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The role of MRI in diagnostic algorithm of cervicofacial vascular anomalies in children

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Summary

Background: Vascular anomalies are usually diagnosed through their clinical picture and history. The purpose of this study was to assess the role of MR imaging in initial assessment of cervicofacial vascular anomalies in children.

Material/Methods: Twenty pediatric patients with vascular anomalies located in the cervicofacial region underwent MRI examination in our department. Images were evaluated for lesion detectability and its signal characteristics (on T1w, T2w images with fat suppression and contrast enhanced T1w sequences); the extent of the lesions and surrounding tissue involvement were also assessed.

Results: In the studied group MR images revealed all anomalies and provided information of their anatomic extent and invasion of surrounding anatomic structures. Nine hemangiomas and six venous malformations were found among studied patients. Two children had multiloculated lesions corresponding to lymphatic malformations. One examination visualized a lesion consisting mainly of dilated vascular channels with an apparent feeding artery, which was consistent with arteriovenous malformation. Two remaining lesions were mixed malformations. Nine patients had lesions limited to subcutaneous tissue. Two masses infiltrated bone structures. There was muscle involvement found in nine cases.

Conclusions: MR imaging is a well-established method for detection and monitoring of vascular anomalies in children. With ultrasound used mostly for initial diagnosis and additional flow assessment, angiography viewed as an invasive therapeutic method and computed tomography used only in specific situations due to its high irradiation dose, magnetic resonance is the best imaging method used in differential diagnosis and topographical characterization of vascular malformations and tumors of cervicofacial area in pediatric patients. Noninvasively and without irradiation, it enables evaluation of the extent and characteristics of lesions and planning proper therapeutic strategy.

Key words: MRI • hemangioma • vascular malformations • pediatrics

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Background

Vascular anomalies comprise a broad group of lesions diagnosed mainly in a pediatric population and constitute an important clinical problem. They may occur in any location, but are most often present within structures of head and neck [1]. According to Mulliken's and Glowacki's classification [2] based on cytological and clinical picture adopted by the International Society for the Study of Vascular Anomalies (ISSVA), vascular anomalies are divided into vascular tumors and vascular malformations [3], as shown in Table 1.

Hemangiomas are benign vascular tumors that present at childhood, with increased cellular proliferation and hyperplasia, characterized by slow involution. On the other hand, vascular malformations are congenital lesions formed out of dysplastic vascular canals that do not vanish at later age. Depending on the type of flow, we distinguish slow flow

Table 1. Classification of vascular anomalies according to ISSVA.

Vascular anomalies	
Vascular tumors	Vascular malformations
· Hemangiomas (neonatal age, congenital- NICH I RICH) · Hemangioendotheliomas · Dermatological acquired vascular tumors (i.a. pyogenic granuloma)	Slow-flow · Capillary · Lymphatic · Venous High-flow · Arterial malformations · Arteriovenous malformations · Arteriovenous fistulas

malformations, i.e. capillary, venous, lymphatic, mixed, or high flow malformations and arteriovenous fistulas [4].

Commencing treatment with laser therapy, sclerotherapy, embolization or surgical excision as well as prognosis depend on the type of anomaly [3,5]. Due to characteristic appearance of lesions and their temporal evolution, diagnosis is based mainly on clinical features and medical history data [3]. Diagnostic imaging studies are necessary for thorough differential and topographic assessment of lesions, and thus making a decision to commence appropriate treatment [6]. The goal of this work is to determine the role of magnetic resonance imaging (MRI) in the diagnosis of vascular anomalies in children.

Material and Methods

Material

Study group consisted of 20 patients aged 2 months to 10 years with clinically diagnosed vascular anomaly referred to MRI examination at the Department of Radiology of the University Clinical Hospital No 1 in Lodz. Data on patient age, sex, anatomical location of the lesion, performed imaging studies and treatment methods are gathered in Tables 2 and 3.

Methods

MRI examinations were performed with a 1.5T Avanto Siemens scanner (Erlangen, Germany) using a craniocervical coil. Study protocol consisted of the following sequences: T1-weighted (TR 409-765, TE 7.8-11 ms) before and after administration of contrast medium with fat saturation and T2-weighted (TR 4440-6340, TE 96-100 ms) with fat saturation performed in three basic planes. Beside two cases (difficulties with establishing i.v. access) patients received intravenous contrast medium (Magnevist in neonates and children up to 7 years old at a dose of 0.2 ml/kg and Gadovist in children older than 7 years at a dose of 0.1 ml/kg). FOV was determined at 23×23 cm, voxel size from 0.5×0.5×3.0 mm to 0.8×0.8×4.0 mm, flip angle between 90 and 150 degrees.

