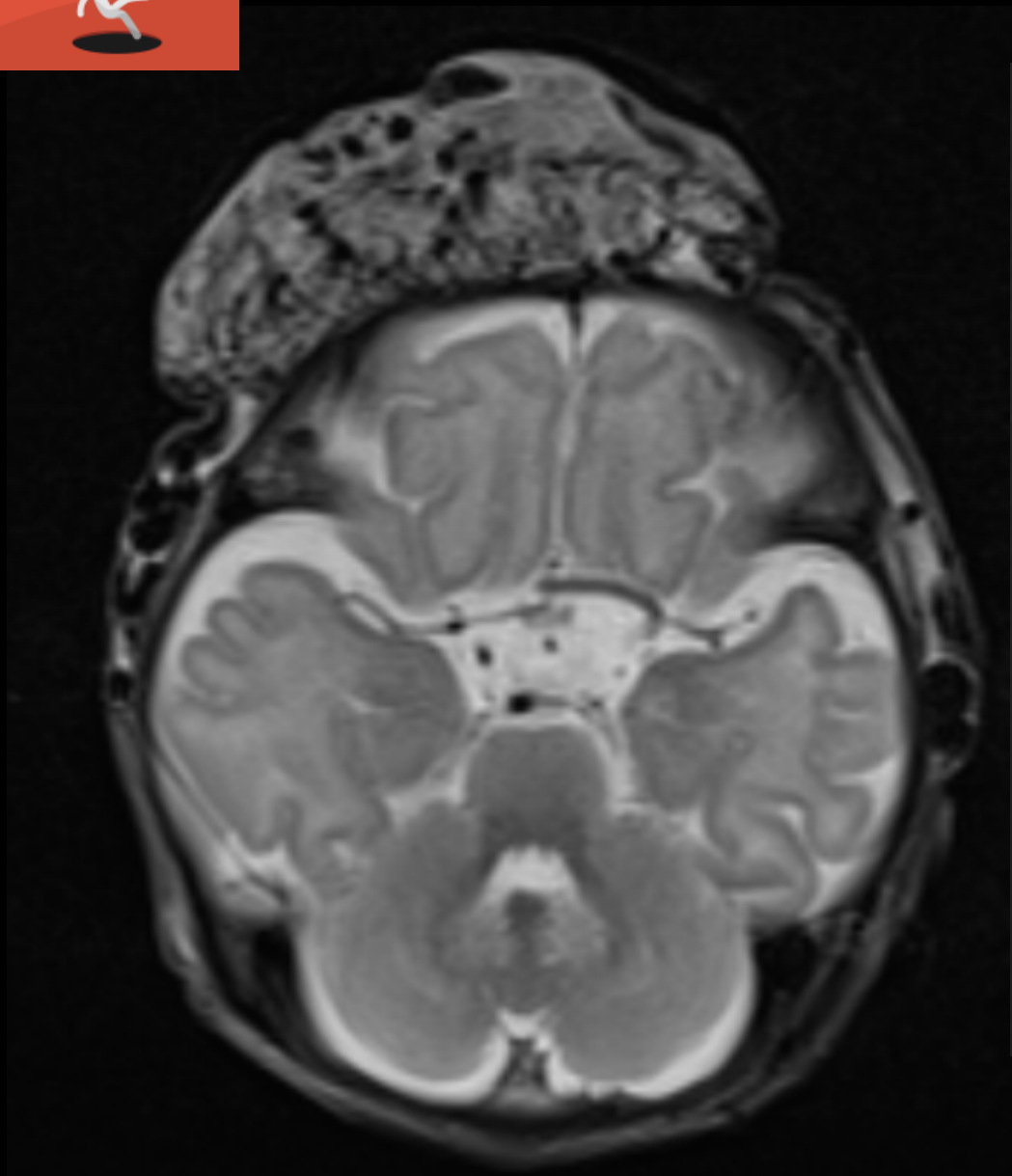
Micro-AVMs confirmed on angiography…. Nidus too small to be seen on MRI/MRA

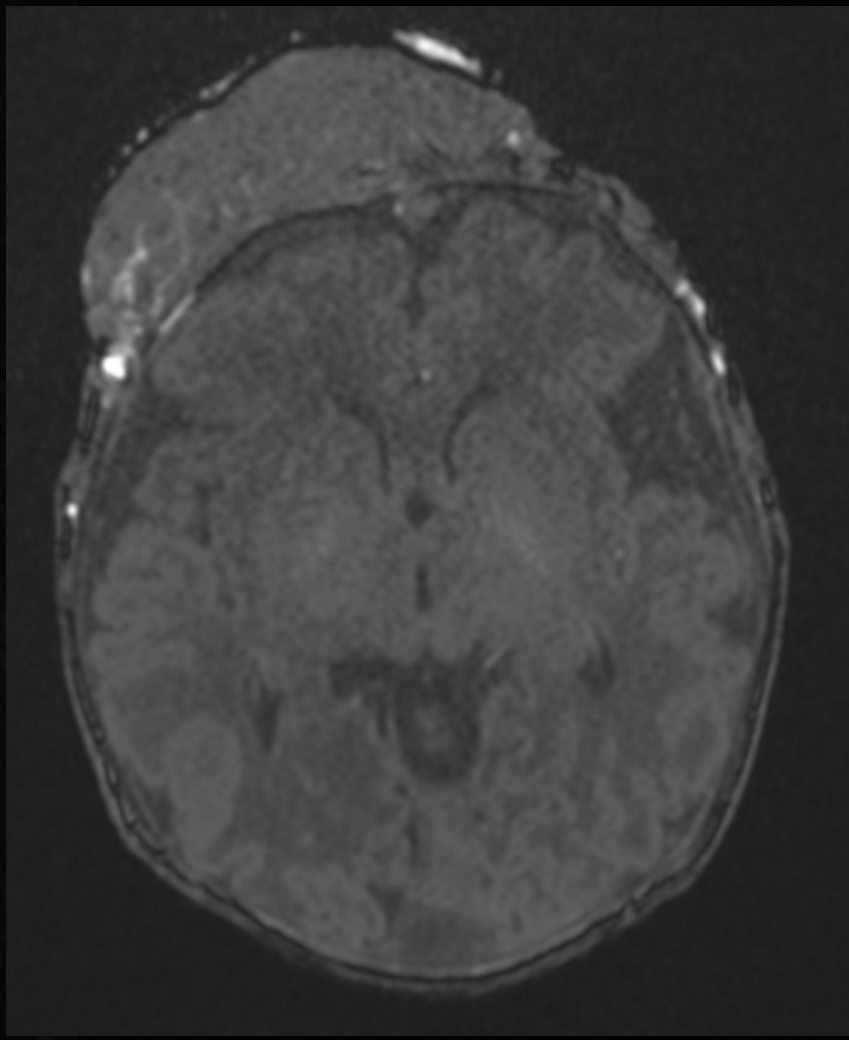
Differential Diagnosis

2) AVM

- High flow and tortuous feeding arteries
- Nidus/AV shunting, NO MASS
- **US: elevated venous peaks**
- **Worsening overtime**
- Clinical: arterial feeding is evident







MRA

Question: what's the diagnosis?

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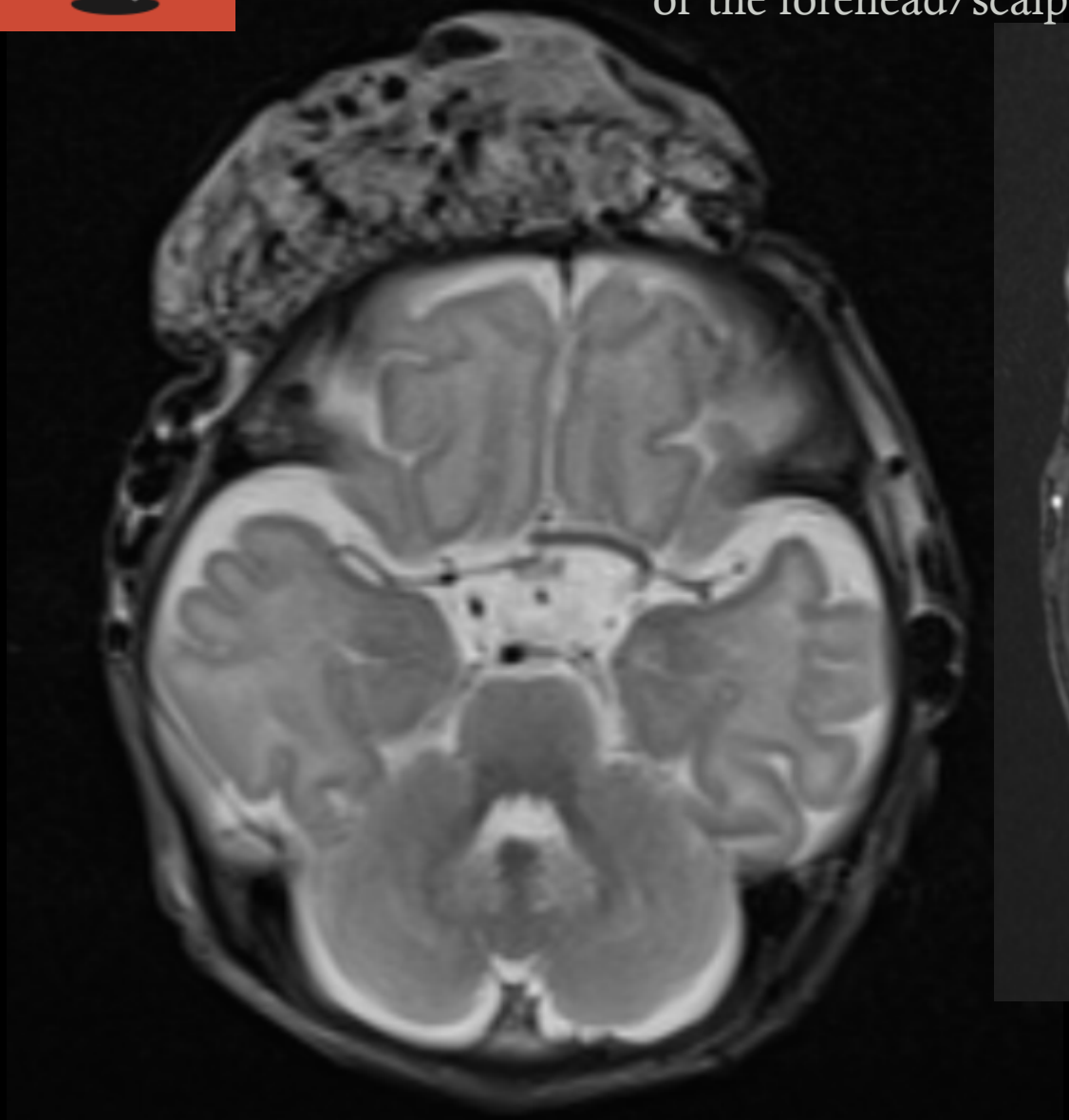
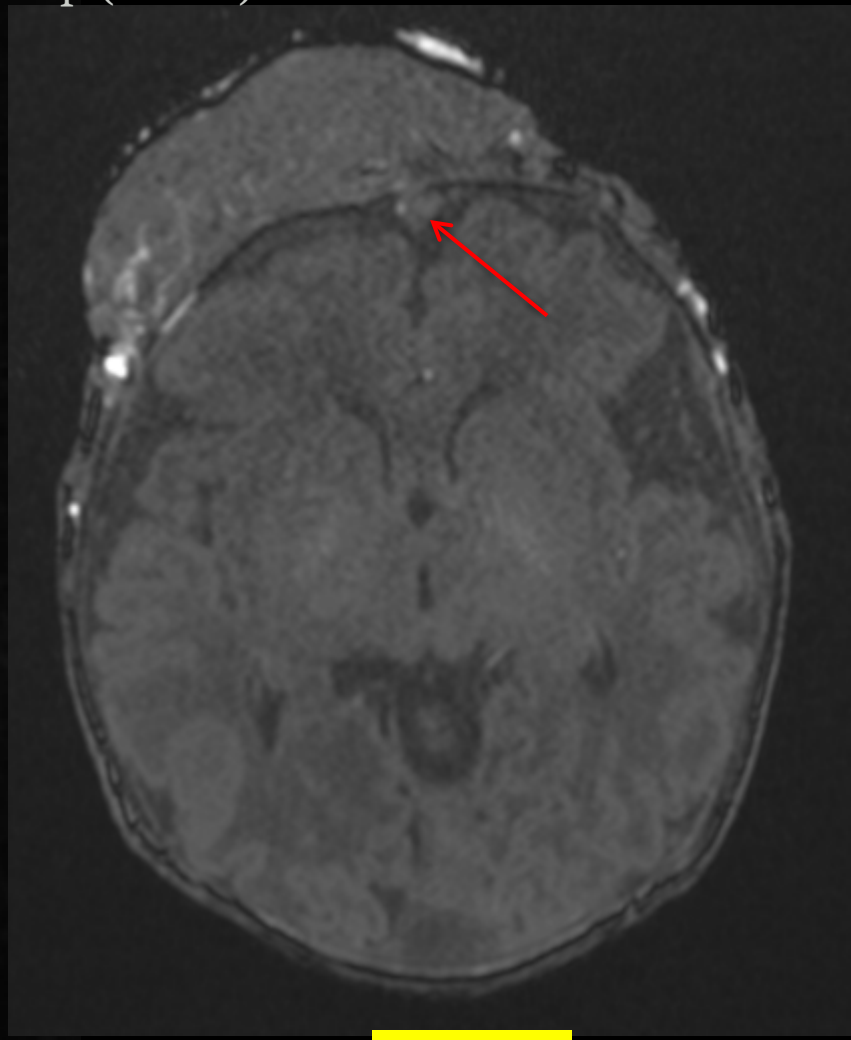
Rapidly involuting congenital haemangioma of the forehead/scalp (RICH)

