Vascular Anomalies: A Cased-based Discussion

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Relevant Disclosure and Resolution

I have no relevant financial relationships or affiliations with commercial interests to disclose.



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All about me!

- Father of 2, husband to another physician • Originally from FW, TX
- Otolaryngology residency in Augusta, GA
- Pediatric fellowship in LR, AR
- Training in...
- ...general pediatric ENT
- ...cleft lip and palate surgery
- ...care for complex vascular anomalies



Why ENT?

- Vascular anomalies may occur anywhere
- Who takes the lead?
- Orthopedics (Peds? Onc?)
- General surgery
 ENT (Peds? H&N?)
- IR

Heme/onc

 $\,$ Vascular anomalies occur $^{\sim}50\%$ in the head and neck, meaning ENT providers have an opportunity to lead the team



• A broad term that encompasses neoplastic and non-neoplastic lesions of blood and lymphatic vessels.

• First, a history lesson...



 ISSVA, was initially founded in 1992 in part to help to clarify the nomenclature of vascular anomalies so that various disciplines could begin to speak the same language.

Beware! Various holdovers from prior to this era persist...

<u>a Coal Madilda: Pathal.</u> 2014 Sep; 18(Suppl 1): S117–S120. doi: 10.4163/0973-020x.141321 Update on the classification of hemangior

Concerning and the Concerning of the Concer



Case #1



5 yo m with persistent skin who underwent course of propranolol in infancy. Not present at birth but rapid growth starting at about 1 m/o, stabilizing at about 6 m/o. Previously was much more protrusive, but seems to have stopped shrinking and stabilized. Lesion involves scalp with minor extension onto forehead, taller than it is wide

- Vertical elliptical incision with extension onto forehead oriented perpendicular to relaxed skin tension lines Very long horizontal incision just behind the hairline, including lots of uninvolved skin to appropriately distribute tension

... serial excision. Almost all surgically-resectable lesions discussed today are benign, can be addressed serially if necessary.

2

4

- Dx: Infantile hemangioma

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0r...

Vascular anomalies

SSVA Classification of Vascular Anomalies @2018 International Society for the Study of Vascular Anomalies Available at 'issva.org/classification' Accessed August 4, 2021



Benign neoplasms

4

Benign vascular neoplasms

Infantile hemangioma



* - Growth is the best clinical factor for delineating between IH and CH

Infantile hemangiomas (IH)

- IHs are the most common tumor of infancy, affecting roughly 1 in 10 <1y
 On histopathology, lesions are GLUT-1 positive (?placental "metastasis"?)
 Lesions most often involve skin and immediate subcutaneous tissues
- Diagnosis is usually clinical, though CT with contrast or MRI might be considered, or trial of beta blockers Growth pattern:
- While some are actually present at birth (leading to confusion with congenital hemangiomas), most IHs appear and start growth phase in the first month of life*
 Growth peaks around 5-6 months
- Long period of involution up to 2-3 years of age
- Because natural progression is toward eventual involution, observation and watchful waiting is the most common recommendation...

acobs AH, Walton RG. The incidence of birthmarks in the neonate. Pediatrics. 1976;58(2):218-222.

Infantile hemangiomas (IH)

- Because natural progression is toward eventual involution, observation and watchful waiting is the most common recommendation...
- But what if we can't just watch and wait for involution? Significant symptoms from: Mass effect Rapid growth and ulceration

Infantile hemangiomas (IH)

Medical therapies
 Beta blockers

Glucocorticoids

Infantile hemangiomas (IH)

Medical therapies
 Beta blockers
 Propranolol – usually PO but also available IV. For patients <8 weeks AGA, inpatient monitoring is recommended by "current" consensus...

