

MRI evaluation of soft tissue vascular malformations

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Abstract

Background: Soft tissue vascular malformations are not uncommonly encountered in clinical practice and are often mistaken for other pathologies. Vascular anomalies are divided into vascular tumors and malformations. Vascular malformations progressively increase in size with increasing age and are classified into low-flow (venous, lymphatic, veno-lymphatic, capillary and capillary venous) and high-flow malformations (arteriovenous malformations (AVM) and arteriovenous fistula (AVF)) depending on the presence or absence of arterial flow. **Aim:** Aim of this study is to evaluate the Magnetic resonance imaging (MRI) features of the soft tissue vascular malformations and to classify the lesions depending on flow pattern. **Materials and Methods:** A total of 52 patients of soft tissue vascular malformations were included in this study over a period of 3 years. All patients underwent detailed clinical examination and dynamic post contrast MRI evaluation. Doppler and ultrasound were done as an adjunct. **Results:** There were 31 females and 21 male patients with age ranging from 9 to 34 years. In total 33 venous, 11 lymphatic, 8 veno-lymphatic malformations were seen. No case of high-flow malformation was seen. Majority of the lesions involved the head and neck region and the extremities. Fat saturated T2WI, STIR, and 3D dynamic post contrast sequences were found to be the most useful MRI sequences. **Conclusion:** MRI is the modality of choice for evaluating the soft tissue vascular malformations. It depicts the extent of the lesion, classifies the lesions into low or high flow and helps in treatment planning.

Key words: Dynamic post contrast; magnetic resonance imaging; soft tissue vascular malformations

Introduction

Soft tissue vascular malformations are a heterogeneous group of lesions which are not uncommonly seen in clinical practice. They can involve any body part and can affect any age and are one of the most common cause of pediatric soft tissue masses.^[1,2] Clinical history and examination are an important part in evaluating these lesions. Imaging plays a crucial role in identifying and classifying the lesions, in determining the extent of the lesion and in helping to decide the treatment protocol.

Plain radiographs may identify phleboliths but generally have a limited role. Ultrasound and Doppler help to classify the lesions and provide information on the character and vascularity. However because of restricted penetration and limited field of view, ultrasound has limited capabilities.^[3] Magnetic resonance imaging (MRI) with its excellent soft tissue resolution is the modality of choice for evaluating these lesions. It images the entire extent of the lesions, it's relation with the adjacent structures and classifies the

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lesions depending on their enhancement characteristics on gadolinium enhanced MRI. It also helps in the decision for treatment planning.^[1]

The International society for study of vascular anomalies has categorized vascular anomalies into vascular tumors and vascular malformations. The vascular malformations have been further subcategorized as being low-flow or high-flow malformations.^[4]

The aim of this study is to evaluate the MR imaging features of soft tissue vascular malformations on dynamic contrast enhanced MRI and classify the lesions on the flow pattern into high and low flow malformations.

Materials and Methods

A total of 52 patients with soft tissue vascular malformations who reported to our hospital during a period of 3 years from 2015 to 2018 were included in the study. The patients underwent detailed clinical examination prior to the imaging.

All patients underwent MRI on a 1.5 tesla GE MRI scanner. The sequences done included T1WI, T2WI, fat saturated T2WI, STIR, and gradient recalled echo (GRE) sequences in 3 planes (axial, coronal, and sagittal). Precontrast fat saturated T1W and 3D postcontrast dynamic imaging was done in all cases [Table 1]. The contrast used was Gadoterate Meglumine (0.1 mmol/kg). Specialized coils (head, body, extremity, etc.) were used depending on the part evaluated.

Majority of the patients underwent ultrasound examination prior to the MRI. Few patients with extremity swellings had undergone plain radiography prior to the MR imaging. The criteria on MRI for diagnosing low-flow malformations used, were the lack of flow voids on spin echo (SE) and fast spin echo (FSE) sequences. High-flow malformations have vessels which show flow voids on SE sequences and hyperintensity on GRE sequences. 3D postcontrast dynamic imaging was evaluated to detect the lack of early arterial flow and venous shunting in the arterial phase of imaging in the low flow vascular malformations. GRE sequences and post contrast scans were evaluated to detect the presence of phleboliths, septa or intravascular thrombus which can

mimic signal voids on SE sequences. Noncontrast CT was done in a few patients to confirm/clarify the presence of calcification/phleboliths if the same was unclear on MRI. Doppler examination was done as an adjunct in all cases for assessing the vascularity and flow pattern.