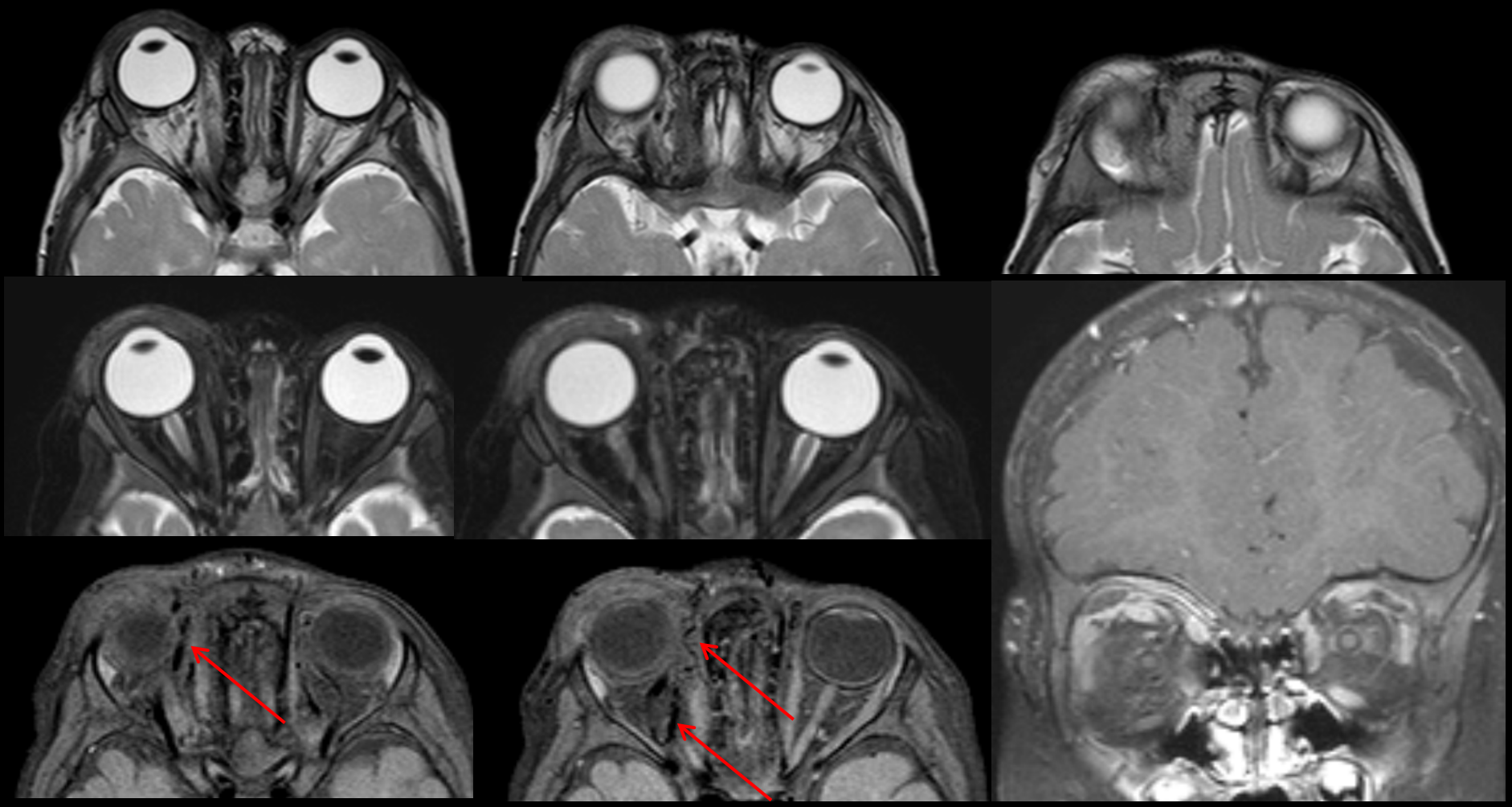




MASS!!!

MRA: no AV shunt demonstrated

1 yo, male, right proptosis



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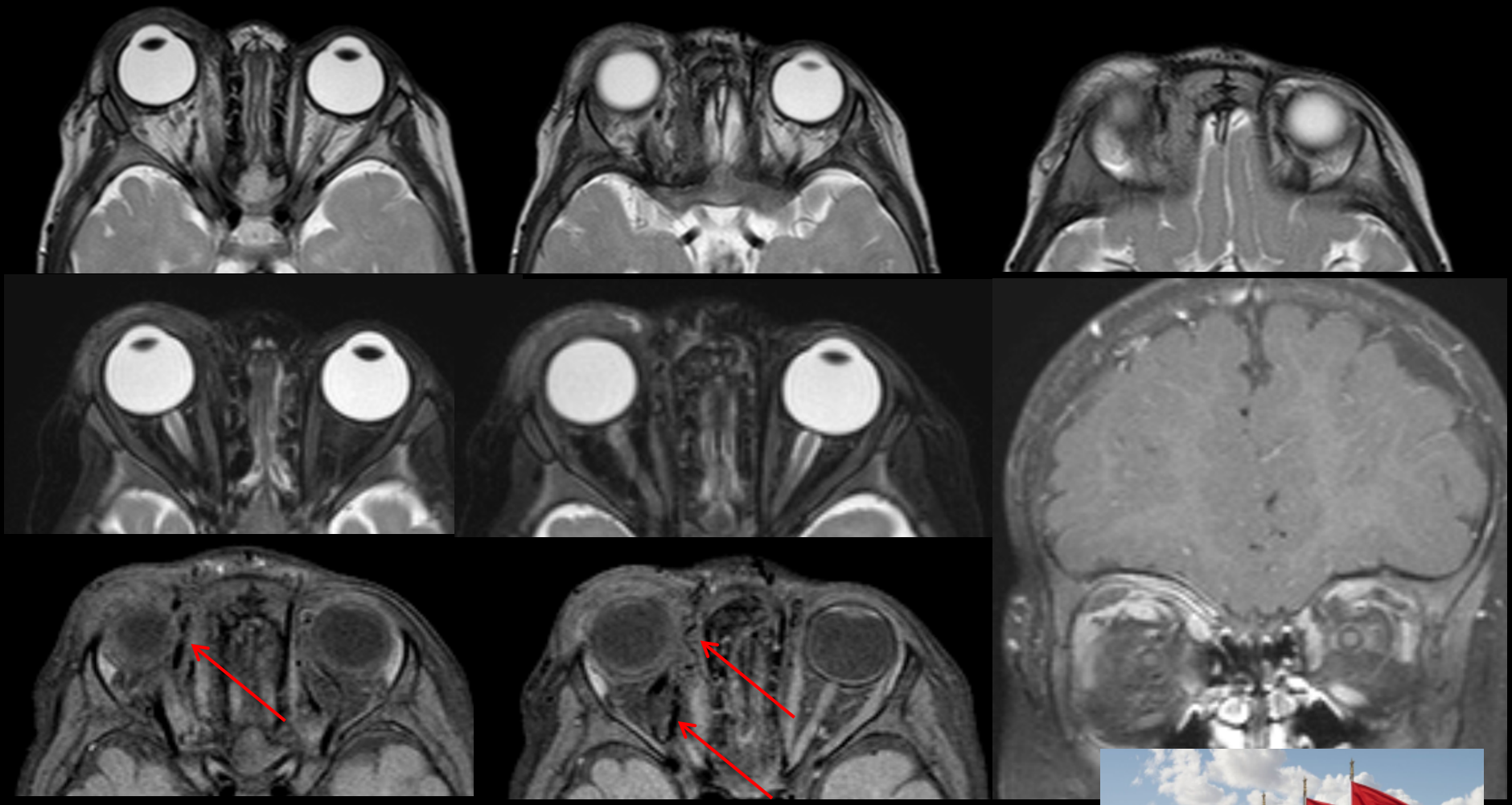


Too many vessels, NO MASS, worsening overtime…. AVM

Vascular anomalies: Description, classification and nomenclature

Deborah R. Shatzkes, MD

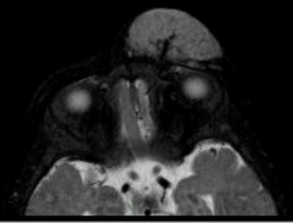
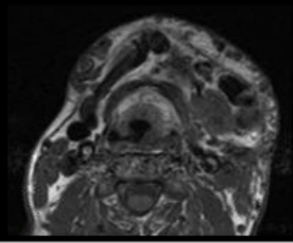
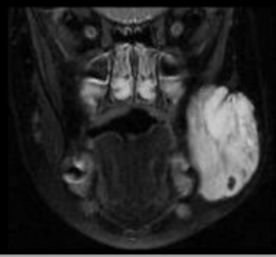
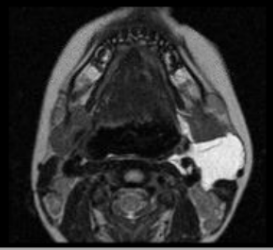
Table 1. Abbreviated 2014 ISSVA Classification of Vascular Anomalies, with examples

Vascular Tumors	Vascular Malformations			
	Simple	Combined	Of major named vessels	Associated with other anomalies
Benign				
Hemangioma (infantile)	CM	VM + LM	Persistent stapedial artery	Sturge-Weber syndrome
Hemangioma (congenital)	VM	CM + AVM	PHACE-associated carotid anomalies	Maffucci syndrome
Pyogenic granuloma	LM	CM + VM		Klippel-Trenaunay syndrome
Borderline	AVM	CM + LM		
Kaposiform hemangio-endothelioma	AVF			
Kaposi sarcoma				
Malignant				
Angiosarcoma				

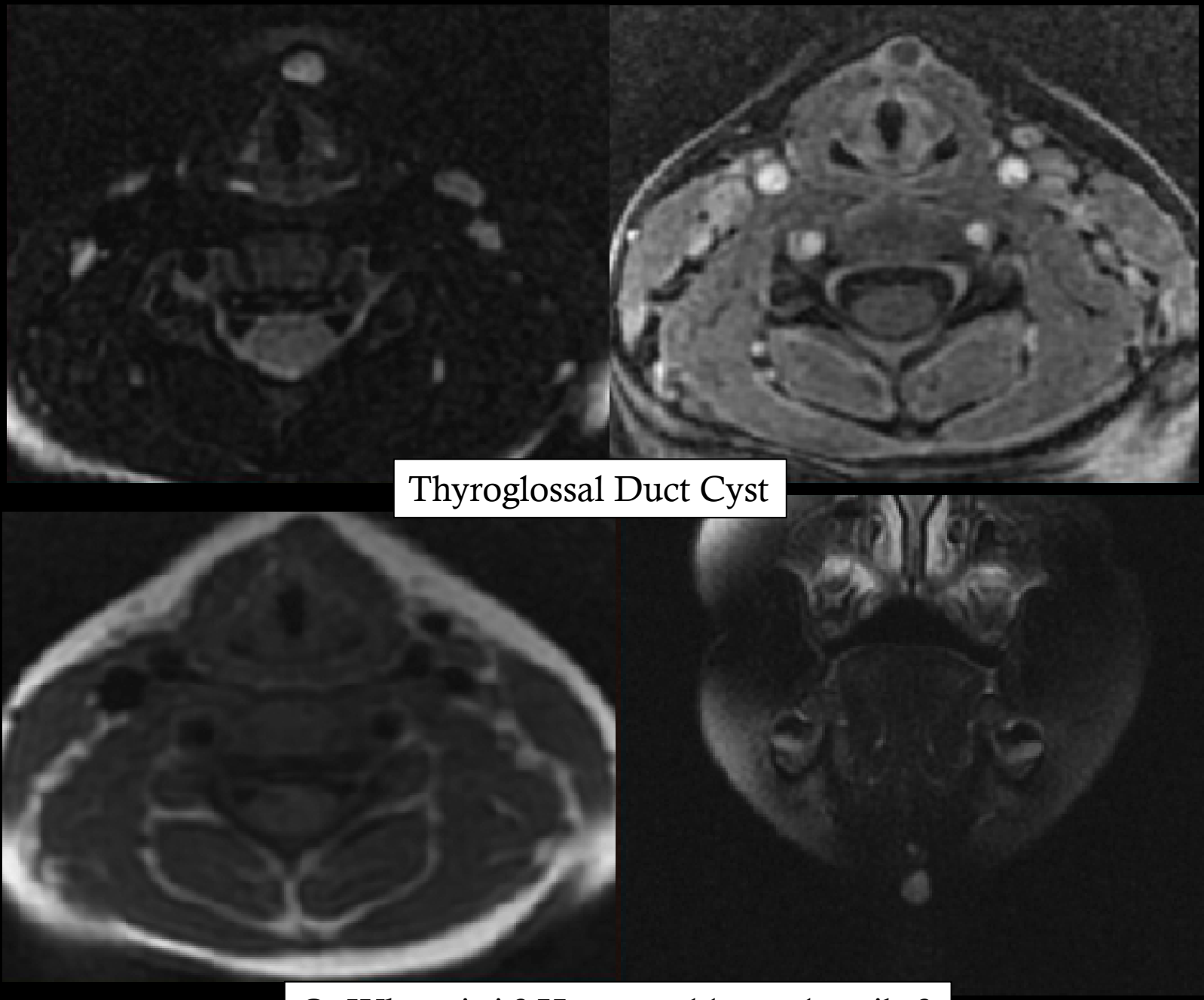
Table 3. Imaging Characteristics of Common Vascular Anomalies

Lesion	Morphology	Enhancement	T2 signal	Comments
Hemangioma (Phase I)	Lobular, hypervascular	++	+	Only moderately increased T2 signal reflects hypercellularity
VM	Trans-spatial, common phleboliths	++	++	High T2 signal reflects low cellularity
LM (macrocystic)	Large cystic spaces	-	++	Image like cysts; hemorrhagic fluid levels common
LM (microcystic)	Multiseptated; very small cystic spaces	+/-	+	Low fluid content; may appear solid
AVM	Vascular flow voids without discrete mass	+	+	Hyperperfusion results in tiss

Haemangioma is a tumour!