Infantile hemangiomas (III) Medical therap Beta blockers Propranolol – monitori , inpatient

Infantile hemangiomas (IH)

Medical therapies
 Beta blockers

- Propranolol usually PO but also available IV. For patients <8 weeks AGA, inpatient monitoring is recommended by "current" consensus, EKG also routinely prior to starting therapy, but some emerging data indicate that this may be unnecessary. (2mg/kg/day is 1//^m therapeutic cardiac dose)
- Timolol topical beta blocker adapted from ophthalmology (glaucoma)
- Atenolol? Cardioselective with possibly lower SE profile. Not as well established. Daily dose
 Glucocorticoids injections can be used to speed progression of involution phase or when urgent reduction in size is indicated (such as in airway hemangiomas)
- Oral preparations can be considered but rarely suitable for long-term use

Infantile hemangiomas (IH)

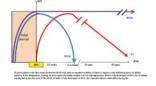
- Surgical excision to resect residuum after involution
- Laser therapy can be considered to make the lesion paler, but is not usually a definitive treatment
- May be useful for large, persisting lesions or for acute bleeding or ulceration

Congenital Hemangiomas (CH)

- Present at birth and do not grow
 Much less common than IH
- Overwhelming majority of CH are rapidly-involuting CHs (RICHs), as opposed to very rare non-involuting CH (NICH)
- Congenital lesions have a much broader differential may require imaging to confirm diagnosis depending on location
- Later in life, patients with NICH may complain of pain in the location of the lesion, prompting consideration of surgical resection
- No known medical therapies for treating CHs

IH vs CH

- IHs grow with time, CHs do not
 IHs are much more common!
- Morphologically indistinctHistopath: IH +ve for GLUT-1
- Diagnostic course of propranolol (or atenolol?) can be considered if clinically necessary to differentiate – beware of missing a sinister diagnosis



Vascular anomalies

Selmetti, C. Vascular Birthmarks: A hidden world behind a word. Indian J of Ped Derm. 2018, Jan(19)1:1-8.



Vascular malformations

Distinct from vascular tumors

Often congenital lesions, arising from aberrant lymphatic, arterial, venous or capillary growth

Neoplasms

Malignant neoplasms (Exceedingly rare)

Locally-aggressive neoplasms Kaposiform hemangioendo-thelioma (KHE) Juvenile Nasal Angiofibroma (JNA)

Benign neoplasms

- Infantile hemangioma Congenital hemangioma Rapidly-involuting congenital Non-involuting congenital Partially-involuting congenita ital hemangioma (nuch) hemangioma (NICH)

/A Classification of Vascular Anomalies ©2018 International Society for the Study of Vascular Anomalies Available at a org/classification / Accessed August 4: 2021

Capillary malformations (CM)

Congenital, well-defined vascular macule or patch (non-elevated discoloration)

Most common head-and-neck vascular malformation
 AKA "port-wine stain"

- · Dilation or ectasia of dermal capillaries
- Dialoutor establish of definited capitalities
 Unless a mixed lesion with other vascular malformations, CMs do not protrude from the skin but the surrounding skin may be slightly thickned, do not bleed significantly
 Functional limitations are uncommon but may be disfiguring
 Designed addising lightly discussed to act acquired
- Dx is usually clinical. Imaging usually not required



Malformations Capillary malformations

Capillary malformations (CM)

- Large "segmental" capillary malformations should prompt workup for Sturge-Weber syndrome (MRI, ophthalmology referral)
- Large CM (usually centered on the ophthalmic division of trigeminal nerve) Glaucoma
- Leptomeningeal vascular malformation
- May also occur in vascular anomaly syndromes including Klippel-Trenaunay syn (KTS)



Nevus simplex, or medial CM

- Light pink skin lesion located anywhere in continuity from glabella-scalp-midline back-sacrum, often Vshaped on the forehead, may involve nose or medial eyelids
- This lesion is histologically indistinct from capillary malformation, but more mild
 Isually lightens with time and does not require
- Usually lightens with time and does not require treatment
- $^{\circ}$ May be treated if still persistent after age $^{\sim}2$



CM management

- Decision to intervene depends on patient and parent preference mostly a cosmetic concern, few functional limitations
- Medical therapy Sirolimus (rapamycin)
- Inhibits mammalian target of rapamycin (mTOR)
- Oral or topical preparations
 If systemic, requires monitoring, dose titration, prophylactic abx (p. carinii infection)
- Laser therapy

