MRI of head and neck vascular anomalies: pearls and pitfalls

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Learning objectives

- Illustrate the typical MRI features of head and neck vascular anomalies
- Describe the commonly encountered imaging pitfalls
- To review the value of different MRI techniques in the diagnosis of vascular anomalies
Background

Soft tissue vascular anomalies comprise a wide spectrum of abnormalities affecting blood and lymphatic vessels. A binary classification system has been adopted by the International Society for the Study of Vascular Anomalies dividing vascular anomalies into tumors and malformations based on their clinico-pathological features [1,2]. Infantile hemangiomas are the most common tumor of infancy and have an anatomic predilection for the head and neck region [3, 4].

Accurate differentiation between vascular tumors (hemangiomas) and vascular malformation is crucial, as their management is completely different [5]. Different types of vascular anomalies are usually diagnosed based on their clinical presentation. Imaging modalities are reserved for alarming or atypical lesions, and prior to surgery or imaging guided procedures. In addition, imaging can aid in the follow-up to assess the response to therapy [5,6].

Segmental facial vascular anomalies can be an indicator of one of the two famous neurocutaneous syndromes: Sturge-Weber and PHACE syndromes. MRI is the modality of choice to detect the associating intracranial abnormalities. Sturge -Weber syndrome is characterized by facial capillary malformation in the distribution of ophthalmic branch of trigeminal nerve in association with leptomeningeal angiomatosis and choroidal angiomia [7]. PHACE syndrome is a neurocutaneous syndrome of Posterior fossa malformation, segmental Hemangioma, coarctation of the Aorta, Cardiac and Eye abnormalities. The hallmark of PHACE syndrome is large segmental hemangioma of trigeminal dermatome pattern [8]. It can be mistaken for a capillary malformation, and hence, clinical misdiagnosis of Sturge-Weber syndrome. However, knowing the evolution pattern of hemangioma avoids confusion with capillary malformation.

Ultrasound and MRI are the two primary imaging techniques employed in the assessment of vascular anomalies. Ultrasound is widely available, easy to use, non-invasive, has low cost, and usually doesn’t require sedation. However, Doppler evaluation may not be possible in all cases due to motion artifact. MRI has the advantage of superior soft tissue resolution enabling the assessment of deep extension of the lesion, and the involvement of nearby structures in absence of ionizing radiation. Different MRI protocols for imaging of vascular anomalies have been adopted. A multidisciplinary center recommended conventional T1-weighted spin-echo, fat saturated T2-Weighted fast spin-echo and flow-weighted sequences with no need for contrast administration [9]. Other researchers highlighted the role of contrast enhancement in the differentiation of venous from lymphatic malformation [10]. Thanks to its high temporal resolution, time resolved MR angiography has been recently added to increase the diagnostic efficacy of the study [11].
Findings and procedure details

**Procedure details**

Patients with extracranial vascular anomalies of the head and neck who underwent MRI at our institution (between January 2013 and March 2015) were included in the study. Patients had been referred for MRI examination for different reasons: equivocal diagnosis due to uncertain history or poor response to therapy, or to define the deep extension of the lesion. Two children with segmental facial vascular anomalies were referred searching for associated intracranial abnormalities.

Children under the age of six years had been sedated by oral chloral hydrate. General anesthesia was scheduled after failure of oral sedation. All studies had been performed with 1.5-T magnet using 16 channel phased-array neurovascular coil. A 15 channel spine coil had been used additionally in one child who had vascular tumor extending to the upper part of the chest wall. Axial T1-weighted spin-echo (TR/TE/flip angle: 645/15 ms/90°); axial T2-weighted fast spin-echo (TR/TE/flip angle: 5,297/110 ms/90°); axial, sagittal and coronal fat saturated T2-weighted fast spin-echo had been acquired in all patients. A non contrast angiography M2DI (multiple 2 dimensional inflow) had been performed in axial plain (TR/TE/flip angle: 12/ 2.4 ms /60°) followed by maximum intensity projection (MIP) coronal reconstruction. A saturation slap had been selected to suppress the venous flow signal. Fat saturated T1-weighted spin-echo was acquired in multiple planes following the injection of 0.1mmol/Kg gadopentate-dimeglumine. Dynamic post contrast MRI techniques had been available in 6 children. 4D TRAK (4D Time-Resolved Angiography using Keyhole) was available in two children and dynamic THRIVE (T1 High Resolution Isotropic Volume Examination) was available in four. 4D TRAK sequence is a 3D TFE resolution sequence containing 24 dynamics (TR/TE/flip angle: 3.1/ 1.2 ms/ 35°). Dynamics had been acquired with a temporal resolution of 1 sec for each phase. Dynamic THRIVE sequence had included six dynamics (TR/TE/flip angle: 3.9/1.8 ms/ 10°). The contrast had been injected in a dose of 0.2 mmol/kg followed by same dose saline flush. Imaging post-processing had been performed to acquire time-intensity curves. Post contrast fat saturated T1-weighted spin-echo images were acquired at the end of dynamic phases. All fat saturated sequences were acquired using SPAIR (Spectral Adiabatic by Inversion Recovery).