	(+) Enhancement	(-) Enhancement
↑ T2 SI	**Hemangioma** • Cellular neoplasm • Triphasic growth • Large vessels in Phase I 	**AVM** • Vascular flow voids • No discrete mass • Bony lysis and overgrowth 
↑ ↑ T2 SI	**Venous Malformation** • Phleboliths • Dynamic enhancement 	**Lymphatic Malformation** • Macrocystic and microcystic • Hemorrhagic fluid levels 

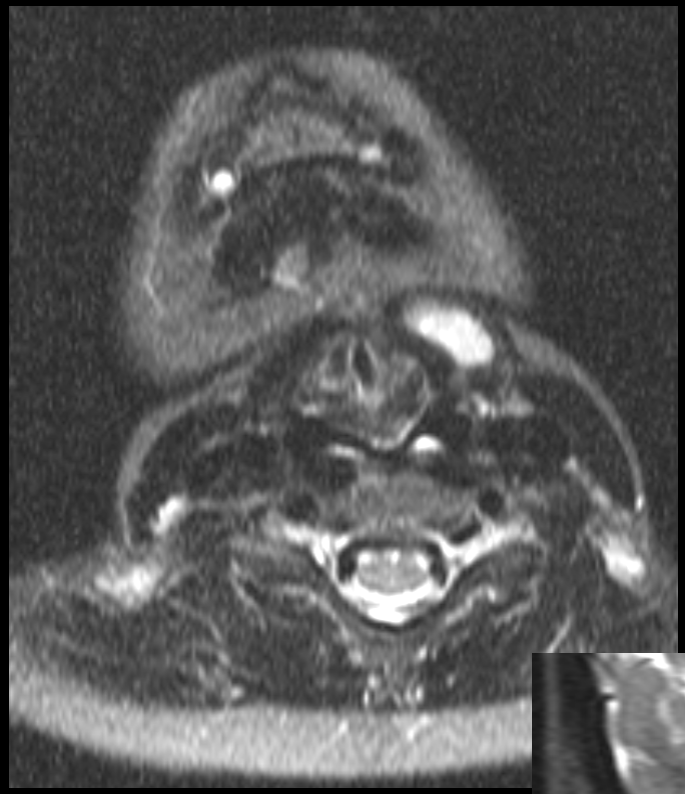
Courtesy: D. Shatzkes

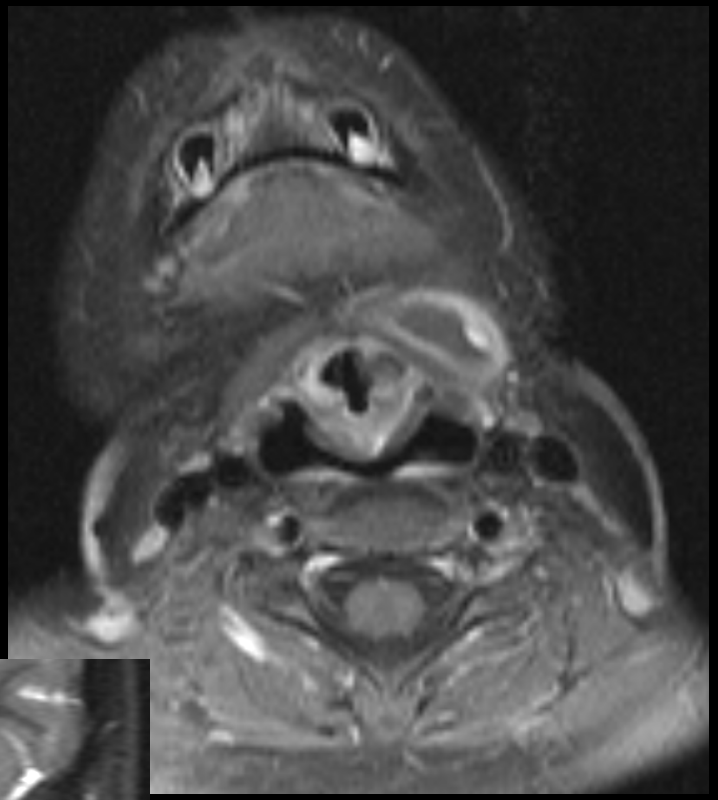


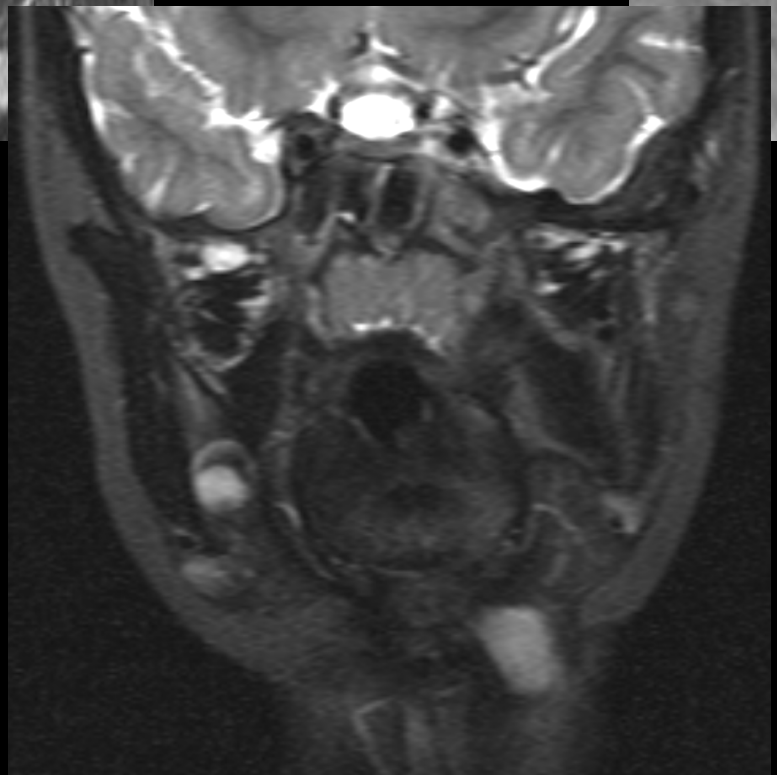
Thyroglossal Duct Cyst

Q: Where is it? How would you describe?

source: Slideshare







Question: what's the diagnosis?

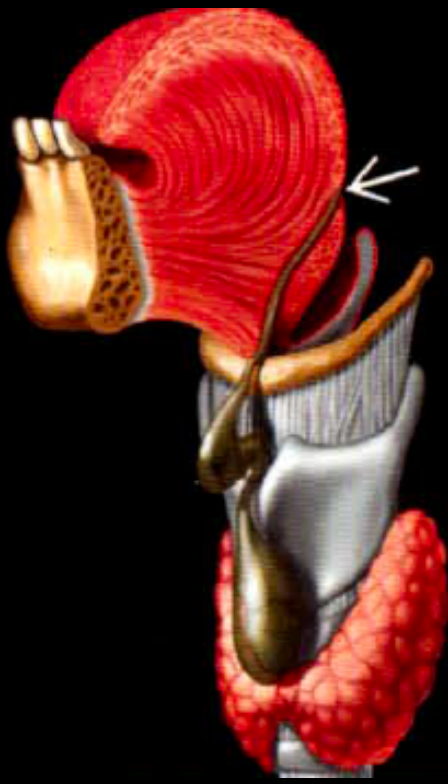
A. 1st Branchial Apparatus Anomaly (BAA)
B. 2nd Branchial Apparatus Anomaly (BAA)
C. Veno-lymphatic malformation
D. TGD cyst

Question: what's the diagnosis?

A. 1st Branchial Apparatus Anomaly (BAA)
B. 2nd Branchial Apparatus Anomaly (BAA)
C. Veno-lymphatic malformation
D. TGD cyst

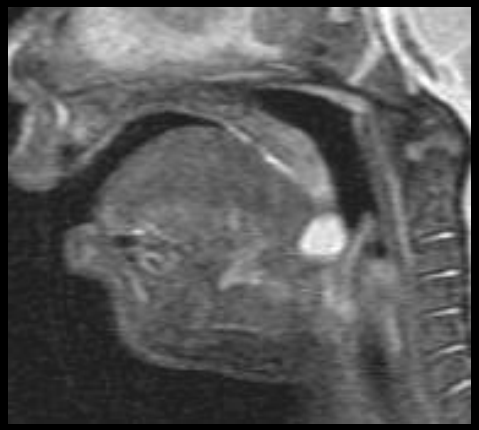
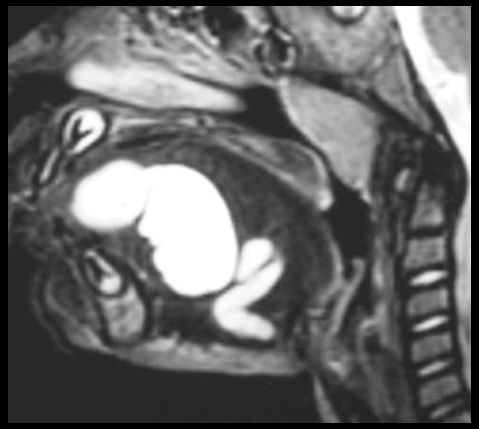
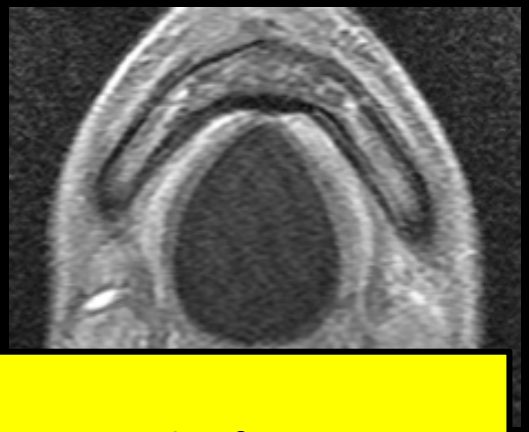
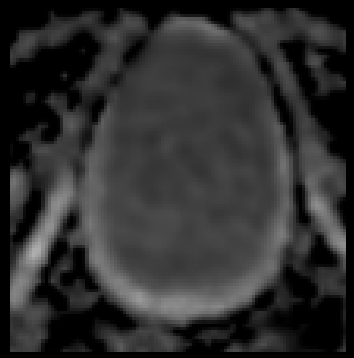
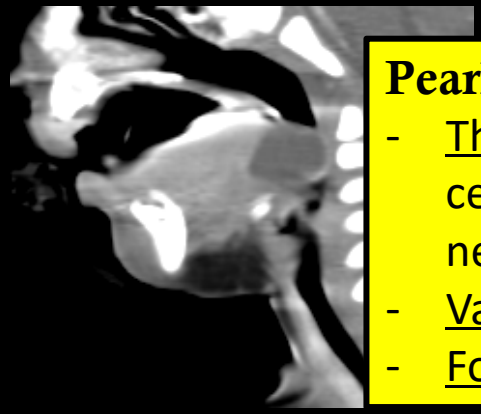
Thyroglossal Duct Cyst

- Remnant of the TGD (Between foramen cecum at tongue base → thyroid bed in infrahyoid neck)
- Most common congenital neck lesion
- Median cyst (could be also paramedian in the infrahyoid neck)
- Thin rim of c.e. is possible (often associated with infection)
- Embedded by strap muscles ("claw sign")



Harnsberger 2004

Midline cyst ddx

Pearl: LOCATION!

- Thyroglossal duct cysts →between the foramen cecum and hyoid bone or within the infrahyoid neck.
- Vallecular cysts→ in the vallecula.
- Foregut Dupl Cyst→ ant 1/3 tongue

Dermoid cyst

Vallecular cyst

Well defined, non enhancing ,fluid filled mass at the base of the tongue , pushing epiglottis against pharyngeal wall

Foregut duplication cyst

Cystic , non-enhancing mass (signal similar to CSF), ant 1/3 tongue ++, midline or midline + left extension, Unilociular or serpiginous

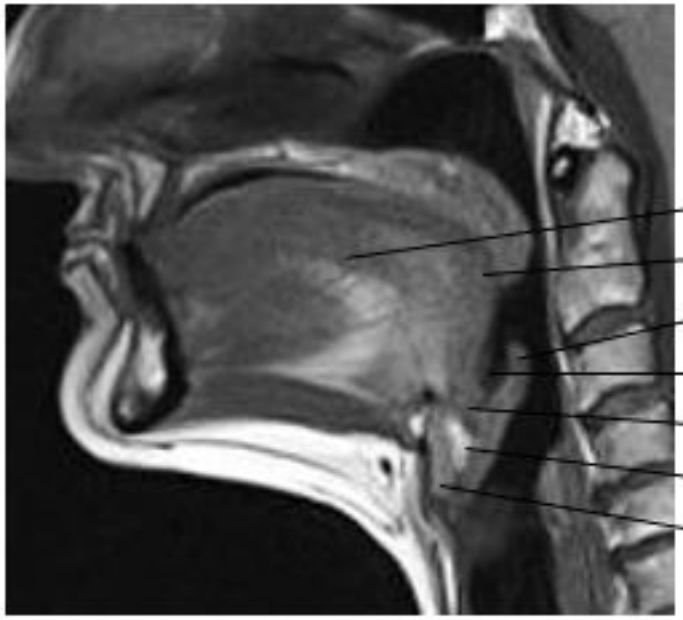
Slide courtesy of Dr. C. Robson

ORIGINAL ARTICLE

Foregut Duplication Cysts in the Head and Neck

Presentation

Stephen M. Kieran ... *MD*



Objective: To re... management of fo... neck in our instit...

Design: An insti... spective review of ... tion cysts of the h...

Setting: Pediatri...