Patients with contraindication to MR evaluation were excluded from the study. Young and claustrophobic patients who were unable to undergo MRI were imaged under sedation.

Results

There were 31 females and 21 male patients ranging from 9 to 34 years. Patients presented with symptoms of progressively increasing asymptomatic swellings to local discomfort or pain.

There were 52 cases of low flow vascular malformations. No case of high-flow malformations was imaged in this study. The low-flow malformations included 33 cases of venous malformations, 11 cases of lymphatic malformations and eight cases of veno-lymphatic malformations [Table 2].

Venous malformations were the most common malformation seen in this study. Of the 33 cases, 21 cases were seen involving the extremities and 12 cases seen in the head and neck region. The lesions appeared hypointense on T1WI and hyperintense on T2WI, STIR, and fat saturated T2W sequences. Phleboliths were seen in 17 cases appearing as hypointense foci on T1WI, T2WI, GRE, and postcontrast images [Figures 1-3]. On dynamic postcontrast images majority of these lesions showed diffuse contrast enhancement on delayed images [Figures 4 and 5]. No arterial vessels or draining veins were seen in the dynamic scans. In total 17 lesions were fairly well defined and localized, whereas 15 lesions were diffuse and infiltrative on imaging. Doppler examination also showed the absence of any arterial flow. Mass like features or any extensive surrounding edema was absent in all lesions. A few lesions showed fluid-debris levels in the dependent portion.

There were 11 cases of lymphatic malformations. Of which 8 cases were seen in the head and neck region and 3 in the extremities. On imaging seven cases had a

Table 1: MRI parameters of various sequences

Parameter	T1WI FSE	T2WI FSE	STIR	T2*GRE	Pre Contrast fat saturated T1WI	3D post contrast T1WI
TR (msec)	440-980	3480-5800	2900	465	1100	7.9
TE (msec)	9.2-13	87.9-107.7	41.6	13.3	10.3	3.4
T1 (msec)	-	-	150	-	-	24
Section thickness (mm)	4	4	4	4	4	1.6
Spacing (mm)	1	1	1	0.5	1	0
Matrix	384×224	320×224	256×192	288×192	384×224	288×288

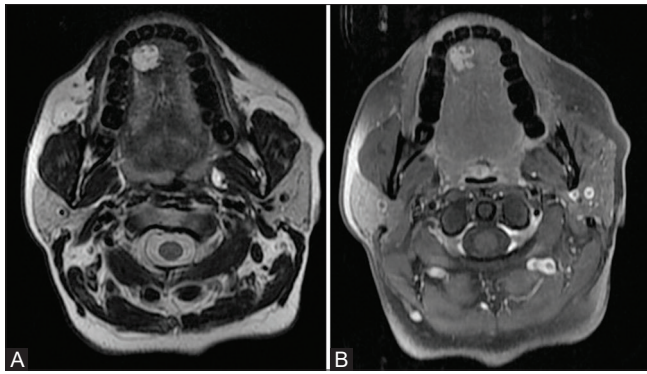


Figure 1 (A and B): Venous malformation tongue: (A) Axial T2W image show an hyperintense lesion in the right genioglossus muscle with phleboliths (B) axial fat saturated post contrast T1W image shows diffuse enhancement in the lesion

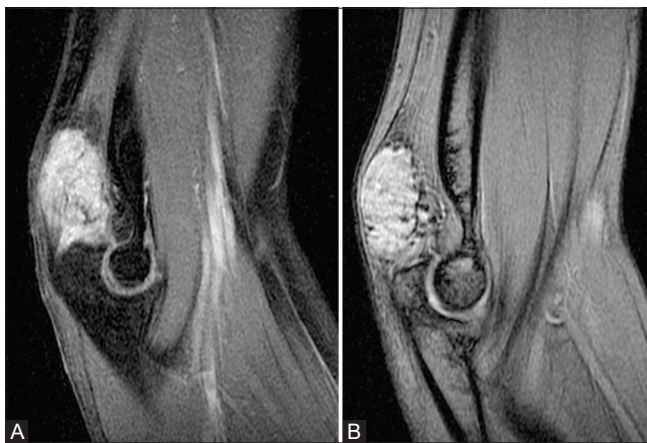


Figure 3 (A and B): Venous malformation elbow: (A) Sagittal STIR and (B) Sagittal gradient echo images show an lobulated hyperintense lesion with multiple hypointense phleboliths within, involving the triceps muscle