**Image analysis**

The vascular anomalies had been diagnosed as vascular tumor (hemangioma) or vascular malformation (fig 1) Fig. 1 on page 8 following the previously described typical MRI features [12,13], and according to the classification proposed by Mulliken and
Glowacki in 1982 [10] and its recent update of international society of study of vascular anomalies (ISSVA) in 2014 [14]. All the detected lesions had been analyzed according to their signal intensity on T1-weighted spin-echo and T2-weighted fast spin-echo, presence of signal voids, fluid levels and the pattern of contrast enhancement. The lesion extension was assessed in fat saturated T2-weighted fast spin-echo and described as focal or diffuse.

**Imaging findings**

Twenty four patients were enrolled in this study. Their mean age was 3 years (age range between 2 weeks to 20 years). Different types of vascular anomalies were encountered. There were nine children with vascular tumors (seven patients with infantile hemangioma, one patient with congenital hemangioma, and one patient with kaposiform hemangioendothelioma); and fifteen with vascular malformations (one patient with arteriovenous malformation, five with venous malformation, five with macrocystic lymphatic malformation, two with combined malformations, one with microcystic lymphatic malformation, and one with capillary malformation).

Following the previously described typical MRI features, MRI confirmed the clinical diagnosis in 11 patients with proper determination of lesion extension or associated intracranial anomalies. Based on the typical MRI features, diagnosis was established in the remaining patients (13 patients) obviating the need for histopathological verification except in one patient whose MRI features were suggestive of an aggressive lesion necessitating biopsy.

Infantile hemangioma was diagnosed in seven patients. The lesions appeared as well defined localized mass (4 children) (fig.2a,b,c) Fig. 2 on page 8. Diffuse lesion affecting both parotid glands, deep structures of the neck and the laryngeal air way in a beard distribution was seen in one patient (fig 3) Fig. 3 on page 10. Diffuse enlargement of the parotid gland was seen in another patient (fig. 4a,b,c) Fig. 4 on page 10. Segmental facial infantile hemangioma appeared as cutaneous thickening in another patient. Infantile hemangioma exhibited the previously reported typical MRI features in the form of hypointense signal on T1-weighted spin-echo, hyperintense signal on fat-saturated T2-weighted fast spin-echo, and associated with marked homogenous contrast enhancement (fig. 2a,b,c) Fig. 2 on page 8. Intra-lesional flow voids were seen in spin echo sequences in all patients (100%), and appeared as flow related enhancement in M2DI (Fig 4 b,c) Fig. 4 on page 10. Post contrast dynamic THRIVE was performed in one child and revealed arterial phase enhancement (fig 3) Fig. 3 on page 10.

There was one case in this series with congenital hemangioma, who presented in the neonatal period with a well defined mass in the right cheek region. MR images
demonstrated similar features as infantile hemangioma apart from being of more heterogeneous signal.

One girl presented with an infiltrative mass in the neck and upper chest that was pathologically proven Kaposiform hemangioendothelioma. It showed early heterogeneous arterial enhancement with a large feeding vessel (fig 5) Fig. 5 on page 11.

One patient had arteriovenous malformation. It demonstrated tubular high flow vessels without soft tissue mass (fig 4 d,e,f) Fig. 4 on page 10.