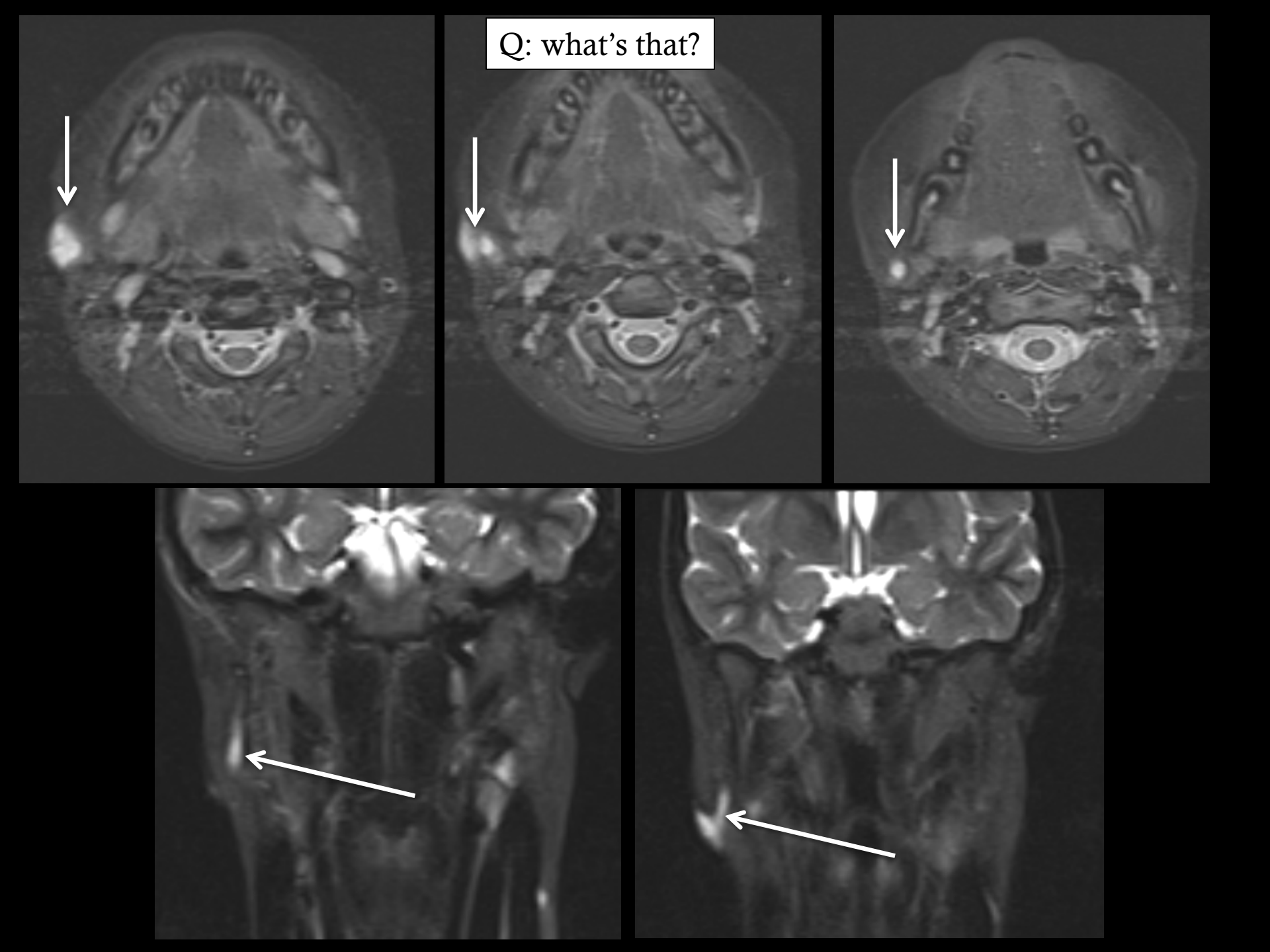
Patients: Twent... confirmed foregut duplication cysts of the head and neck were identified. Fourteen patients (64%) were male. The median age at diagnosis was 1.5 years (age range, 5 days to 7 years).

Main Outcome Measures: Clinical data, including age, presenting symptoms, anatomical site(s), evaluation, treatment, and complication, were recorded and analyzed.

Results: Presentation varied depending on anatomical site ...%) being asymptom... l cavity (n=12), oro...), and neck (n=3). ...3 patients and con... ng (n=8), computed ...aphy (n=1), demon... s. All patients under... l on excising the cyst, ...al tissues. No patient ...up.

Conclusions: Foregut duplication cysts of the head and neck, although uncommon, should be included in the differential diagnosis of cystic head and neck lesions. Preoperative imaging is recommended to differentiate these lesions from other congenital head and neck masses. Surgical excision biopsy with complete removal of the mucosal lining is curative, with no instances of recurrence in our series.

Arch Otolaryngol Head Neck Surg. 2010;136(8):778-782


Q: what's that?

Question: what's the diagnosis?

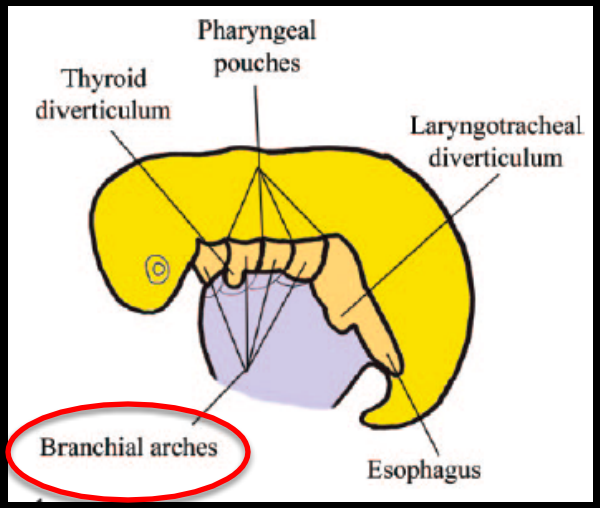
A. 1st Branchial apparatus anomaly (BAA)
B. 2nd Branchial apparatus anomaly (BAA)
C. Veno-lymphatic malformation
D. TGD cyst

Question: what's the diagnosis?

A. 1st Branchial apparatus anomaly (BCA)
B. 2nd Branchial apparatus anomaly (BCA)
C. Veno-lymphatic malformation
D. TGD cyst

Branchial Apparatus Anomalies

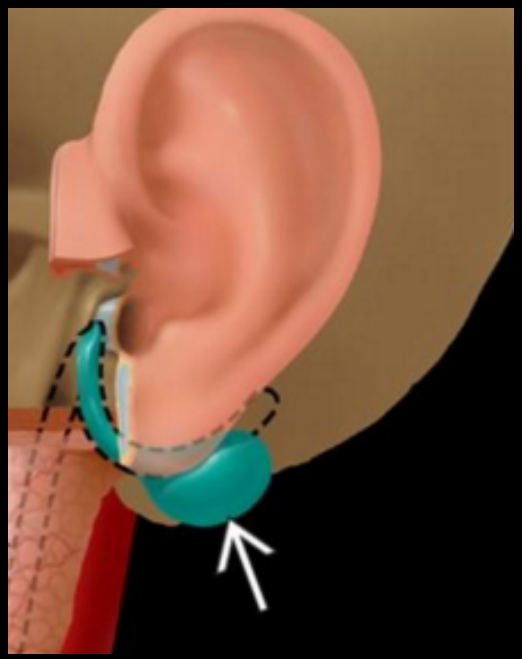
- Congenital malformations during development of the branchial apparatus
- Pits, tags, cyst , sinuses and fistulae from the 1st , 2nd, 3rd and 4th branchial arches
- 2nd branchial apparatus anomaly is the most common: 95%



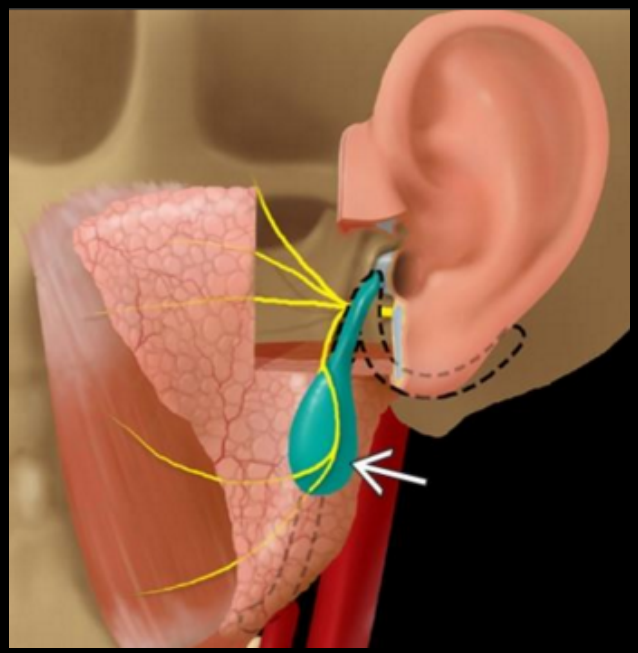
Head and neck region at 4 weeks gestation (Meuwly et al 2005)

1st Branchial Apparatus Anomaly

- Benign, congenital cyst in or adjacent to parotid gland, EAC, or pinna
- Several classifications related to embryology or location