Table 2: Type and number of low flow malformations

Type of malformation	Number of cases (n=52)
Venous malformation	33
Lymphatic malformation	11
Veno-lymphatic malformation	08

macrocytic appearance with large cysts of varying sizes appearing hyperintense on T2WI, STIR, and fat saturated T2 sequences, and hypointense on T1WI. In total 5 lesions showed fluid-fluid levels. The lesions were mostly infiltrative and diffuse in nature. On post contrast imaging the lesions did not show any significant enhancement except for some lesions showing thin peripheral/septal enhancement [Figure 6]. Doppler studies confirmed the absence of any significant vascularity.

There were 8 cases of veno-lymphatic malformations seen mainly in the extremities. On post contrast images, these lesions showed patchy enhancement with the absence of any arterial component [Figures 7 and 8].

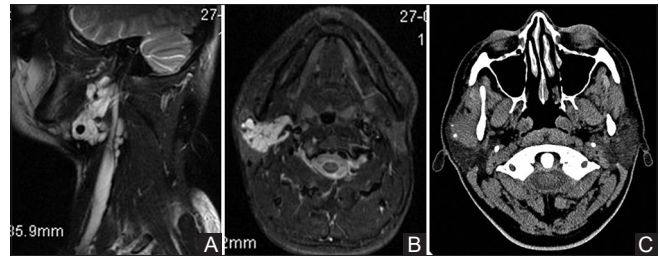


Figure 2 (A-C): Right parotid venous malformation: (A) Sagittal T2W and (B) axial fat saturated T2W images show an multilobulated hyperintense lesion in the right parotid gland with multiple phleboliths within (C) Axial non-contrast CT image confirms phleboliths within the lesion

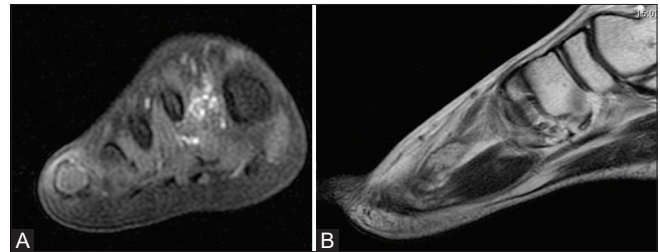


Figure 4 (A and B): Venous malformation right foot: (A) Coronal post contrast T1W fat saturated image show an enhancing lesion in the interosseous muscle (B) The lesion appears hyperintense in the Sagittal T2W image

Sclerotherapy was the main treatment offered to most of the patients. The results of sclerotherapy were varied among the patients. A few lesions were kept under close observation.

Discussion

Vascular anomalies are classified into two categories, vascular tumors, and vascular malformations. Tumors consist mainly of hemangiomas. Vascular malformations are further categorized as low flow (venous, capillary, lymphatic and mixed) and high flow (arteriovenous malformations (AVM) and arteriovenous fistulas (AVF)) depending on the flow dynamics of the lesion. Lesions with an arterial flow component are considered high flow and those without are classified as low flow.^[4-6]

MRI with its superior soft tissue resolution is the modality of choice for evaluating soft tissue vascular malformations. It can show the extent of the lesion, its relation to the surrounding structures and depict excellent anatomical details. The imaging should be done in all three planes, i.e., axial, coronal, and sagittal. Fat saturated T2WI and STIR sequences in all three planes are particularly helpful in depicting the extent of the lesion. In these sequences the malformations appear bright against a dark background. Contrast enhanced 3D T1W sequences are used for evaluating the vascularity of the lesions.^[7-9] However, for evaluating the functional analysis of the lesions dynamic time resolved 3D fast GRE imaging is the sequence of choice. This sequence acquires images with high spatial and temporal resolution and is able to separate the arterial