The following criteria were taken into consideration during evaluation of each study: possible assessment of lesion as positive or negative, demarcation of a lesion – good or poor, signal type – hypo, iso- or hyperintense, signal homogeneity, internal structure, degree and homogeneity of contrast enhancement, extent of lesions and degree of displacement and/or infiltration of neighboring structures.

Table 2. Patients' characteristics.

No.	Age	Sex	Location
1.	7 years	M	Neck
2.	5 years 7 m	M	Face- cheek
3.	5 m	F	Face-cheek, canthus
4.	1 year 5 m	F	Face-preauricular area, cheek
5.	11m	F	Neck
6.	2 m	F	Neck
7.	11 m	F	Face- upper lip
8.	2 years	F	Face-cheek, oral cavity antrum
9.	4 years 8 m	M	Neck, preauricular area
10.	6 m	M	Neck
11.	1 year 2 m	F	Face- cheek
12.	6 m	M	Face- cheek
13.	7 years 9 m	F	Face- mandibular area
14.	11 m	F	Face- cheek
15.	4 years 4 m	M	Face- cheek
16.	11 years 2 m	F	Face- cheek, preauricular area
17.	2 years 7 m	F	Face- cheek, lower lip
18.	10 years	F	Neck
19.	8 years 8 m	F	Neck
20.	4 years 6 m	M	Face- cheek

Results

Table 4 presents characteristics of vascular lesions in MRI. In the analyzed group we found 9 hemangiomas, 6 venous malformations, 2 lymphatic malformations, 1 arteriovenous malformation and 2 mixed type malformations. MRI technique enables detecting all anomalies, evaluating their anatomical extent and degree of infiltration of neighboring structures. These lesions were hypo- or isointense in T1-weighted images and hyperintense in T2-weighted images. Following application of contrast medium we acquired strong contrast enhancement in all cases. Nine of 20 lesions were contained within subcutaneous tissue. Significant displacement of surrounding organs was visible in six cases. Involvement of neighboring structures was visualized in 9 patients, including involvement of masseter muscle in five cases and of other facial muscles in four subjects. However, bone infiltration was observed in two cases and encompassed temporal bone and mandible respectively.

Discussion

Clinical assessment of vascular craniocervical anomalies in children remains crucial for the diagnostic process.

Table 3. Other diagnostic methods and treatment.

No.	Diagnosis	Other examinations	Treatment
1.	Hemangioma	US	-
2.	Lymphatic malformation	US	Surgical removal and bleomycin injection (several)
3.	Congenital hemangioma	Blood cell scintigraphy	Propranolol; laser therapy
4.	Mixed malformation	–	–
5.	Hemangioma	–	–
6.	Lymphatic malformation	US	Antibiotics, referral to surgery
7.	Hemangioma	–	Surgical removal
8.	Venous malformation	US	Sclerotherapy (three times)
9.	Venous malformation	US	Surgical removal, sclerotherapy (several times), laser therapy
10.	Hemangioma	US	No treatment
11.	Hemangioma	–	Steroid injections (Polcortolon 40, Dexaven)
12.	Hemangioma	US	No treatment
13.	Arteriovenous malformation	US, angiography	Embolization with ONYX
14.	Hemangioma	US	No treatment
15.	Venous malformation	–	Surgical removal, sclerotherapy
16.	Venous malformation	Phlebography	Sclerotherapy
17.	Venous malformation	–	Sclerotherapy and surgical removal
18.	Mixed malformation	–	–
19.	Venous malformation	US	Sclerotherapy (two times) and surgical removal
20.	Hemangioma	US	Surgical removal

However, proper radiological assessment in diagnostic imaging studies is necessary to confirm the diagnosis and precisely determine structure of lesions, their size and topographic relationships before planning most effective treatment [3]. Beside MRI technique ultrasonography – including Doppler technique, conventional angiography and computed tomography are also used in the assessment of vascular anomalies [7].