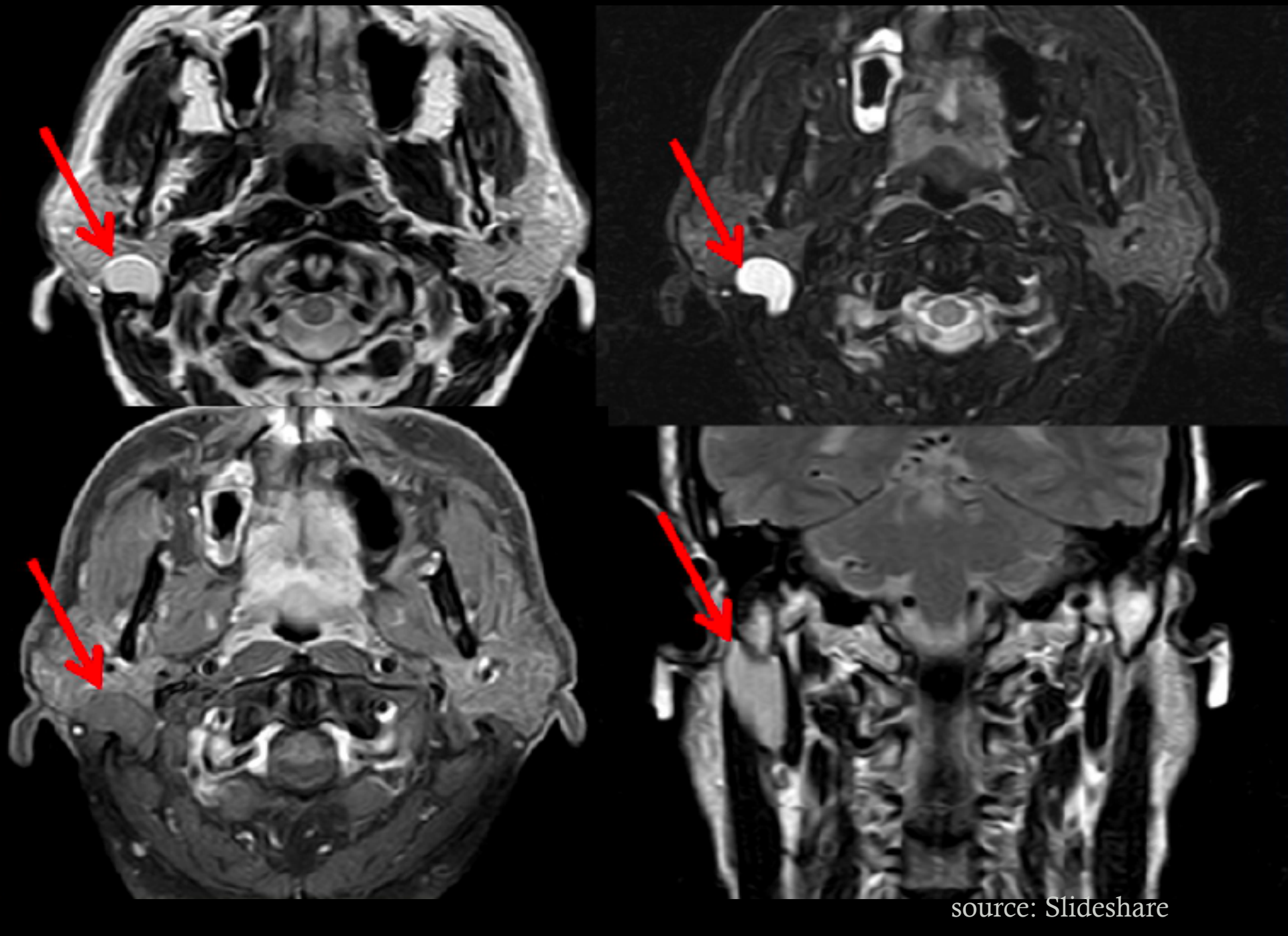


Postero-inferior to auricle



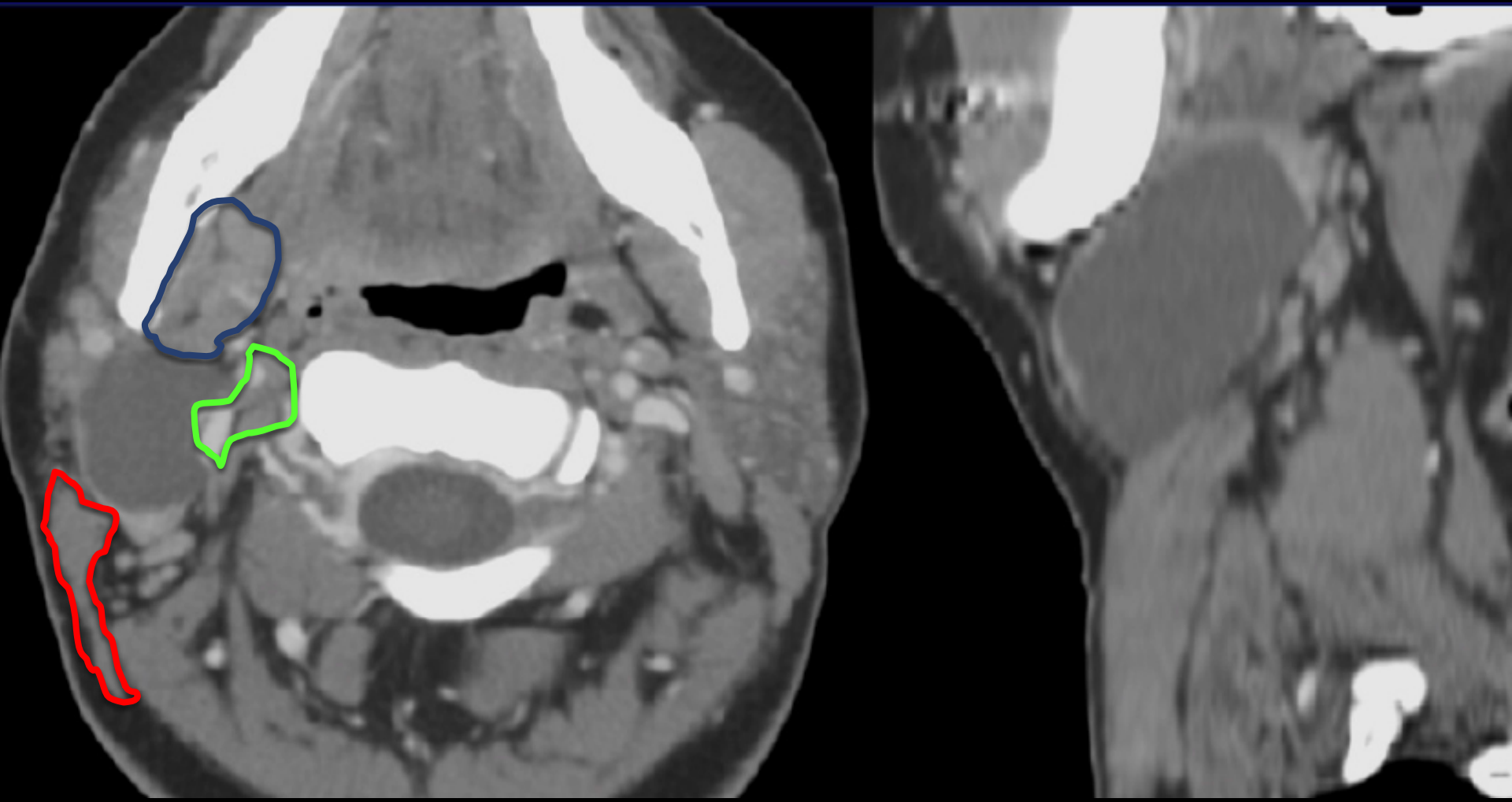
Adjacent to parotid gl./mandible angle

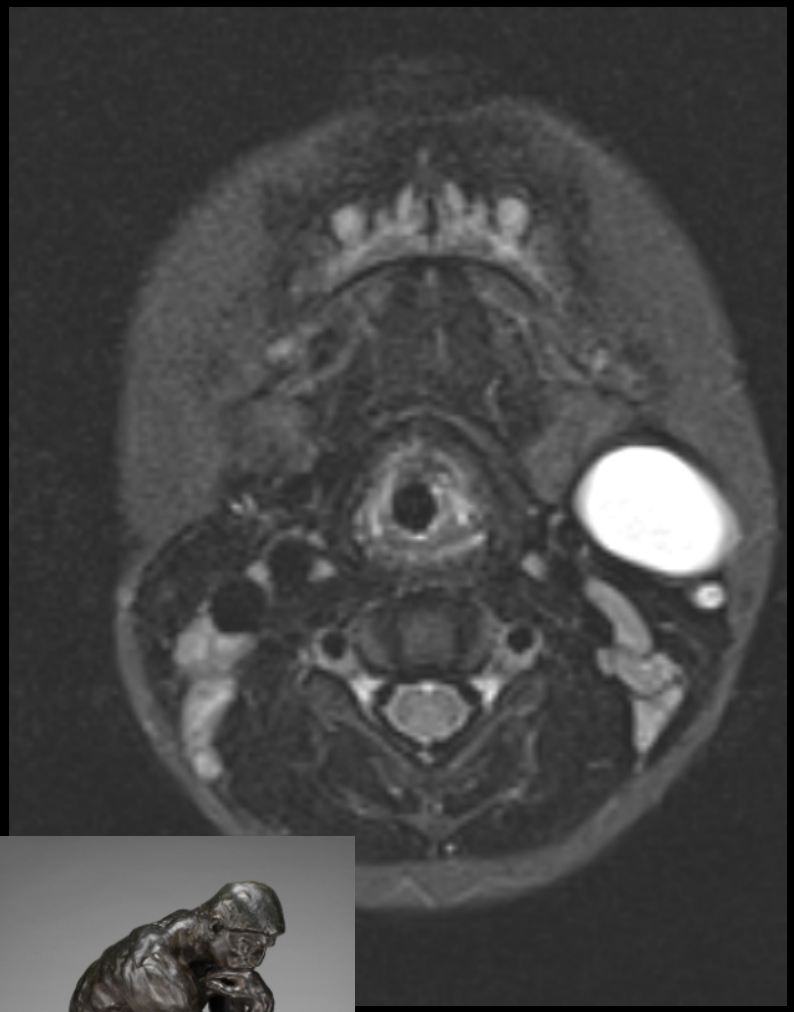
B. Koch 2015

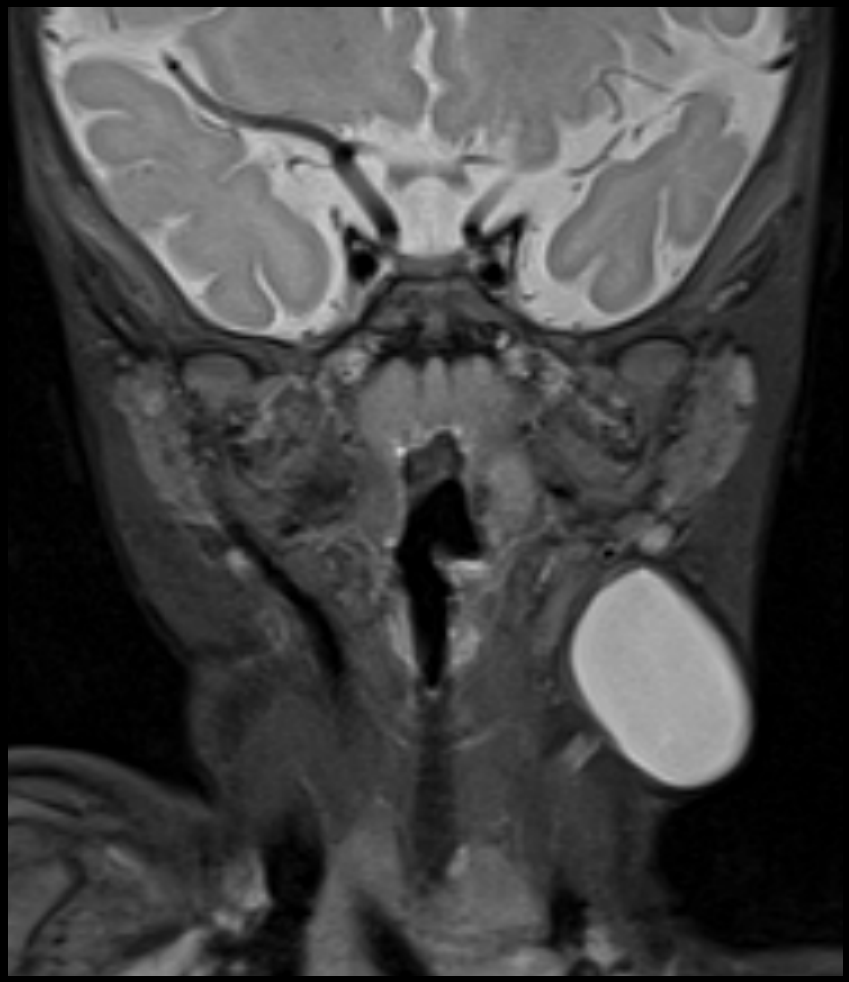


source: Slideshare

Clue for the diagnosis: what are the 3 anatomical bounders of this lesion?



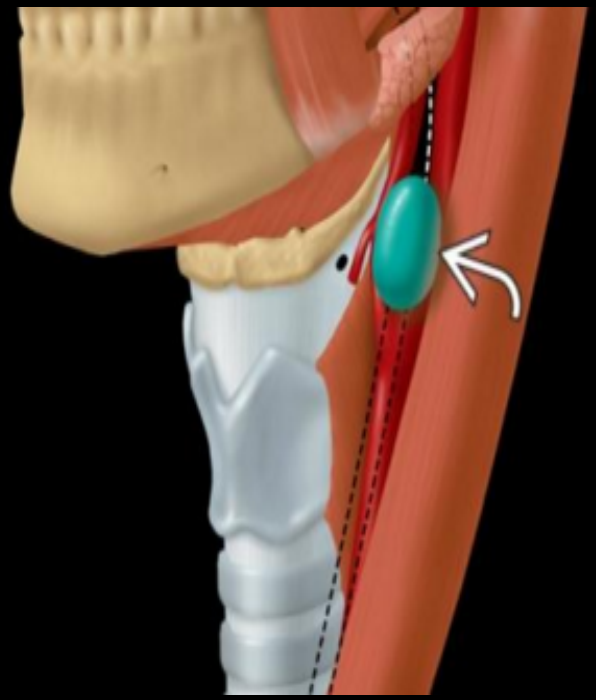




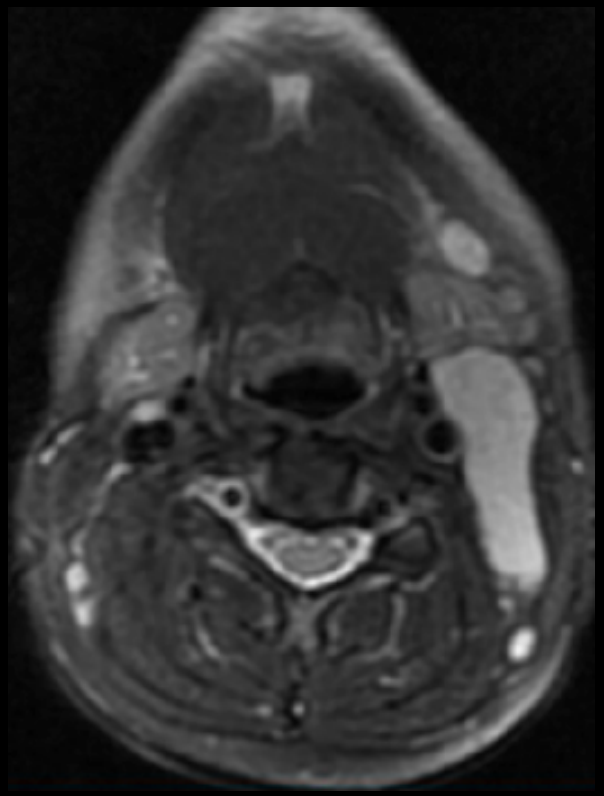
Same location → 2BAA!

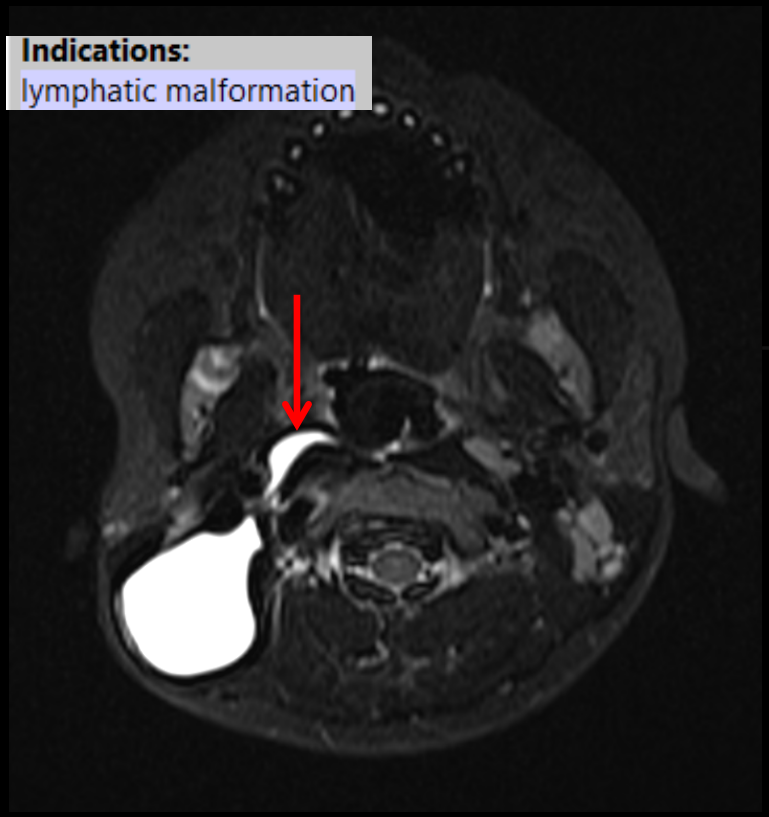
2nd Branchial Apparatus Anomaly

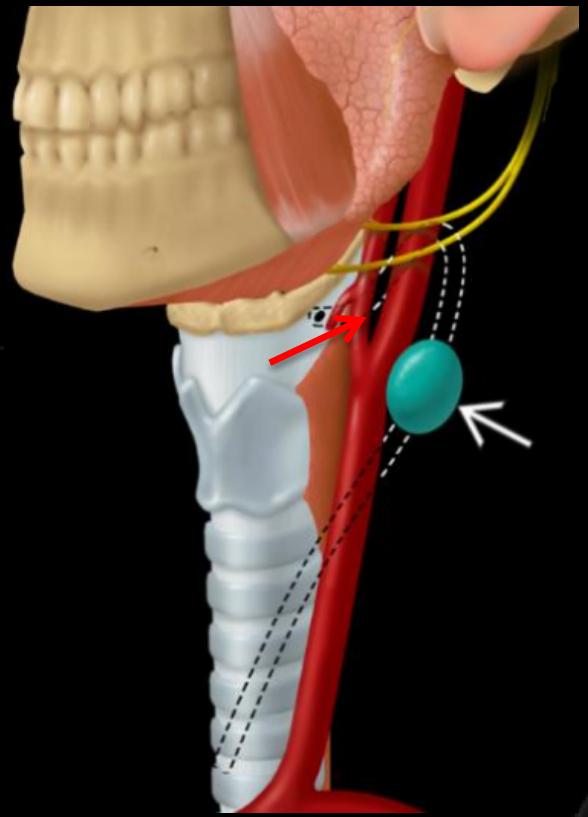
- Typical location: Antero-medially to the SCM (superior 1/3), posteriorly to the submandibular gland, laterally to the carotid space



B. Koch 2015





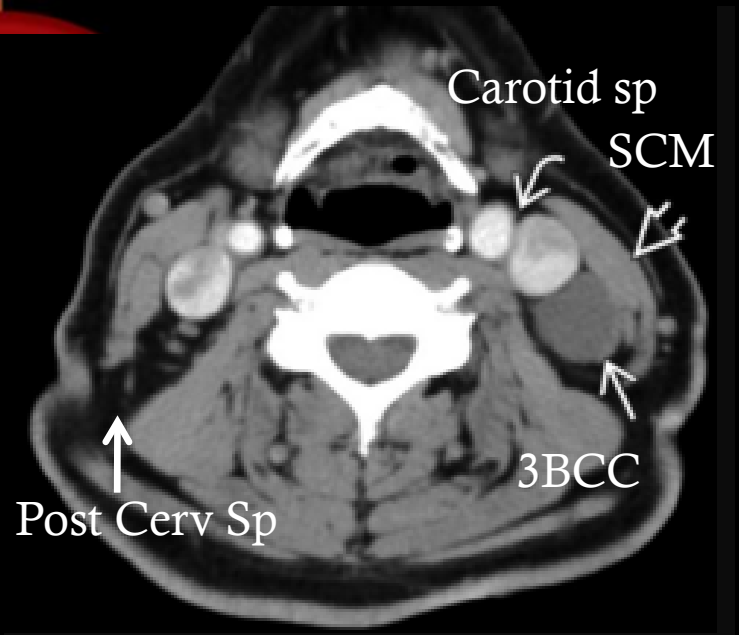


3rd Branchial Apparatus Anomaly

-Medially to the middle 1/3 of the SCM

-Lower than 2nd BCC

-In the **posterior cervical space**



B. Koch 2015

Pt.1 : 21 day-old: infected “mass” left neck

Q: what are the arrows indicating?