Five children had venous malformation. It appeared as well defined lobulated mass lesion in three patients and infiltrative lesion in two. These lesions demonstrated hypointense signal on T1-weighted spin-echo, hyperintense signal on fat saturated T2-weighted fast spin-echo with post contrast enhancement (fig 2 d,e,f) Fig. 2 on page 8. The pattern of contrast enhancement was patchy in two patients (fig 2f) Fig. 2 on page 8 and homogenous in three (fig 6d, 7c) Fig. 6 on page 12 Fig. 7 on page 13. Fluid levels (fig 2e, 8c) Fig. 2 on page 8 Fig. 8 on page 14 were seen in two patients (40%). Signal voids of fibrous striations, phleboliths (fig 8b) Fig. 8 on page 14 or dilated veins (fig 2e, 6) Fig. 2 on page 8 Fig. 6 on page 12 were seen in three patients (60%). Dynamic post contrast MRI was acquired in three patients (4D TRAK in one patient, and dynamic THRIVE in two) and showed enhancement in the venous phase (fig 7, 9) Fig. 7 on page 13, Fig. 9 on page 15 with no prominent arterial feeders.

Macrocystic lymphatic malformation was seen in five patients. Typically, it appeared as well defined multiloculated cystic mass exhibiting hypointense signal on T1-weighted spin-echo, and hyperintense signal on T2-weighted fast spin-echo (fig. 2 g,h,i) Fig. 2 on page 8. However, some of the locules exhibited the reverse (hyperintense signal on T1-weighted spin-echo and hypointense signal on T2-weighted fast spin-echo) likely due to intracystic hemorrhage and/or high protein content (fig 10) Fig. 10 on page 16. Characteristically, macrocystic lymphatic malformation showed marginal contrast enhancement of the cystic spaces (fig 2 g,h,i) Fig. 2 on page 8. Intra-lesional fluid levels were seen in three children (fig 8.d,e,f) Fig. 8 on page 14.

Microcystic lymphatic malformation was seen in one patient and appeared as diffuse sheets of abnormal signal with no appreciable contrast enhancement (fig 11 a,b) Fig. 11 on page 17. Intralesional flow voids were not detected. No flow related enhancement was seen within the lesion in M2DI. Combined venous and microcystic lymphatic malformation was seen in one patient. The venous component appeared as an enhancing soft tissue mass with a draining vein (fig 11 c,d) Fig. 11 on page 17. Combined microcystic and macrocystic lymphatic malformation was seen in one child (fig 12) Fig. 12 on page 18.
Intracranial abnormalities were detected in two children with facial vascular anomaly (fig 13) Fig. 13 on page 18. The first was PHACE syndrome associating an infantile hemangioma. The second was Sturge-Weber syndrome associating a capillary malformation.
Fig. 1: Algorithm for differentiating vascular anomalies using MRI as a single investigation

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Fig. 2: MRI demonstrates the typical features of common vascular anomalies according to their signal characteristics and pattern of enhancement. a,b,c Axial MRI of 7-month-old girl with infantile hemangioma a T1-WI the lesion exhibits hypointense signal (*) with multiple flow voids (arrows). b) axial T2-weighted fast spin-echo with fat-sat showing hyperintense signal of the lesion. c) post contrast T1-weighted spin-echo with fat suppression shows homogenous contrast enhancement (*). d,e,f) Axial MRI of 17-year-old boy with venous malformation. d) T1-WI showing hypointense lesion in the right mandibular region with fluid level (arrow). e) Axial T2 weighted fast spin-echo with fat suppression demonstrates lobulated hyperintense lesion with fluid level (black arrow). A marginal vein with flow void is seen (white arrow) which can be mistaken for signal void of high flow vessel. f) post contrast T1-weighted spin-echo with fat suppression showing patchy enhancement of the lesion and its draining vein (arrow). g,h,i Coronal
MRI of 2-month-old boy with macrocystic lymphatic malformation. g T1-WI showing hypointense lesion in the neck (*). h T2-weighted fast spin-echo with fat suppression showing hyperintensity of the lesion (arrow). i Post contrast T1- weighted spin-echo showing marginal enhancement of the cystic spaces (arrows).

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Fig. 3: Coronal MRI of a 3-month-old girl with infantile hemangioma at the beard distribution. a) T2-wighted fast spin-echo with fat suppression showing hyperintense lesion involving both parotid glands (*) with large feeding vessels (arrow). b) Post contrast T1 weighted spin-echo with fat suppression reveals deep laryngeal extension with narrowing of the air way (arrow). c) Colour map of the THRIVE sequence during arterial phase confirms the high flow nature of the lesion. d) Time-Intensity curve of post contrast dynamic THRIVE shows early and persistent enhancement.

