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ROLE OF 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 the low-flow (venous, lymphatic, venous-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. Vascular Malformations belong to the spectrum of orphan diseases and can involve all segments of the vascular tree: arteries, capillaries, and veins, and similarly the lymphatic vasculature. The classification according to the International Society for the Study of Vascular Anomalies (ISSVA) is of major importance to guide proper treatment. Imaging plays a crucial role to classify vascular malformations according to their dominant vessel type, anatomical extension, and flow pattern.

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 the flow pattern.

MATERIAL AND METHOD:
The study was an Observational study has been conducted in the Department of Radiology. A total of 50 patients with soft tissue vascular malformations who reported to our hospital. The patients underwent detailed clinical examinations 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). Pre-contrast fat-saturated T1W and 3D postcontrast dynamic imaging were done in all cases. The contrast used was Gadoterate Meglumine (0.1 mmol/kg). Specialized coils (head, body, extremity, etc.) were used depending on the part evaluated. Patients with contraindications to MR evaluation were excluded from the study.

RESULTS:
Venous malformations were the most common malformation seen in this study. Of the 30 cases, 18 cases were seen involving the extremities and 12 cases were 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 13 cases appearing as hypointense foci on T1WI, T2WI, GRE, and post-contrast images. On dynamic postcontrast images, the majority of these lesions showed diffuse contrast enhancement on delayed images. No case of high-flow malformations was imaged in this study. The low-flow malformations included 32 cases of venous malformations, 10 cases of lymphatic malformations, and eight cases of venous-lymphatic malformations.

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, and helping in deciding the management and in 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.

KEYWORDS: Dynamic Post Contrast, Magnetic Resonance Imaging and Vascular Malformation

INTRODUCTION

Vascular malformations (VM) are vascular spaces lined by flat epithelium with an estimated prevalence of 4.5% in the population.[1] It is usually present at birth, with enlargement proportional to the child's growth, but sudden expansion may be seen in infection, hormonal changes, or trauma.[2] They are the most common childhood soft tissue masses.[3] VM occurs due to errors in morphogenesis however they exhibit normal cell turnover, unlike vascular tumors.[4,5] These lesions can be diffuse or focal, simple or combined based on the subtype of vessels involved.[6] Appropriate distinction between different VM lead to improved management of lesions, therefore VM

can be classified by the type of vessel component (capillary, venous, lymphatic, arterial, and hybrid subtype) and according to blood flow dynamics (high and slow flow lesions).[7] Vascular anomalies (VAs) comprise a wide spectrum of vascular lesions, which can involve any part of the body, including vascular tumors with altered cellular proliferation, and vascular malformations with underlying mesenchymal and angiogenetic dysplasia.[8,9] Whereas infantile hemangiomas as the most frequent vascular tumor regularly regress with the patient's age, vascular malformations never regress on their own, but frequently increase in size and grow concomitantly or even overproportioned with the child. The most accepted classification of VAs, grounded in biological, histopathological, hemodynamic, and clinical findings, is provided by the International Society for the Study of Vascular Anomalies (ISSVA).[10]

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.[11] 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, and their relation with the adjacent structures and classifies the lesions depending on their enhancement characteristics on gadolinium-enhanced MRI. It also helps in the decision for treatment planning.[12]

Doppler ultrasonography (US) has been the first diagnostic modality utilized in the management of patients with VM, due to low-cost, non-ionizing technology and the ability to provide lesions' flow characteristics[13], however magnetic resonance imaging (MRI) has proven advantageous to define the extent of the lesions and guide appropriate treatment.[14] As prenatal MRI distinguishes the location, morphological components, and flow components of a lesion, it provides information allowing for distinct in utero differential diagnosis of tumors and vascular malformations.[15,16] Further MR angiographic techniques without intravenous contrast, such as time-of-flight (TOF) or phase contrast (PC) sequences, which do not expose the patient to contrast agents, are less used in the evaluation of VAs owing to their high susceptibility towards different flow velocities and associated flow-related artifacts.[17] Post-contrast MRI is useful to examine slow-flow malformations including the extent of their drainage in the venous system and characterization of mixed veno-lymphatic malformations. Imaging should be able to assess the exact flow dynamics in order to differentiate the various types of vascular malformations (fast-flow vs. slow-flow lesions), as to date there is a high rate of misdiagnosis.[18] The International Society for the 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.[19] 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.

MATERIAL AND METHODS

The study was an Observational study has been conducted in the Department of Radiology. A total of 50 patients with soft tissue vascular malformations who reported to our hospital. The patients underwent detailed clinical examinations 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). Pre-contrast fat-saturated T1W and 3D postcontrast dynamic imaging were done in all cases. The contrast used was Gadoterate Meglumine (0.1 mmol/kg). Specialized coils (head, body, extremity, etc.) were used depending on the part evaluated. Patients with contraindications to MR evaluation were excluded from the study. Young and claustrophobic patients who were unable to undergo MRI were imaged under sedation. Informed written consent was taken from the patients or their guardians willing to participate in the study.

MRI Examination:

The majority of the patients underwent ultrasound examinations 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 that 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 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. Non-contrast 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.

Imaging Technique:

To permit subsequent image subtractions, we collected 3D data sets both before and after rapid infusion of 0.3 mmol of gadobenate dimeglumine (Multi-Hance, Bracco, Milan, Italy), flushed with 20 mL of saline. The contrast material was administered at a rate of 2.0 mL/sec using an automated injection system (Spectris, Medrad, Pittsburgh, PA). After acquiring the early arterial phase data set, we obtained late arterial and early and late venous phase MRIs approximately every 30 sec. Images were reconstructed in the coronal plane. Conventional venography was performed in patients with venous vascular malformations or hemangiomas with and without supra malleolar compression by injecting iobitridol and saline (1:1 ratio) into the dorsal vein of the first toe of the affected limb. Furthermore, the superficial varicose veins of two vascular malformations (one in the calf and one in the hand) were directly punctured. Flow and distribution after an instant injection of nonionic contrast material were observed under fluoroscopy.