Pt.2

Pearl → Clinical presentation as LEFT neck infection!

… controversies…

ORIGINAL RESEARCH

B. Thomas
M. Shroff
V. Forte
S. Blaser
A. James

Revisiting Imaging Features and the Embryologic Basis of Third and Fourth Branchial Anomalies

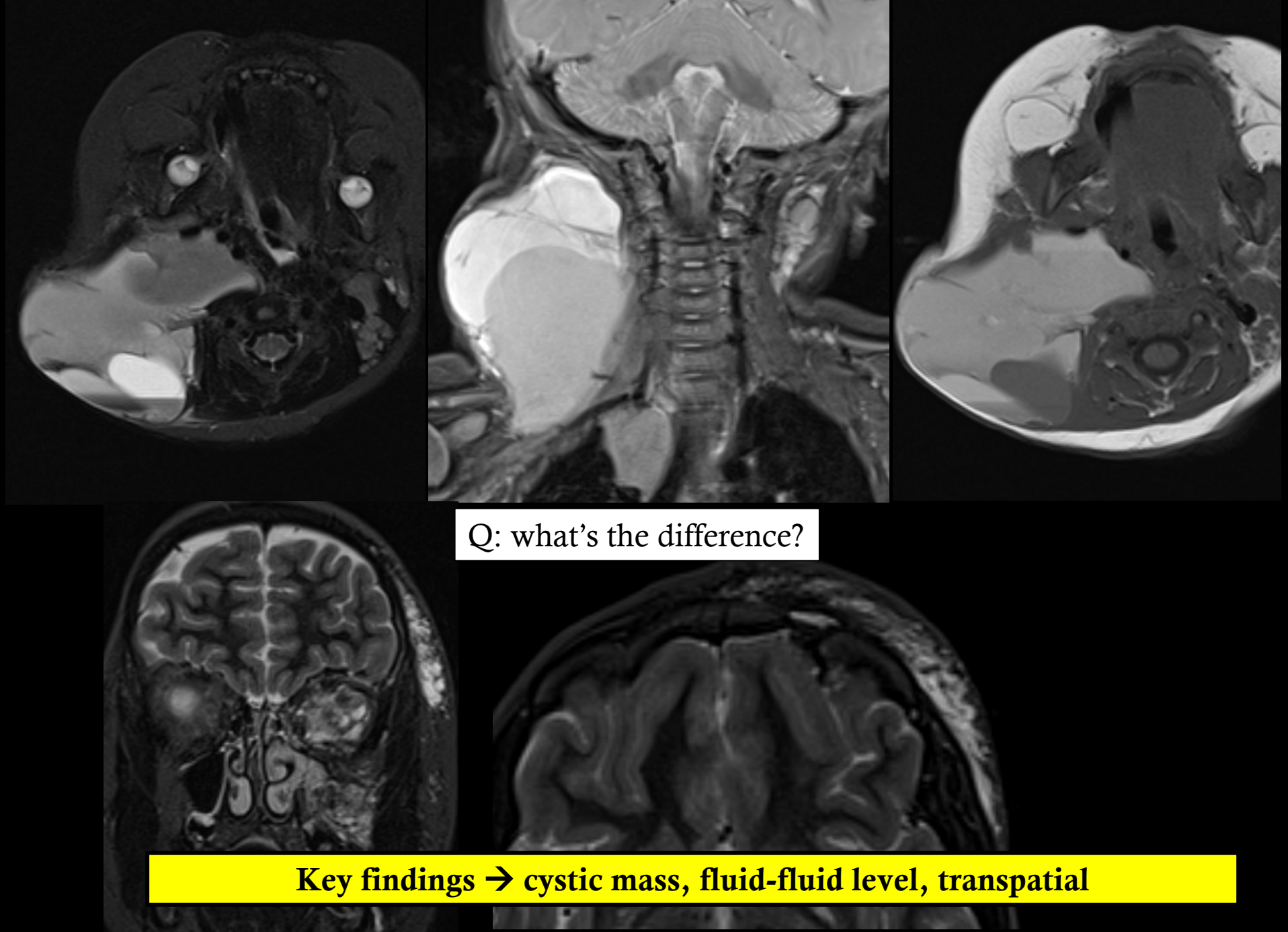
BACKGROUND AND PURPOSE: There is wide discrepancy between common clinical and radiologic presentations of branchial sinuses arising from the pyriform fossa and the theoretic course of third and fourth branchial arch anomalies. The purpose of this study was to revisit the clinical presentations and imaging features of such anomalies in children.

MATERIALS AND METHODS: A retrospective review of institutional and diagnostic imaging data bases from 1998 to 2008 for reported cases of third and fourth branchial cleft anomalies was conducted. Clinical presentation, pharyngoscopy results, and imaging features in all the patients were evaluated. Surgical and histopathology correlation in patients who underwent excision of the tract was also obtained.

RESULTS: Twenty reported cases described as third or fourth branchial apparatus anomalies were identified. There were 12 females and 8 males with a mean age of 84.6 months. The most common presentation was an inflammatory neck mass (18/20, 90%) almost always involving the thyroid gland. Most lesions were on the left side (16/20, 80%). Pharyngoscopy showed a sinus opening at the piriform fossa in 18/20 (90%) cases. None of the cases followed the classic theoretic pathway of third and fourth arch remnants. Histopathology showed tracts lined with pseudostratified squamous epithelium or ciliated columnar epithelium often associated with inflammatory changes in 17 surgically resected cases.

CONCLUSIONS: Branchial sinuses arising from the pyriform fossa often present with an inflammatory neck mass involving the thyroid lobe, most often on the left side. Imaging and surgical findings suggest that they arise from the embryonal thymopharyngeal duct of the third branchial pouch, because they do not follow the hypothetic course of third or fourth arch fistulas.

ABBREVIATIONS: Adj = adjacent; E = epiglottis; H = hyoid bone; Infl neck mass = inflammatory neck mass; L = left side; MRI = MR imaging; Noninfl = noninflammatory; Piriform S = piriform sinus opening; R = right side; SI = signal intensity; TC = thyroid cartilage; TG = thyroid gland; Thyroid Inv = thyroid gland involvement; US = ultrasonography



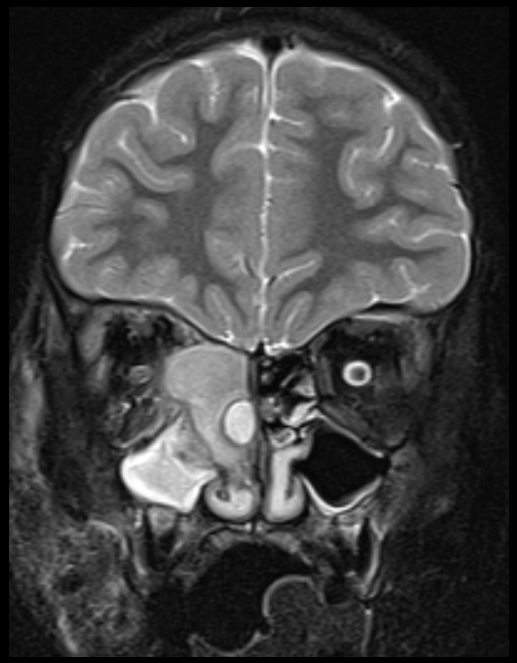
Q: what's the difference?

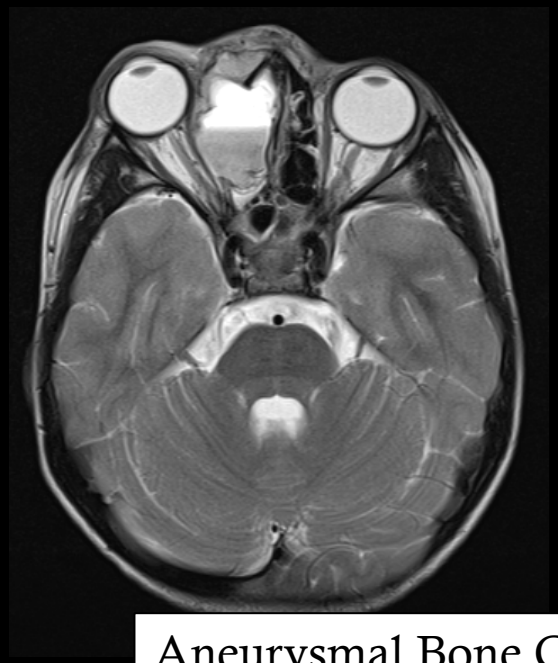
Key findings → cystic mass, fluid-fluid level, transpatial

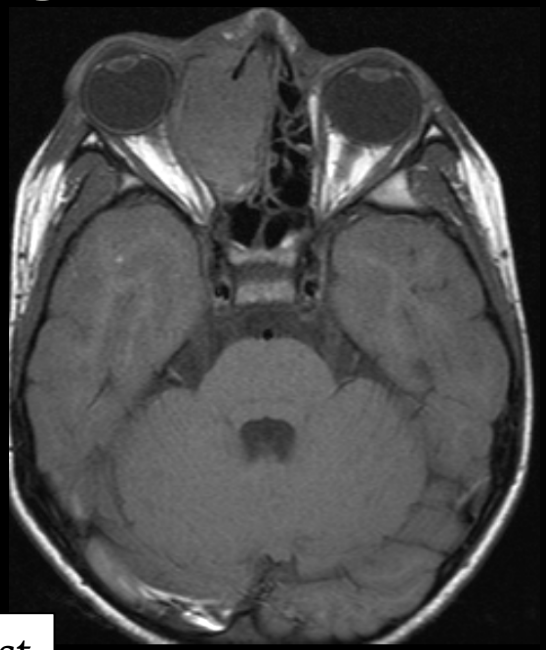
Lymphatic Malformation

- Uni- or multiloculated, non-enhancing, cystic neck mass.
- Micro- and macro cystic
- Often trans-spatial, with fluid-fluid levels (hemorrhage and high proteinaceous components)
- Venolymphatic Malf. : Combined elements of venous malformation & lymphatic malformation (contrast enhancement of the venous elements)

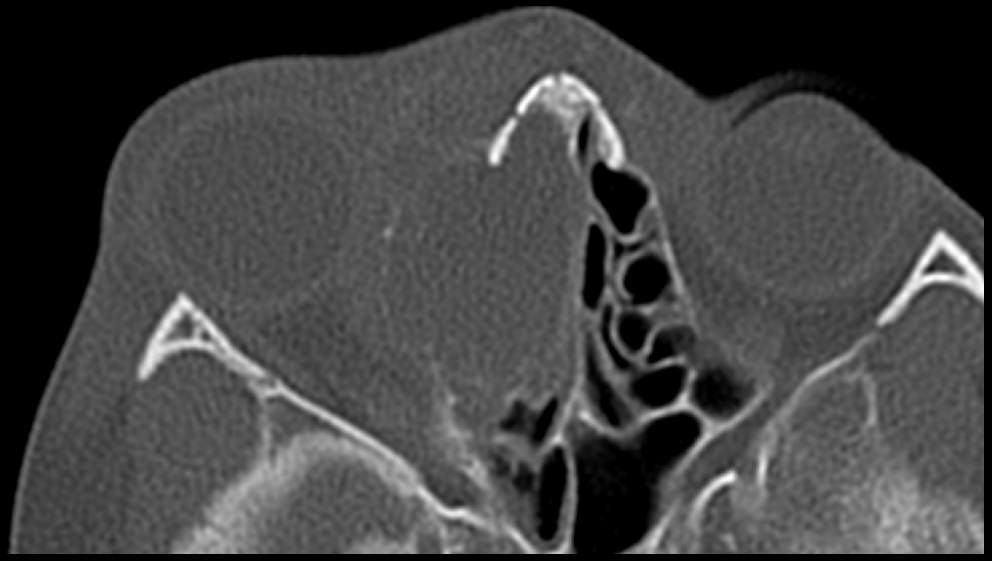
Fluid-fluid levels: Diagnosis?