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**Fig. 4:** MRI of two patients with high flow lesions a,b, c 9-month-old girl with infantile hemangioma involving the parotid. a axial T2-weighted with fat suppression showing hyperintense signal (*) with multiple signal flow voids (arrows). b axial source image of M2DI showing hyperintense signal of the flow related enhancement. c coronal gray scale inverted MIP reconstruction showing the high flow feeding vessels (arrows). d,e,f 20-year-old patient with arteriovenous malformation of the right cheek d axial T2-WI showing multiple serpigenous flow voids involving the right cheek (arrows) e axial source image of M2DI showing hyperintense signal of the flow related enhancement. f Axial gray scale inverted MIP reconstruction showing the high flow vessels (arrows).

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Fig. 5: MRI of 2-year-old girl with pathologically proven Kaposiform hemangioendothelioma dated since birth. a) coronal T1-WI shows infiltrative hypointense mass. b) Coronal T1 post contrast shows heterogeneous enhancing mass. c) Coronal gray scale inverted MIP reformat during the arterial phase of 4-D TRAK sequence showing a large mass involving the neck and upper chest wall (arrows) with a large feeding artery (arrowhead). d). Time-Intensity curve of post contrast dynamic study shows marked early enhancement.

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Fig. 6: MRI of a 5-month-old girl with venous malformation dated since birth. a,b axial T1-weighted spin-echo shows hypointense lesion (*) with intraliesional tubular signal void (short arrow) which can be mistaken for high flow arterial feeder; however, by tracing this structure it is seen draining into the left innominate vein (arrowhead), in addition the left common carotid artery (long arrow) is not enlarged excluding the possibility of being a high flow lesion. c coronal T2-weighted fast spin-echo shows hyperintense lesion (*). d Coronal post contrast T1-weighted spin-echo with fat suppression shows homogenous enhancement with intraliesional fibrous septations.

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Fig. 7: MRI of 6-month-old girl with a lump in the right cheek proved to be venous malformation. She was initially diagnosed as infantile hemangioma. She was referred to characterize the lesion as the lesion was stationary in size and not responding to propranolol therapy. a axial T1WI shows hypointense lesion. b axial T2-WI with fat suppression shows hyperintense signal of the lesion. Intraloesional hypointense bands can be mistaken for high flow vessels (arrows). c post contrast T1-weighted spin-echo with fat suppression shows homogenous enhancement. d gray scale inverted sagittal MIP 4D- TRAK reveals no enhancement in the arterial phase with lesion enhancement during the venous phase of the study, confirming its slow flow nature. e) colour map during the venous phase nicely demonstrates the lesion (arrow).

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Fig. 8: MRI of venous malformation and lymphatic malformation. a,b,c MRI of 6-month-old girl with intramuscular venous malformation. a Axial T1-weighted spin-echo shows hypointense lesion within the masseter muscle (*) characterized by presence of phleboliths (arrow). b axial T2-weighted fast spin-echo shows hyperintense lesion (*) with phleboliths (arrow). c axial post contrast T-weighted spin-echo with fat suppression shows intralesional enhancement with a fluid level which indicates stasis of the blood within the venous channels. d,e,f MRI of 4-year old boy with macrocystic lymphatic malformation dated since birth with recent rapid increase in size as a result of intralesional hemorrhage. d axial T1-weighted spin-echo shows a well defined lesion with fluid level due to intralesional hemorrhage. e sagittal T2-weighted fast spin-echo shows multiloculated lesion with multiple fluid levels (arrows). f axial post contrast T-weighted spin-echo with fat suppression shows marginal enhancement of the lesion (arrows).

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**Fig. 9:** MRI (dynamic THRIVE sequence) of 17-year-old boy with venous malformation. a) arterial phase shows absence of lesion enhancement (*). b,c venous phase shows progressive patchy enhancement of the lesion (arrow). d) axial colour map confirms the low flow nature of the lesion. e) coronal gray scale inverted MIP reformat during the arterial phase showing normal angiography with no evidence of high flow vessels. f) coronal gray scale inverted MIP reformat during the venous phase showing dilated vein draining the lesion (arrow). g) Time-Intensity curve confirms delayed contrast enhancement.

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**Fig. 10:** 3-year-old boy with macrocystic lymphatic malformation. Coronal T1-WI(a) and T2WI (b) shows multiloculated mass with mixed hyper and hypointense signal.