Ultrasound allows for initial assessment of size of lesions, their morphology and, importantly, evaluation of vascular flow [8,9]. It is particularly useful for evaluation of small, superficial lesions [10]. Widespread access, low invasiveness, chance to assess flow and low cost of this study make it the most frequently used diagnostic imaging method in patients with craniocervical vascular anomalies [11]. Being a dynamic study, giving a possibility to apply pressure, it allows for differentiation of hemangiomas, venous and lymphatic malformations and arteriovenous fistulas [8,12]. Ultrasonography is also used as a good tool for follow-up in order to assess their evolution and for initial assessment of possible complications. Limitations of this method are associated with relatively small tissue range related to the depth of penetration by ultrasounds, minor ability to differentiate and dependence on diagnostician's experience [13]. In the analyzed group, 11 of 20 patients were referred for ultrasound examination at the first stage of diagnostic process, although due to the need for further evaluation, diagnostics were broadened to include MRI.

Although another diagnostic method – conventional angiography – cannot assess the full extent of pathology, it enables detection of afferent and efferent vessels [14]. It is currently used in case of arteriovenous malformations, mainly as an effective therapeutic modality, for embolization of high flow vascular lesions [9,15,16]. Among the examined patients, embolization of arteriovenous malformation was performed at three stages in one subject, leading to clinically and radiologicaly apparent improvement.

In case of computed tomography, angiography option can visualize vascular lesions and foci of abnormal vessels together with their supplying vessels [17,18]. CT is excellent at visualizing phlebolites within lesions [19]. However, it is not always possible to clearly differentiate between neighboring tissues and examined lesion [20]. Although in a comparison study Kakimoto et al. did not observe a significant differences in detection of vascular anomalies with CT and MRI [19], the role of CT in a pediatric population remains limited to bone lesions due to the use of ionizing radiation. In such cases, CT facilitates precise determination of degree of bone infiltration [3,17] and enables planning and conducting multidisciplinary treatment involving a pediatrician, vascular surgeon, orthopedic surgeon and a

Table 4. MRI characteristics of vascular anomalies in the studied group.

No.	Diagnosis	Demarcation (well demarcated/ weakly demarcated)	Character of signal in MRI sequences: T1-weighted	Character of signal in MRI sequences: T2-weighted	Signal homogeneity	Signal loss within a lesion	Internal structure of a lesion	Contrast enhancement (++/+/–)	Homogeneity of contrast enhancement	Lesion extent (skin, subcutaneous tissue, infiltration of muscles and bone structures)	Afferent and efferent lesions	Degree of displacement of neighboring tissues
1.	Hemangioma	Well demarcated	Iso	Hyper	Quite homogeneous	+	–	++	Quite homogeneous	Subcutaneous tissue suprascapular	Numerous vessels involved in venous drainage	++
2.	Lymphatic malformation	Weakly demarcated	Iso	Hyper	Heterogeneous	-	Multicystic	++	Quite homogeneous	Subcutaneous tissue half of the face	–	+
3.	Hemangioma	Well demarcated	Iso	Hyper	Quite homogeneous	–	–	++	Quite homogeneous	Subcutaneous tissue of upper palpebra, angle of the eye, infiltrating the orbit	–	+
4.	Mixed malformation	Weakly demarcated	Iso	Hyper	Quite homogeneous	–	Foci of tortuous and dilated vessels within a tumor	++	Quite homogeneous	Skin, subcutaneous tissue of half of the face and neck, masseter muscle	–	++
5.	Hemangioma	Well demarcated	Iso	Hyper	Quite homogeneous	+	–	++	Homogeneous	Subcutaneous tissue	Supplied by right thyreocervical trunk, drainage through occipital vein	+
6.	Lymphatic malformation	Well demarcated	Iso/hypo	Hyper	Heterogeneous	–	Multicystic	+(wall)	Homogeneous within walls	Subcutaneous tissue	–	++
7.	Hemangioma	Well demarcated	Iso	Hyper	Homogeneous	–	–	++	Homogeneous	Entire thickness of upper lip, muscles of the upper lip	–	+
8.	Venous malformation	Weakly demarcated	Iso	Hyper	Heterogeneous	–	Numerous bands of fluid	++	Heterogeneous	Masseter muscle, pterygoid muscle, parotid gland, sublingual salivary gland and base of the tongue	–	++
9.	Venous malformation	Quite well demarcated	Iso	Hyper	Heterogeneous	–	Microlobular	+	Heterogeneous	Parotid gland, subcutaneous tissue, masseter muscle	–	+
10.	Hemangioma	Quite well demarcated	Hypo	Hyper	Heterogeneous	+	Numerous vessels	++	Quite homogeneous	Subcutaneous muscle	–	+

Table 4 continued. MRI characteristics of vascular anomalies in the studied group.