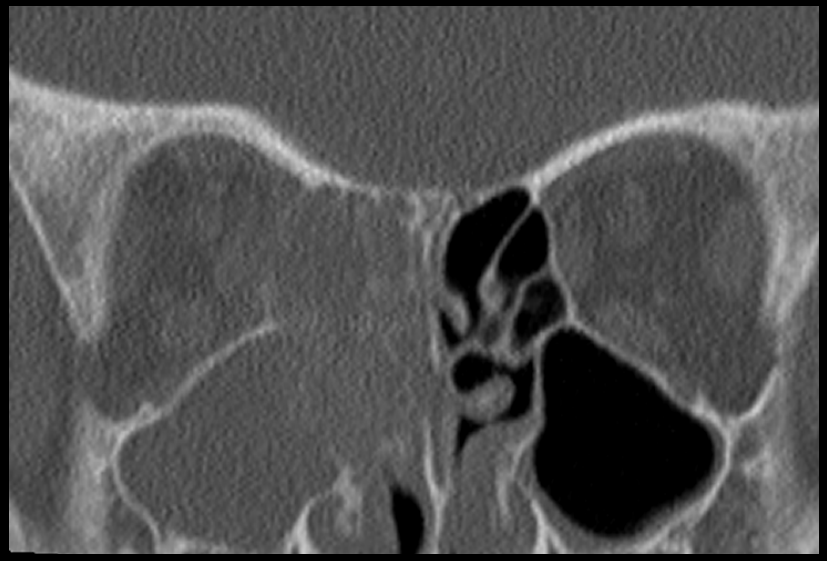


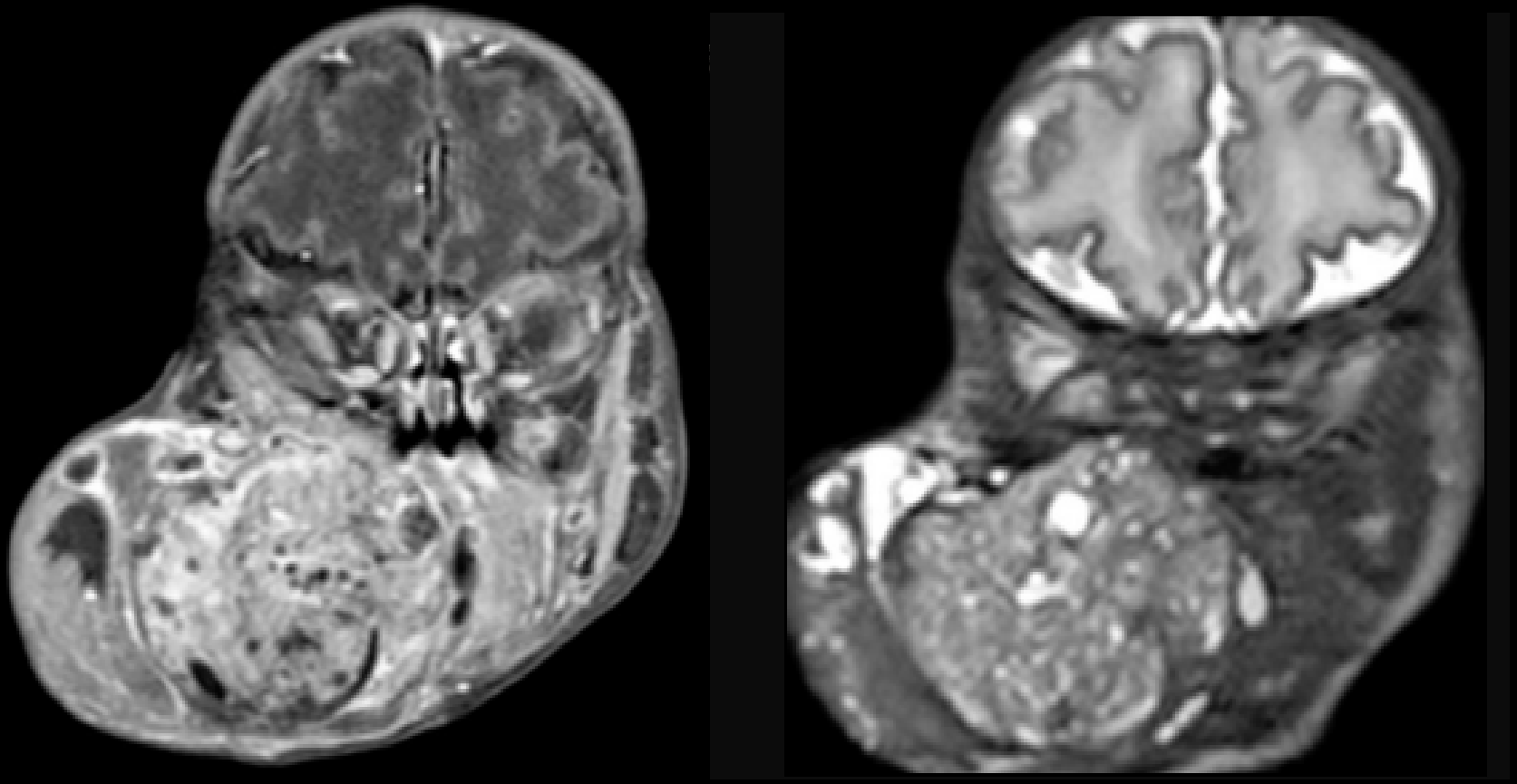




Aneurysmal Bone Cyst

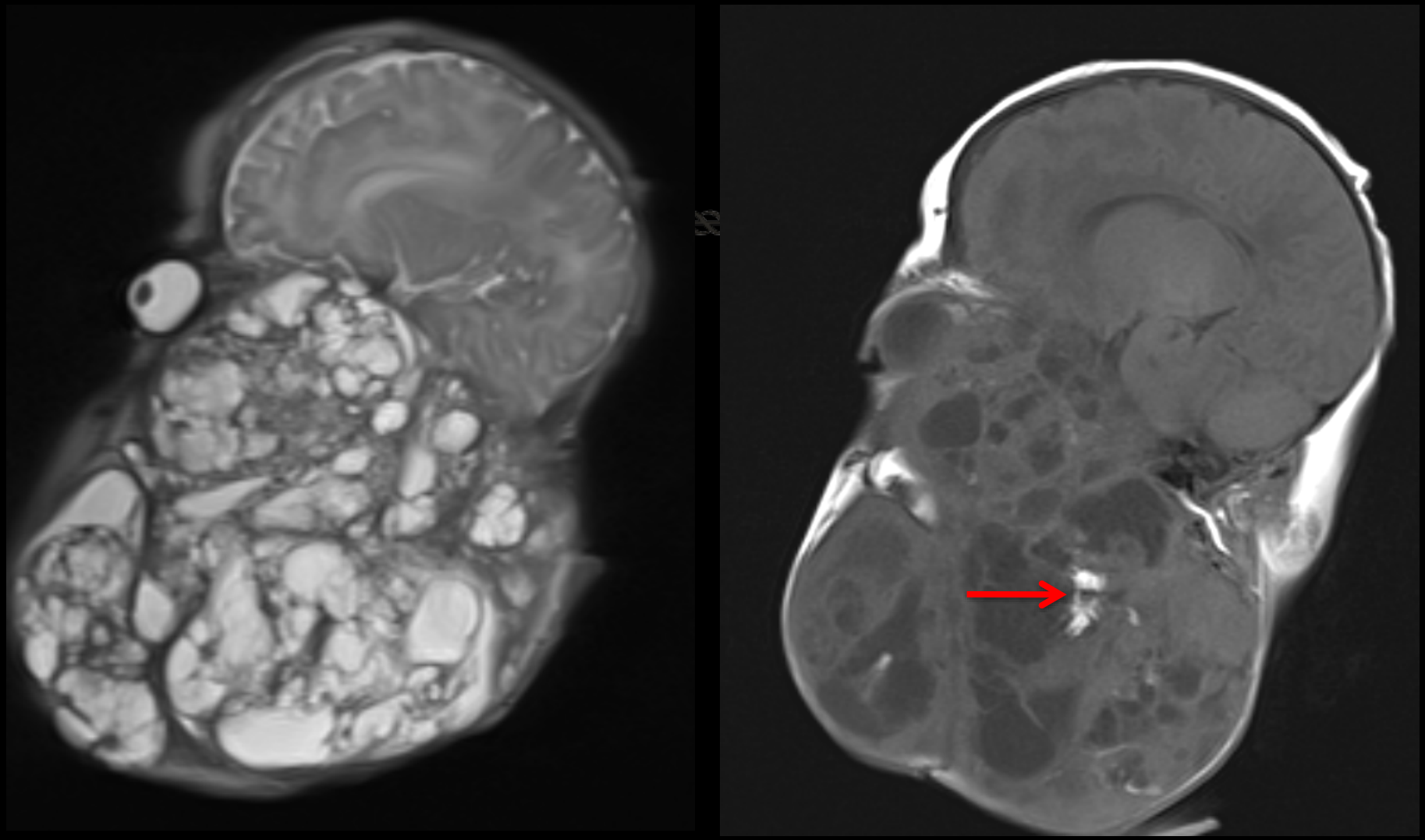






Question: what's the diagnosis?

A. dermoid
B. teratoma
C. Hairy polyp
D. Sarcoma

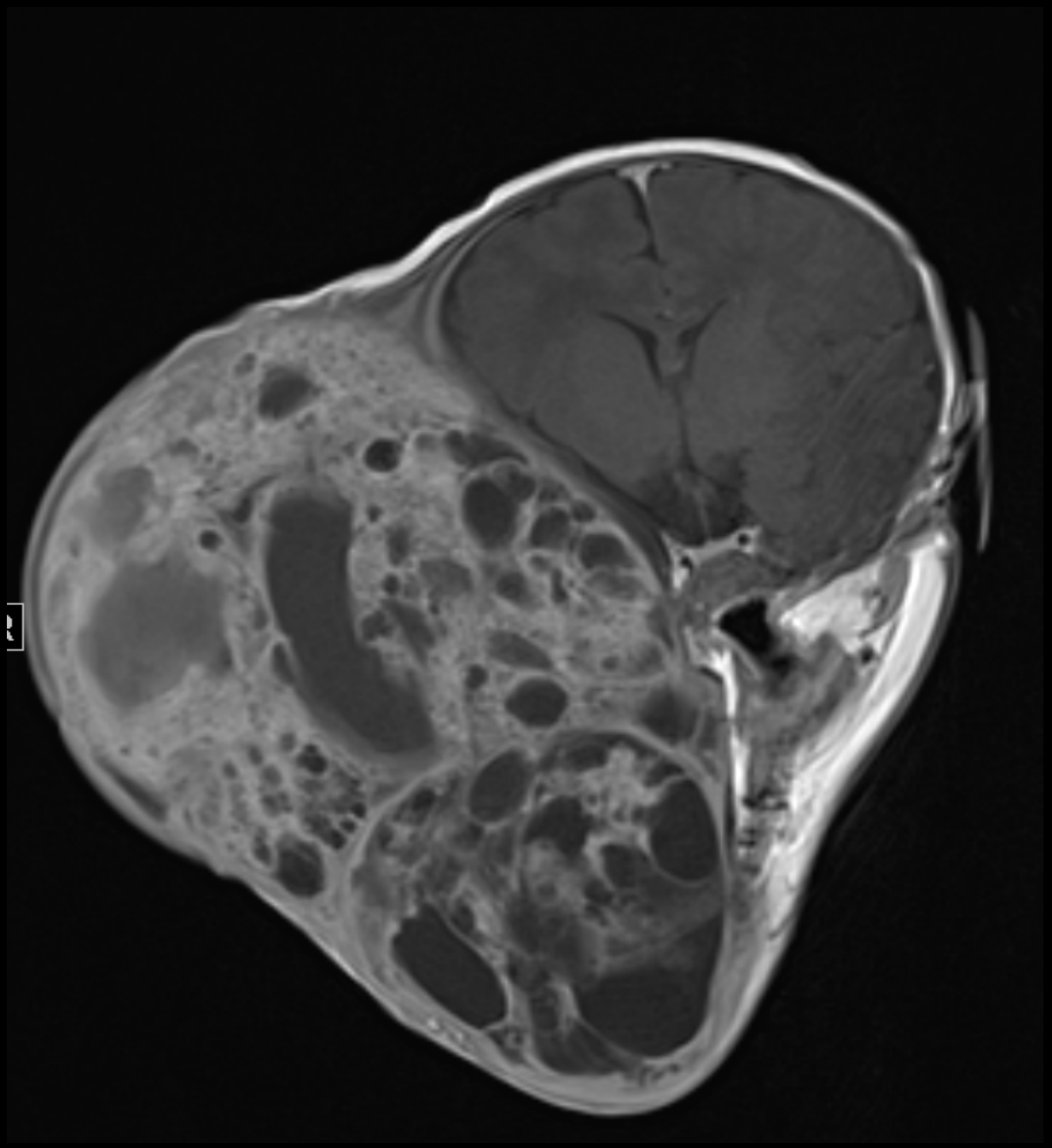


Mixed solid-cystic mass with fat content

Pearl: look for T1 hyperintensity of the fat!

Question: what's the diagnosis?

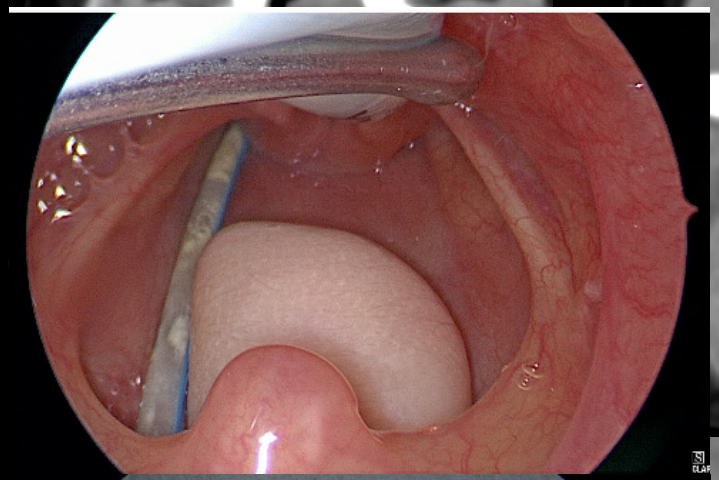
A. dermoid
B. teratoma
C. Hairy polyp
D. Sarcoma

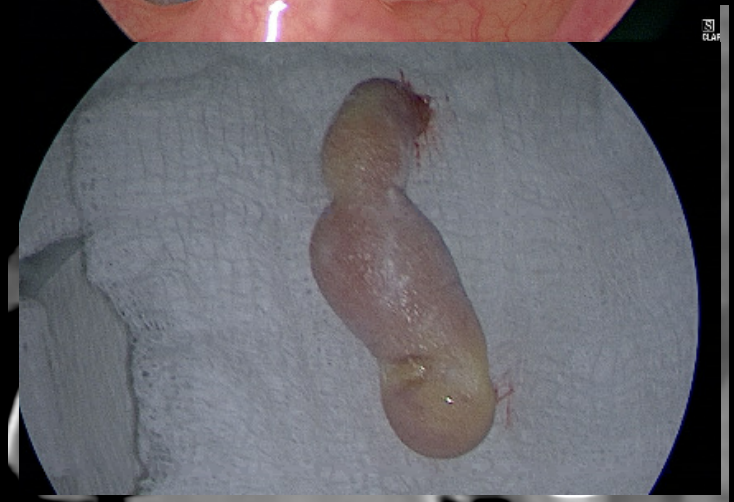


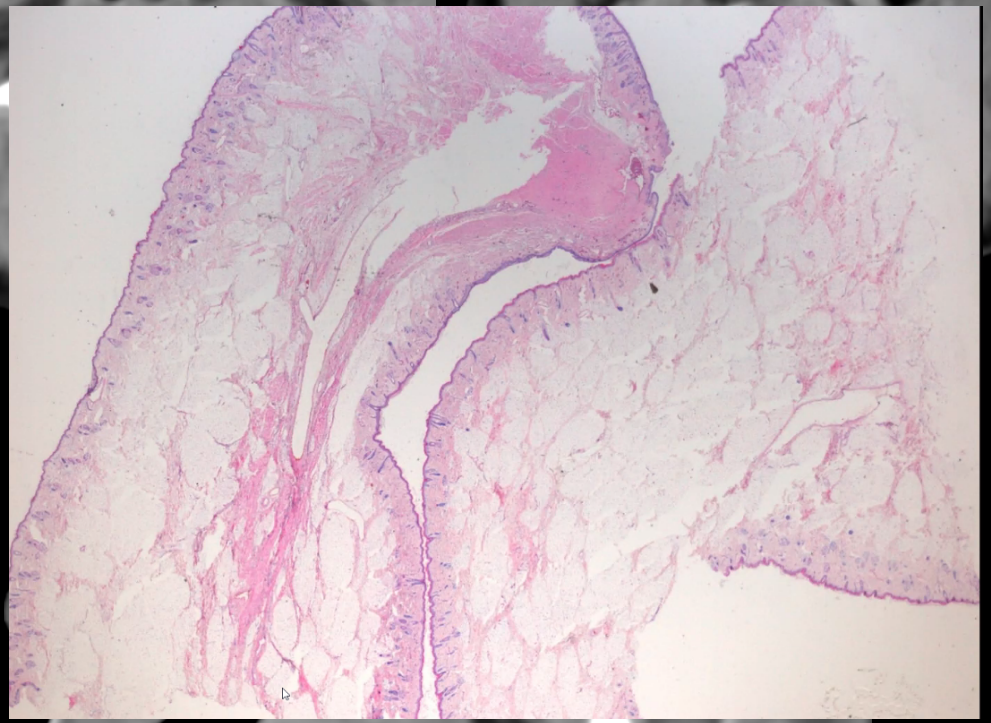
Teratoma

- Anterior neck, off/midline mass containing all 3 germ layers
- **Mixed (cystic and solid) with fat and calcium**
- DD: Lymphatic Malf (fluid with no fat, calcium or solid components), Goiter (homogeneous, respects limits of the thyroid gland)

… let's stay focused on the fat as key finding …







Question: what's the diagnosis?