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**Fig. 11:** a,b 11-month-old boy with microcystic lymphatic malformation. a. Coronal T2-WI with fat-sat showing rounded and tubular high signal. b post contrast T1-WI with fat suppression showing absence of contrast enhancement. c,d 15-year-old girl with microcystic lymphatic malformation with small venous malformation. c axial T2-WI with fat-sat showing sheets of hyperintense signal. d Post contrast T1-WI with fat suppression showing enhancement of the venous component with prominent draining intracranial vein (arrow).
**Fig. 12:** 5-month-old girl with combined micro and macrocystic lymphatic malformation. a) coronal T2-WI with fat suppression shows multiple cystic spaces (arrows). b,c) coronal T2-WI with fat suppression and post contrast fat suppression at a more anterior section shows infiltrative area of high signal that does not show contrast enhancement (arrows). d) axial post contrast T1WI with fat-sat shows marginal enhancement of the cystic spaces of macrocystic lymphatic malformation (black arrows) with no enhancement of the microcystic lymphatic malformation (white arrows). e) axial colour map of dynamic THRIVE during the arterial phase shows confirms the low flow nature of the lesion.

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**Fig. 13:** Syndromes associating vascular anomalies. a,b 2-year-old boy with segmental facial infantile hemangioma (not shown) with PHACE syndrome. a Axial T2-WI showing right cerebellar hypoplasia (*) as a posterior fossa malformation. b axial T2-WI with Fat-Sat reveals non visualization of the left internal carotid artery (circle). c,d 5-year-old boy with facial capillary malformation and Sturge-Weber syndrome. c Axial T2-WI shows left cerebral hemiatrophy (*), dilated deep veins (arrows). d T2* sequence shows gyral low signal intensity consistent with gyral calcifications (arrows).

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Conclusion

Apart from arteriovenous malformations that have multiple vessels with no associated considerable soft tissue component [9,10], most vascular tumors and malformations demonstrate soft tissue masses with similar hypointense signal on T1WI, and hyperintense signal on T2WI. Fluid levels are characteristic features of macrocystic lymphatic malformation (table 1) Table 1 on page 22. However, presence of fluid levels can be a feature of venous malformation as well. The pattern of contrast enhancement helps in differentiation. Macrocystic lymphatic malformation has characteristic marginal enhancement of the cystic spaces, while venous malformation has typically a patchy pattern of enhancement [12]. Minimal if any enhancement is seen in cases of microcystic lymphatic malformation (table 2) Table 2 on page 22.

The differentiation between Hemangioma and venous malformation can be challenging both clinically and radiologically. Presence of high flow vessels is the main determinant of hemangioma. High flow vessels can be depicted as flow voids on conventional MR sequences. However, the presence of hypointense fibrous striation or dilated veins can be falsely interpreted as flow voids of high flow vascular tumors leading to misdiagnosis as hemangioma. Moreover, venous malformation can demonstrate complete enhancement similar to hemangioma, and phleboliths can be found occasionally in hemangioma [11]. In challenging cases, flow-weighted MR angiography will not demonstrate the serpiginous high flow vessels in venous malformation due to the lack of hypertrophied feeding arteries. In addition, 4D TRAK or dynamic THRIVE with time-intensity curves can be problem-solving techniques enabling the differentiation between hemangioma and venous malformation [11]. Hemangioma shows early arterial homogenous enhancement with multiple feeding vessels, while venous malformation remains unenhanced in the arterial phase and only shows enhancement during the venous phase (table 2) Table 2 on page 22.

Contrast enhanced sequences can be challenging in children because of difficulties in obtaining IV access; in addition, contrast injection can be prohibited in certain situations especially in neonates with immature kidneys or children with impaired renal function when the benefit of contrast injection doesn't outweigh the possible risk of nephrogenic systemic fibrosis. Also, the use of only non-contrast sequences leads to decrease in the timing of examination especially in children under sedation, and substantially reduces the cost of the examination. For these issues, non contrast MR angiography is considered a suitable alternative for patients who are not tolerating gadolinium contrast [15], but it fails to provide the hemodynamic characteristics of vascular anomalies that are required for lesion categorization. In our series we have found contrast injection and dynamic sequences very helpful to increase the diagnostic confidence, differentiate venous and lymphatic malformation, and increase the conspicuity of venous component in cases with combined veno-lymphatic malformations.
Vascular anomalies in the head and neck are mostly diagnosed on clinical basis; however, when the history is uncertain or the diagnosis is equivocal, a well tailored MR examination can be a single valuable diagnostic tool providing structural and functional information. Knowing the typical imaging features and potential pitfalls is crucial to avoid the misinterpretation and faulty diagnosis.
**Table 1:** Characteristic features of vascular tumors and malformations

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**Table 2: Commonly encountered imaging pitfalls in the diagnosis of vascular anomalies by MRI**

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