Radiolab May 21, 2021

The Dirty Drug and the Ice Cream Tub

Play + 46 min



ies ©2018 International Society for the Study of Vascular Anomalies Available at

1

Neoplasms

Malignant neoplasms (Exceedingly rare)

21 yo f with extensive surgical history including previous tracheostomy (now decannulated), tongue reduction, multiple open neck surgeries who presents to clinic with known history of lymphatic malformation and progressive oral pain and snoring

No known history of sclerotherapy. Repeat MRI with involvement of oral tongue, BoT, and perimandibular tissue What treatment options should we offer? Why is this different from previous patient?

- Injection sclerotherapy and oral tongue coblation for mucosal involvement

Completed third round of sclerotherapy (often performed in series)

-Microcystic LM

- Rarely a great candidate for excision

Locally-aggressive neoplasms Kaposiform hemangloendo-thelioma (KHE) Juvenile Nasal Angiofibroma (JNA)

Benign neoplasms
 Infantile hemangioma
 Congenital hemangioma
 Rapidy-involuting congenital
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 Partiality-involuting congenital

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Malformations Capillary malformations

Lymphatic malformations (LM) Macrocystic LM Microcystic LM (Mixed LM)

Lymphatic malformations (LMs)



Lymphatic malformations (LMs)

For small isolated lesions with high confidence in the diagnosis and minimal functional limitations, watchful waiting may be an appropriate approach

- Spontaneous regression has been demonstrated to be more common in head and neck lesions that are lateralized, infrahyoid and macrocystic (Secondary infection can induce involution in some lesions)
- Unfortunately, lateralized, infrahyoid macrocystic lesions are also more amenable to surgical resection and sclerotherapy than higher, medial microcystic lesions
- Surgical excision of primarily macrocystic lesions, or of macrocystic component of mixed lesions
- · Sirolimus (usually for extensive or multifocal lesions, esp as result of syndrome)
- Sclerotherapy

IIIa-Velez J, et al. Active Observation as an Alternative to Invasive Treatments for Pediatric Head and Neck Lymphatic bimations. Laryngoscope. 2021 Jun;131(6):1392-1397.

Sclerotherapy for vascular anomalies

- "Scarring therapy" for lymphatic, venous or arteriovenous malformations
- Chemical initiating includes in and/or around vascular anomaly to induce scarring of vessel channel and/or surrounding tissue. Reduces burden of disease/limits lesion growth
 In pediatric patients, virtually always under anesthesia for better control of delivery
 Sometimes necessitates ultrasound or other image guidance (IR)

- In venues or AV malformatics actions requiring laser therapy, may be performed in conjunction (surgeon)
 Specific agents (increasing pain/inflammation, some feel this increases efficacy)
- Bleomycin antineoplastic agent
 Doxycycline antibiotic
- OK-432 (streptococcal antigen developed and used in Japan) Sodium tetradecyl sulfate (STS)
- Ethanol

Sclerotherapy for vascular anomalies

- As we will discuss, indications include:

LMs
VMs
AVMs

- IR (justifiably) feels it is critical to directly inject the lesions without any "spill" around the malformation
- However, microcystic LMs are known to improve from injection sclerotherapy, where the injection is both in and around the lesion

- 10 yo f with h/o progressive R facial swelling for 5 years, previously diagnosed as a parotid hemangioma and parotid lymphatic malformation. Now with progressive right facial pain/tenderness.
- Able to induce acute swelling with jaw clenching or Valsalva. CT neck with contrast
- Venous malformation of parotid
- Tx options
- Schurd and the second s







- 8 yo male who was incidentally found on exam to have purple staining of the right aspect of the oropharynx
- Asymptomatic
 MRI confirmed venous malformation involving right oropharynx and pterygoid plexus
- Tx Do nothing?