A. JNA
B. teratoma
C. Hairy polyp (dermoid)
D. Sarcoma

Question: what's the diagnosis?

A. JNA
B. teratoma
C. Hairy polyp (dermoid)
D. Sarcoma

Head and Neck Pathol (2016) 10:213–216
DOI 10.1007/s12105-015-0632-2

SINE QUA NON RADIOLOGY-PATHOLOGY

Hairy Polyp of the Nasopharynx Arising from the Eustachian Tube

Judy Wu[1] · Jefree Schulte[2] · Carina Yang[1] · Fuad Baroody[3] · Daniel Thomas Ginat[1]

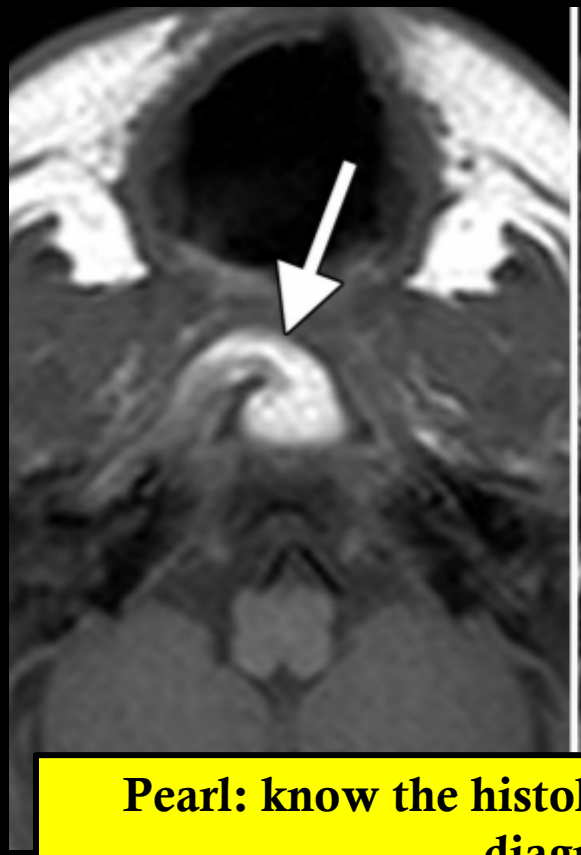
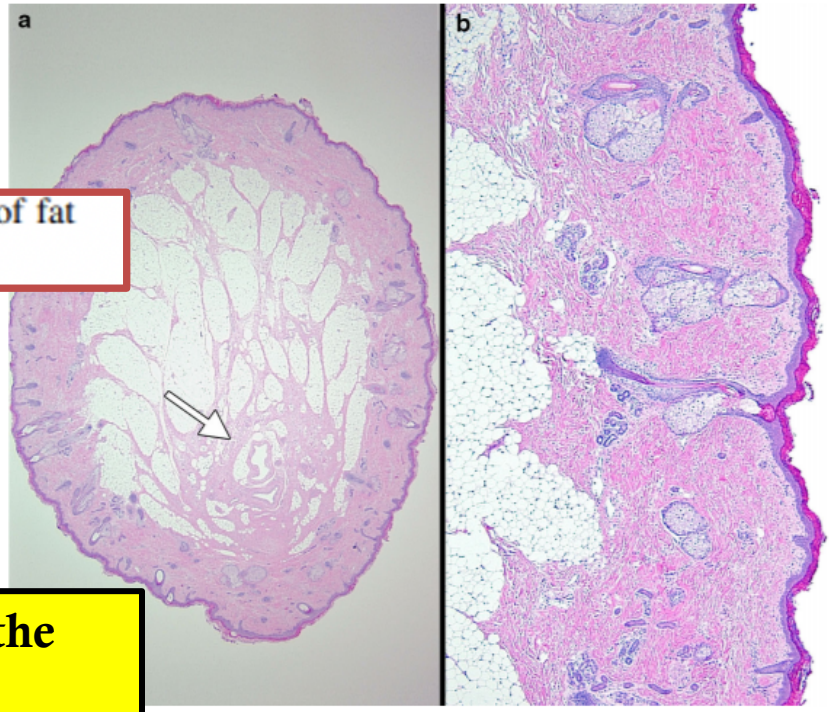


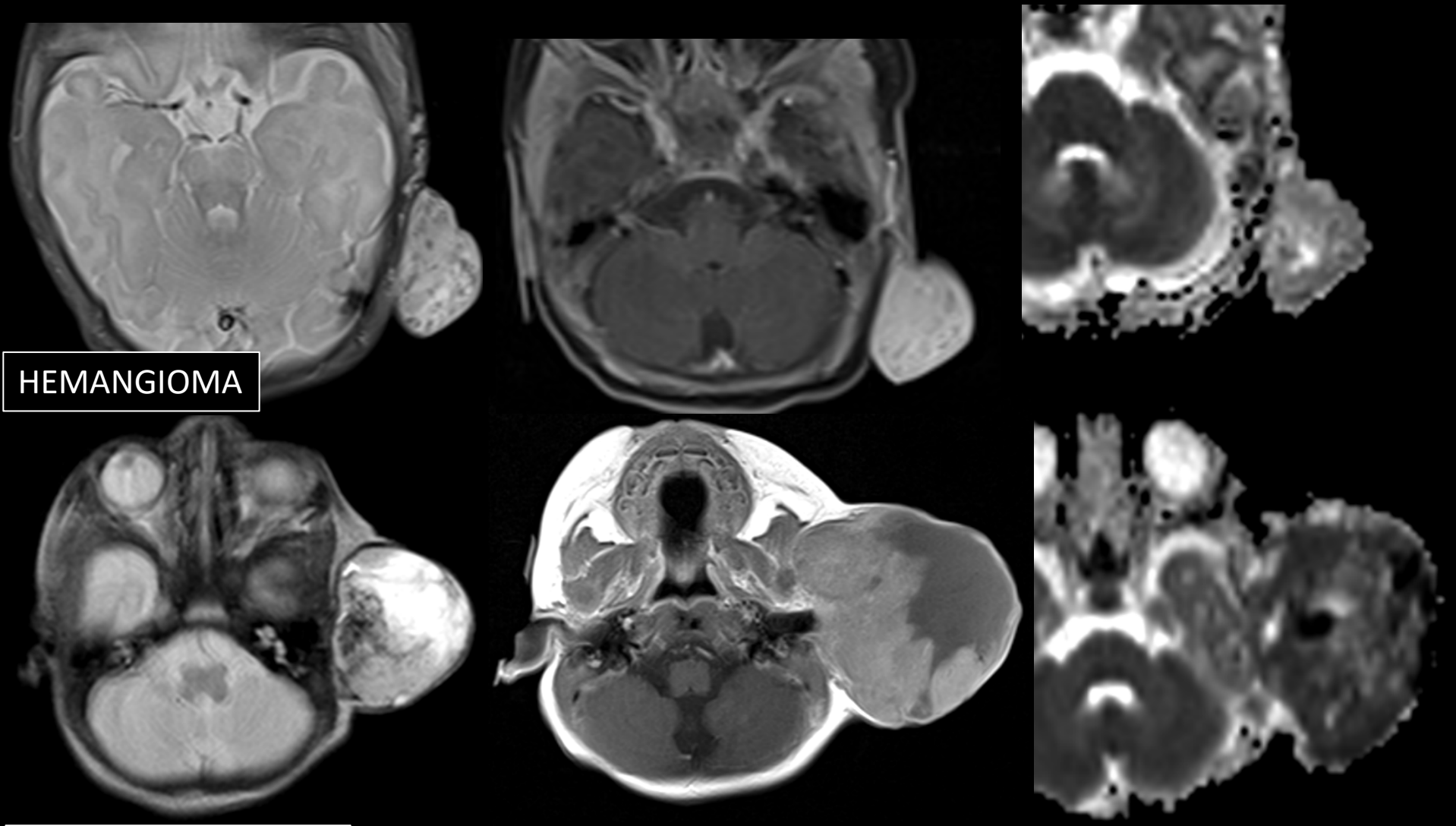
Fig. 4 Hematoxylin and eosin stained cross sectional photomicrographs of the hairy polyp obtained at 1.25X (**a**) and 3X (**b**) original magnification show skin containing multiple pilosebaceous units overlying a mesenchymal core

predominantly comprised of fat and a fibrovascular stalk



Pearl: know the histology, you will guess the diagnosis !

Same (neonatal) age, same location, different imaging characteristics!



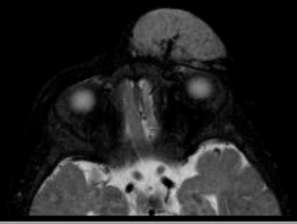
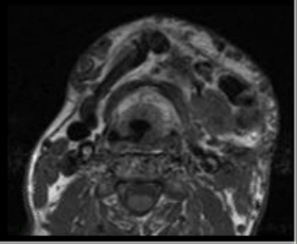
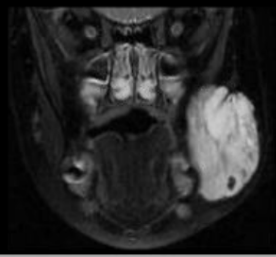
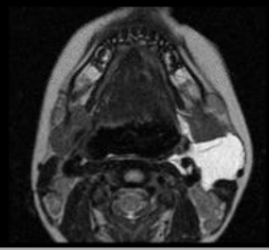
HEMANGIOMA

INFANTILE SARCOMA

Pearl: low ADC, necrosis, heterogeneous enhancement -→ malignant!

Courtesy: Dr. A. Biswas - GOSH

Tips of the Day

	(+) Enhancement	(-) Enhancement
↑ T2 SI	**Hemangioma** • Cellular neoplasm • Triphasic growth • Large vessels in Phase I 	**AVM** • Vascular flow voids • No discrete mass • Bony lysis and overgrowth 
↑ ↑ T2 SI	**Venous Malformation** • Phleboliths • Dynamic enhancement 	**Lymphatic Malformation** • Macrocystic and microcystic • Hemorrhagic fluid levels 

Pearl: IN CASE OF CYSTIC LESIONS: LOCATION LOCATION LOCATION!

Courtesy: D. Shatzkes

Suggested lectures…

Pearls, Pitfalls, and Mimics in Pediatric Head and Neck Imaging

Felice D'Arco, MD[a], Lorenzo Ugga, MD, PhD[b,*]

KEYWORDS

• Head and neck • Pediatrics • Diagnostic imaging • CT • MR imaging

KEY POINTS

- Most of the neck masses in children have some preference for specific locations, which can help in the differential diagnosis. However, there is substantial overlap in possible localizations of different entities, and some masses in children are typically transpatial.
- Identification of one or more key imaging findings in the right clinical context is often enough to reach a precise radiological diagnosis.
- The modality of choice for the study of head and neck masses in children is MR imaging, with protocols that are different from standard brain imaging.
- Clinical presentation, especially the dermatologic findings, together with the patient's age is important for the correct diagnosis, to guide management and predict evolution over time of the lesion.
- Radiologists need to be familiar with the most recent nomenclature of pediatric head and neck masses.

Neuroimag Clin N Am 32 (2022) 433–445

Neuroradiology 2022

Neuroradiology
https://doi.org/10.1007/s00234-022-02950-9

GUIDELINES

Guidelines for magnetic resonance imaging in pediatric head and neck pathologies: a multicentre international consensus paper

Felice D'Arco[1,2] · Livja Mertiri[1,3] · Pim de Graaf[4] · Bert De Foer[5] · Katarina S. Popovič[6] · Maria I. Argyropoulou[7] · Kshitij Mankad[1] · Hervé J. Brisse[8,9] · Amy Juliano[10] · Mariasavina Severino[11] · Sophie Van Cauter[12,13] · Mai-Lan Ho[14,15] · Caroline D. Robson[16] · Ata Siddiqui[2,17] · Steve Connor[2,17,18] · Sotirios Bisdas[19,20] · on behalf of the Consensus for Magnetic Resonance Protocols Study (COMPS) Group

Received: 16 February 2022 / Accepted: 5 April 2022

Abstract

The use of standardized imaging protocols is paramount in order to facilitate comparable, reproducible images and, consequently, to optimize patient care. Standardized MR protocols are lacking when studying head and neck pathologies in the pediatric population. We propose an international, multicenter consensus paper focused on providing the best combination of acquisition time/technical requirements and image quality. Distinct protocols for different regions of the head and neck and, in some cases, for specific pathologies or clinical indications are recommended. This white paper is endorsed by several international scientific societies and it is the result of discussion, in consensus, among experts in pediatric head and neck imaging.

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