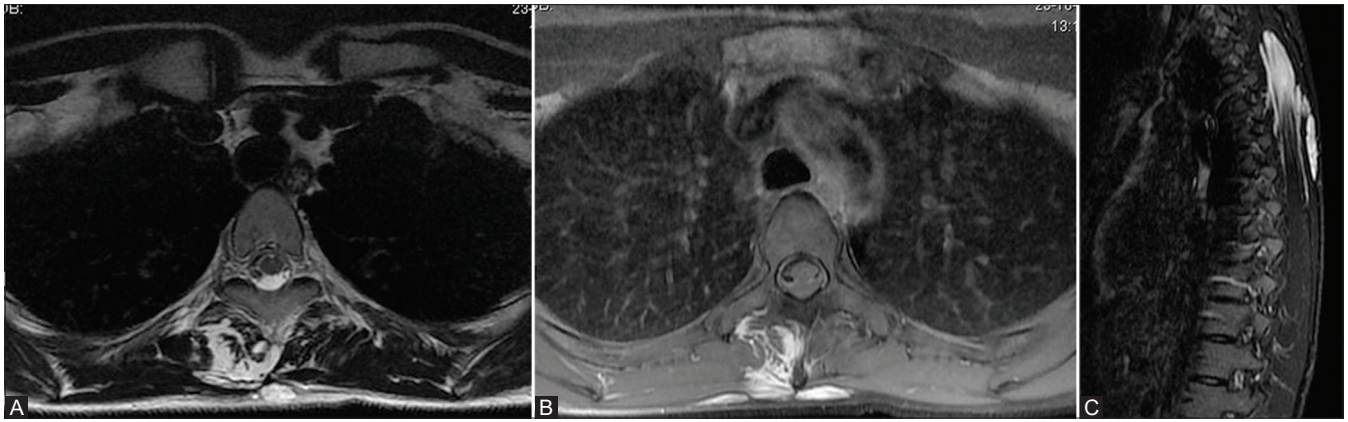


Figure 5 (A-C): Venous malformation paravertebral muscles: (A) Axial T2W image show an hyper intense lesion in right paravertebral muscles. (B) Axial and (C) Sagittal post contrast T1W fat saturated images show diffuse enhancement in the lesion