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Venous malformations (VM)

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Venous malformations (VM)

- Dilated venous channels with (often a network of) thin-channeled walls and abnormal smooth muscle
 Slow flow of blood with poor drainage, resulting in expansion of the lesion to invade or impinge on normal tissue
- Unlike LMs, VMs do not regress spontaneously
- Unlike LMS, VMS do not regress spontaneously
 Multimodal distribution of presentation –
 congenital, early puberty, pregnancy
 Women with history of venous malformation (in
 remission or active disease) should be on
 estrogen-sparing contraception



Venous malformations (VM)

Symptoms depend on location of the lesion and resultant mass effect

- Mucosal surfaces may bleed, cutaneous bleeding is less common Lesions may be painful when localized thrombosis occurs
- ("phleboliths")
- High risk of LIC in large lesions D-dimer is often elevated
- Hematology involved to assist with anticoagulation (to reduce pain while definitive management is rendered, may also prevent thrombus perioperatively for IR sclero)



VM treatment

- Similar to lymphatic malformations:
 Sirolimus for residual/diffuse lesions
- Sclerotherapy
- Surgical resection often immediately following IR embolization
- Laser
- Generally, surgery can/should only proceed once overlying epithelium has been cured of involvement with laser treatments – reduces morbidity of reconstructive procedures, VASTLY improves cosmetic result



Laser therapy for vascular anomalies

Selective photothermolysis

- Takes advantage of differential between Ox-Hgb and Deox-Hgb absorption spectra
- Pulsed-dye laser for "red" lesions (hemangiomas, AVMs, CM)
- ND:YAG for "blue" lesions (VM, CM, AVMs)
 ND:YAG has deeper penetration (less melanin absorption)



Laser and Sclerotherapy for vascular vascular malformations

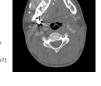
- Often, combined laser/sclerotherapy 3-4 sessions are scheduled q3 months, then reassess for response
- Laser is important for reducing morbidity of surgical resection (epithelium-sparing surgery)
- Sclerotherapy may also be delayed until after resection (resection may be more challenging after sclerotherapy, especially if done "blindly")

13 yo m with known history of mandibular AVM presents to ED with profound oral cavity hemorrhage 6 months after undergoing coil embolization which provided temporary control of the same. CTA demonstrates mandibular marrow space AVM with extension into perimandibular lower lip.

Tx options?

- Stop the bleeding. Ideally with a selective temporizing embolization and/or laser

- Curative resection after embolization (radical resection vs curettage?)



Vascular anomalies

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Macrocystic LM Microcystic LM Venous malformations (VM) Arteriovenous malformations (AVM)

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Malformations Capillary malformations

Lymphatic malformations (LM)

Arteriovenous malformations (AVM)

- Most difficult vascular anomaly to treat, thankfully also the rarest
- May be thought of as arteriovenous fistula, with a nidus of abnormal capillaries that shunt oxygenated blood in a high-flow fashion
- Because of shunting, surrounding tissue is chronically paradoxically ischemic, leading to neovascularization and growth of the lesion, as well as enlargement of feeding/draining vessels
- Symptoms usually related to bleeding, which can be hemodynamically significant and life-threatening late stage includes high-output HF



Arteriovenous malformations (AVM)





AVM treatment options

- Laser therapy for epithelial involvement (same rule regarding epithelium-sparing surgery for VM applies here)
- Sclerotherapy
- Embolization/resection if focal and surgically accessible. For arterial embolizations, wake patient up afterward for neuro exam pre-operatively (defensive medicine)
- For acute bleeding, laser therapy may be used, esp for mucosal bleeding
- Embolization should be super-selective large feeding vessels should be spared if
 possible for future access. AVM will virtually always recruit from adjacent vasculature
 later.