Duplex Sonography:

All 50 patients underwent duplex color sonography of the affected extremity. After identifying the relevant vasculature, we assessed the feeding and draining vessels and measured flow using color-flow Doppler sonography. Changes in flow patterns were analyzed during Valsalva's maneuver. All examinations were performed by experienced staff radiologists on an advanced sonography scanner with probes ranging from 4.5 to 7.5 MHz. The value of MR angiograms alone for the assessment of lesion extension and interventional planning was compared with the values of MRI and all other vascular imaging techniques.

STATISTICAL ANALYSIS

Data were analyzed by statistical software SPSS version 20. To eliminate any recognition bias, we masked all patient-related data on the images, which could be viewed as hard copies or as

electronic files on a workstation. MRI data sets were evaluated separately in random order by a panel consisting of two board-certified radiologists who reached their findings by consensus. Both interpreters were unaware of the results of the other examinations. After establishing the technical adequacy of the examination, we assessed MRI data sets for the type of lesion visualized (i.e., venous malformation, arteriovenous malformation, and hemangioma); any abnormalities in the feeding arterial or draining venous systems; and the involvement (if any) of muscle, bone, and joints by the lesion

RESULT: -

There were 30 female and 20 male patients ranging from 10 to 34 years. Patients presented with symptoms of progressively increasing asymptomatic swellings to local discomfort or pain.

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)	437-970	3478-5802	2905	454	1104	6.7
TE (msec)	8.2-12	85.6-104.5	39.4	12.2	11.2	2.3
T1 (msec)	-	-	150	-	-	24
Section thickness (mm)	3	3	3	3	3	1.3
Spacing (mm)	1	1	1	0.5	1	0
Matrix	384×224	320×224	256×192	288×192	384×224	288×288

Venous malformations were the most common malformation seen in this study. Of the 30 cases, 18 cases were seen involving the extremities and 12 cases were 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 13 cases appearing as hypointense foci on T1WI, T2WI, GRE, and post-contrast images. On dynamic postcontrast images, the majority of these lesions showed diffuse contrast enhancement on delayed images

No arterial vessels or draining veins were seen in the dynamic scans. In total 15 lesions were fairly well-defined and localized, whereas 14 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 were absent in all lesions. A few lesions showed fluid-debris levels in the dependent portion.

Table 2: Type and number of low-flow malformations

Type of malformation	Number of cases (n=50)
Venous malformation	32
Lymphatic malformation	10
Vino-Lymphatic malformation	8

There were 50 cases of low-flow vascular malformations. No case of high-flow malformations was imaged in this study. The low-flow malformations included 32 cases of venous malformations, 10 cases of lymphatic malformations, and eight cases of venous-lymphatic malformations.

There were 10 cases of lymphatic malformations. Of these 8 cases were seen in the head and neck region and 2 in the extremities. On imaging, seven cases had a macro cystic 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.

There were 8 cases of venous-lymphatic malformations seen mainly in the extremities. On post-contrast images, these lesions showed patchy enhancement with the absence of any arterial component Sclerotherapy was the main treatment offered to most of the patients. The results of sclerotherapy 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.[20]

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, and 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.[21] 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 flow from the venous flow and reduce artifacts and hence is able to accurately classify the lesions.[22] T2- weighted GRE sequences can detect calcification or hemorrhage within a lesion and show high-flow vessels as having high intensity.[23]

Mulliken and Glowacki 1982[24] with further revisions in 1996 based on pathologic features by the International Society for the Study of Vascular Anomalies (ISSVA), is now widely accepted by various subspecialists. The correlation of this system with clinical history and treatment options makes this approach clinically useful. Awareness of these atypical features helps radiologists not to misdiagnose these lesions. AVMs also

require diagnostic angiography and they present as dilated arteries with early opacification of dilated veins in cases where embolization is performed during the angiography, we see that additional arteries become apparent after original vessels are embolized. [25,26]

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 that increase on Valsalva and are compressible if superficial. They can be seen alone or combined with lymphatic malformations in syndromes like Proteus, Maffuci, 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, provides the best clue for their diagnosis. On post-contrast 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.[27,28] Venous malformations were the most malformation seen in this study and most lesions showed characteristic imaging features.

Commonly associated with other malformations, they are classified as microcystic and macrocytic. They often present in early life as smooth non compressible soft tissue masses, usually in the posterior neck and axilla. They are infiltrative lesions that appear hyperintense on T2WI and STIR and commonly have fluid-fluid levels. On post-contrast images, usually do not enhance or may show only peripheral or septal enhancement. If associated with venous malformations the lesions may show diffuse enhancement.[29,30]

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 consists of the single vascular channel between an artery and vein and is 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 types of lesions.[31] 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.

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, and helping in deciding the management and in 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 is suspicious features and should be evaluated with a biopsy. As misdiagnosis and false classification of vascular malformations are frequent, imaging modalities easy to use and interpret are of utmost importance. In addition, the recently improved understanding of pathogenesis and progression forces visualization of the underlying biological activity and angiogenesis in vivo. Thus, novel techniques for diagnosis, patient/treatment selection, and treatment monitoring are required to improve patient outcomes. The attractiveness of imaging and treating the same molecular target is appealing. A few pioneering imaging studies including PET and MSOT have successfully targeted highly specific aspects of vascular malformations while attesting to the feasibility of such methods.

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