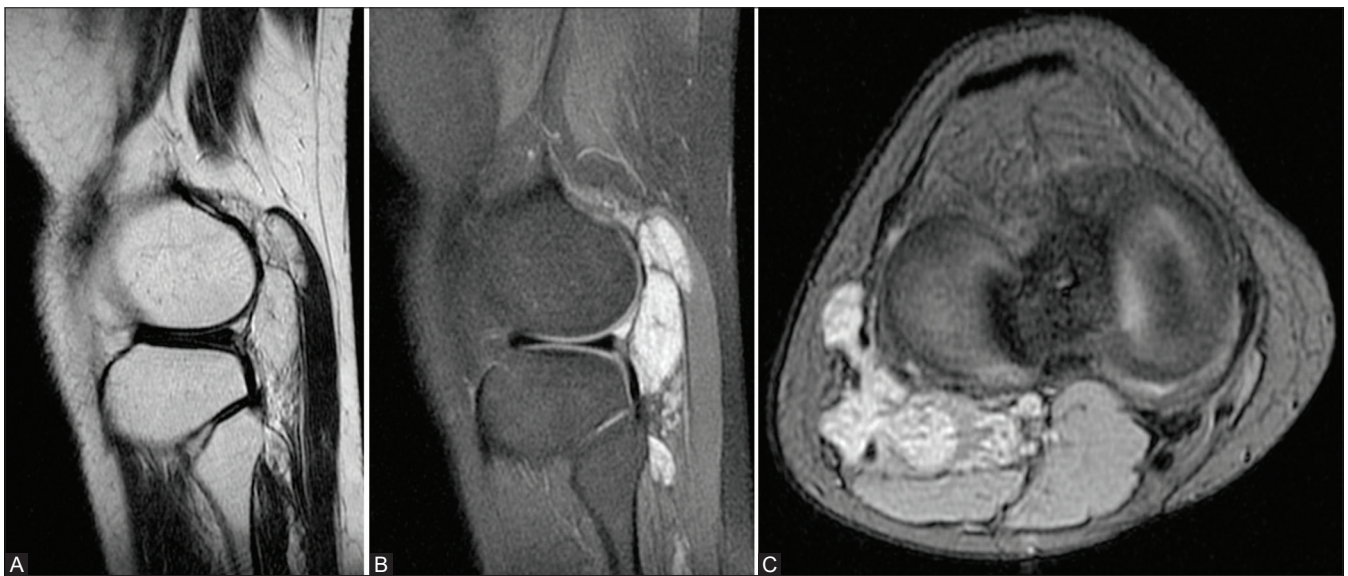


Figure 6 (A-C): Lymphatic malformation right knee: (A) Sagittal T2W (B) Sagittal STIR (C) axial gradient echo images show an multilobulated hyperintense, septated lesion in the lateral head of gastrocnemius

flow from the venous flow and reduce artifacts and hence is able to accurately classify the lesions.^[1,9] T2* weighted GRE sequences can detect calcification or hemorrhage within a lesion and show high flow vessels as having high intensity.^[10]

Vascular tumors are endothelial neoplasms and include infantile hemangiomas (most common lesion), congenital hemangiomas and rare Kaposi form hemangioendotheliomas.^[2,11] Infantile hemangiomas are benign tumors with a female preponderance, characterized by early rapid proliferative growth and complete regression by 7 to 10 years. Most lesions require no treatment.^[1] Most commonly occurring in the face and neck region their diagnosis is often clinical and MR imaging is needed to assess the deep lesions/guide therapy or in follow-up.^[12] In the proliferating phase, the lesions appear well defined and show high signal on T2WI and early uniform intense

enhancement. Involuting lesions show increasing fatty replacement and less enhancement. The presence of perilesional edema should alert for other more ominous tumors, such as rhabdomyosarcoma, fibrosarcoma, hemangiopericytoma, soft tissue sarcomas, etc., which must be ruled out. Any atypical imaging findings should be suspect and must be evaluated with a biopsy.^[13,14]

Vascular malformations are congenital lesions present at birth. These lesions grow with age and are classified as low flow and high flow depending on the flow characteristics and the presence/absence of arterial flow.

Venous malformations consist of dysplastic vascular channels and present with deformity, pain and impaired mobility. Often located in the head and neck region, extremities and the trunk, and they present as swellings which increase on Valsalva and are compressible if

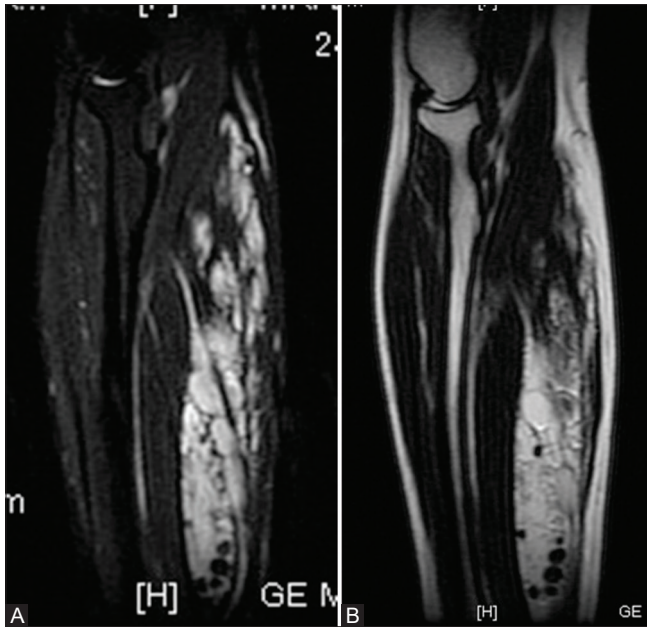


Figure 7 (A and B): Veno-lymphatic malformation of forearm: (A) Sagittal STIR and (B) Sagittal T2W images show phleboliths and fluid-fluid levels within the lesion involving the brachioradialis muscle

superficial. They can be seen alone or combined with lymphatic malformations in syndromes like Proteus, Maffucci, and blue rubber bleb nevus syndrome. T2WI and STIR sequences are very helpful to map the extent of the lesions, which can involve multiple tissue planes and invade adjacent tissues such as muscle, tendon and bone. The lesions can have fluid-fluid levels due to hemorrhage. The presence of phleboliths, appearing as signal voids on all sequences, provide the best clue for their diagnosis. On postcontrast images, the lesions show gradual filling in and delayed scans are helpful to detect diffuse enhancement. Typically, there is the absence of AV shunting, early arterial enhancement and enlarged feeding vessels.^[11,15] Venous malformations were the most malformation (33 cases) seen in this study and most lesions showed characteristic imaging features.