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Extracranial arteriovenous malformations: natural progression and recurrence after treatment

AVM prognosis

PNEC 201968 OC NUMPRESENTATION PAGE 201968 OC NUMPRESENTATION Largest case series on extracranial AVMs, from Boston Children's Hospital vascular anomalies center

Extremely high rate of recurrence, timeframe dependent on the lesion stage and mode of treatment

No data on frequency of using laser or sclerotherapy, reduction of recurrence/progression rate Measuring success based on recurrence rate alone may be wrong-headed? Table 4. Time to Recurrence after Treatment of Arteriovenous Malformation*

cation ces) (%)

plog-Head and Tech Durgery CETED 142, 184 188 NAL RESEARCH-HEAD AND NECK SURGER Clinical course of arteriovenous malformations of the head and neck: A case series Gresham T. Richter, MD, and James Y. Suen, MD, Little Rock, AI

AVM prognosis

This terrible prognosis is contradicted by other, smaller case series demonstrating drastically better results

10-patient case series of AVM patients who'd failed prior treatment with other centers, all with improved or stable disease and improved QoL at last f/u

Quite prone to publication bias!

Limited resection may afford lower morbidity especially if revision surgery is so often necessary.

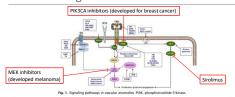
AVM treatment paradigm has shifted from treating to cure toward managing the disease with expectation of repeat therapy in a more controlled setting



systematic review of pediatric mandibular rteriovenous malformations Sys

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Medical management of vascular malformations

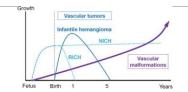


. dams DM, Ricci KW. Vascular Anomalies: Diagnosis of Complicated Anomalies and New Medical Treatment Options. H Incol Clin North Am. 2019 Jun;33(3):455-470. doi: 10.1016/j.hoc.2019.01.011. PMID: 31030813.

Neoplasms Malformations Capillary malformations Malignant neoplasms (Exceedingly rare) Lymphatic malformations (LM) Macrocystic LM Microcystic LM Locally-aggressive neoplasms Kaposiform hemangioendo-thelioma (KHE) Juvenile Nasal Angiofibroma (JNA) Venous malformations (VM) Benign neoplasms Infantile hemangioma Congenital hemangioma Rapidly-involuting congenital Non-involuting congenital Partially-involuting congenital Arteriovenous malformations (AVM) ma (NICH) Mixed lesions (Beyond scope) nangi A Classification of Vascular Anomalies ©2018 International Society for the Study of Vascular Anomalies Available at a org/classification* Accessed August 4: 2021

Tissue hypertrophy

- Many vascular anomalies can present with surrounding tissue hypertrophy
- Etiology may be multifactorial
- Somatic mutation involving surrounding tissue?
 Increased blood flow to involved area promoting asymmetric growth?
- Known component of vascular anomaly syndromes
 Klippel Trenaunay (KTS), Diffuse capillary malformation with overgrowth (DCMO), many others Involved extremities (especially lower extremities) may require growth plate arrest (epiphysiodesis) during adolescence
- Surgical approach may be complicated by the presence of the lesion, requiring multidisciplinary coordination



Conclusion and Clinical Pearls

- Infantile hemangiomas (IHs) are the most common vascular anomaly, and follow a predictable pattern of early growth and late involution.
- IHs that require treatment often respond well to propranolol during the early proliferative phase. Atenolol is an emerging treatment option that may have fewer side effects.
- IHs grow, congenital hemangiomas (CHs) do not. CHs are overwhelmingly most likely to be rapidly-involuting CHs (RICHs) that also will likely not require treatment.
- Vascular malformations, on the other hand, typically do not involute spontaneously, with the exception of select lymphatic malformations.

Additional Resources

ISSVA Classification of Vascular Anomalies ©2018 International Society for the Study of Vascular Anomalies Available at "issva.org/classification".

 Head and Neck Vascular Anomalies: A Practical Case-Based Approach. Richter GT, Suen JY. 2015. Plural Publishing, San Diego, CA.

Very happy to chat about any referrals for vascular lesions with unclear diagnoses, and happy to see patients vascular lesions outside head and neck or in adults!
 Many referrals are funneled through orthopedics or through dermatology, but if wait times are long or urgency is critical, please feel free to contact me:

• Colin-fuller@ouhsc.edu

• 817-454-1615 (cell)

Thank you!



Questions