Lymphatic malformations consist of chyle filled cysts lined by endothelium and are the second most common malformations. Commonly associated with other malformations, they are classified as microcystic and macrocystic. They often present in early life as smooth non compressible soft tissue masses, usually in the posterior neck and axilla. They are infiltrative lesions which appear hyperintense on T2WI and STIR and commonly have fluid-fluid levels. On post contrast images they usually do not enhance or may show only peripheral or septal enhancement. If associated with venous malformations the lesions may show diffuse enhancement.^[13,14] In total 11 lymphatic and 8 veno-lymphatic malformations were seen in this study.

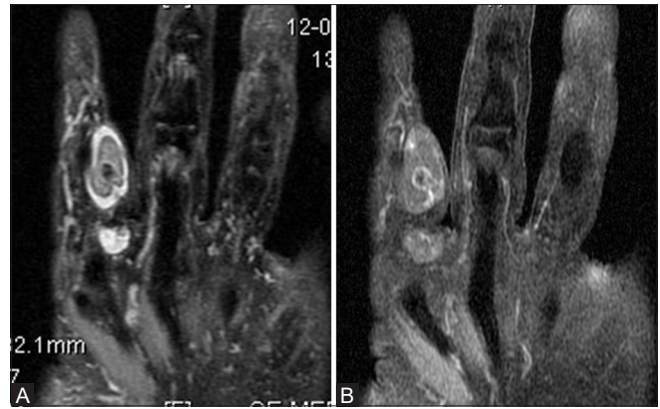


Figure 8 (A and B): Veno-lymphatic malformation right index finger: (A) Coronal STIR and (B) post contrast Coronal T1W fat saturated images show a large phlebolith and fluid-fluid level within the lesion

Capillary malformations are usually superficial and involve the skin and mucosal surface and are diagnosed clinically. They occur in syndromes such as Klippel Trenaunay, Parkes Weber, and Sturge Weber. Capillary venous malformations on imaging are similar to venous malformations except that they show early homogenous enhancement compared with delayed enhancement in the latter.^[1,16]

High-flow vascular malformations include infantile hemangiomas, AVM, and AVF. On imaging AVMs consist of enlarged arteries and draining veins with a nidus which are well demonstrated on time resolved dynamic 3D MR angiography. Congenital AVF consist of single vascular channel between an artery and vein and are seen usually in the head and neck regions. In this study no cases of high-flow malformations were seen.

The clinical implications and importance of classifying the lesions as low or high flow are in the fact that the treatment options vary for both the type of lesions. Percutaneous sclerotherapy with agents like ethanol is the treatment of choice for low flow malformations. These lesions undergo fibrosis and progressive regression with time. Laser therapy and at times surgery can also be used in specific clinical settings. The aim of therapy in high flow vascular malformations is the eradication of the nidus which is achieved by trans-arterial embolization. Surgical excision following pre op embolization can also be attempted in some lesions.^[17,18]

Conclusion

Soft tissue vascular malformations are not infrequently encountered and are often mistaken for other lesions. Dynamic contrast enhanced MRI is the modality of choice for classifying the lesions, mapping their entire extent, helping in deciding the management and in the follow up of the lesions. Special emphasis needs to be laid on the inclusion of the correct sequences during the MR

examination. Heavily T2 weighted and STIR sequences are very helpful to detect the extent of the lesions. Ultrasound and Doppler are helpful in characterizing the lesions and assessing the vascularity and are useful adjuncts to MRI. The presence of mass like features and extensive surrounding edema are suspicious features and should be evaluated with biopsy.

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Nil

Conflicts of interest

There is no conflict of interest.

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