No.	Diagnosis	Demarcation (well demarcated/ weakly demarcated)	Character of signal in MRI sequences T1-weighted	T2-weighted	Signal homogeneity	Signal loss within a lesion	Internal structure of a lesion	Contrast enhancement (++/+/–)	Homogeneity of contrast enhancement	Lesion extent (skin, subcutaneous tissue, infiltration of muscles and bone structures)	Afferent and efferent lesions	Degree of displacement of neighboring tissues
11.	Hemangioma	Quite well demarcated	Hypo	Hyper	Homogeneous	+	Numerous tortuous and dilated vessels within a lesion;	++	Quite homogeneous	Skin, subcutaneous tissue, platysma, muscles of facial expression, cartilaginous part of nares	Drainage through facial vein	+
12.	Hemangioma	Weakly demarcated	Iso	Hyper	Homogeneous	+	Microlobular, numerous vessels within a lesion	++	Homogeneous	Subcutaneous tissue	–	+
13.	Arteriovenous malformation	Quite well demarcated	Iso/hypo	Hyper	Heterogeneous	+	Numerous vessels	+	Heterogeneous	Mandibular body	+	++
14.	Hemangioma	Quite well demarcated	Iso	Hyper	Homogeneous	+	–	+	Homogeneous	Subcutaneous tissue, masseter muscle, parotid gland	+	+
15.	Venous malformation	Well demarcated	Iso	Hyper	Heterogeneous	+	Heterogeneous, spongeous	Na	Na	Subcutaneous tissue	–	+
16.	Venous malformation	Weakly demarcated	Iso	Hyper	Homogeneous	–	–	++	Heterogeneous	Subcutaneous tissue, muscles (masseter, pterygoids), alveolar processes	–	+
17.	Venous malformation	Weakly demarcated	Iso	Hyper	Heterogeneous	–	–	+	Heterogeneous	Entire thickness of lower and upper lip up to the mucosa	–	+
18.	Mixed malformation	Weakly demarcated	Iso	Hyper	Heterogeneous	–	Fluid-filled and solid spaces	+	Heterogeneous	Subcutaneous tissue, pyramid of temporal bone, tongue, mandibular angle	–	++
19.	Venous malformation	Weakly demarcated	Iso	Hyper	Heterogeneous	–	–	+	Heterogeneous	Subcutaneous tissue of lateral part of neck	–	+
20.	Hemangioma	Well demarcated	Iso	Hyper	Homogeneous	–	–	na	na	Subcutaneous tissue	–	+

radiologist. None of the patients from our group had computed tomography performed.

Due to good tissue contrasting, multilevel imaging and absence of ionizing radiation MRI is an excellent method for the diagnosis of vascular anomalies in children. It can

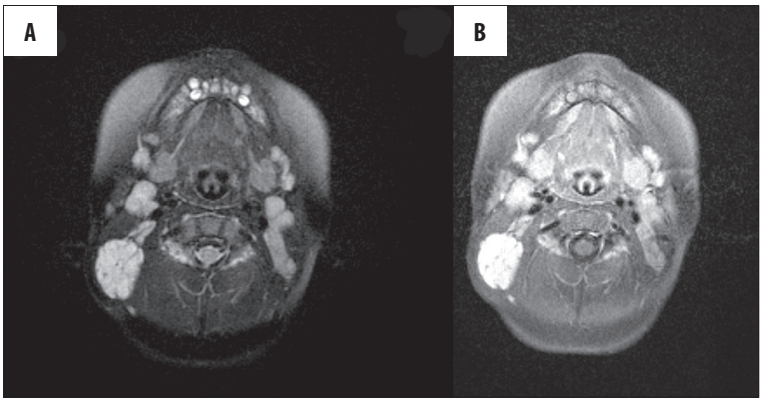

Figure 1. 11-month-old girl with a hemangioma of the right cervical region. (**A**) T2-weighted image with fat suppression, transverse plane. (**B**) T1-weighted image with fat suppression after contrast administration, transverse plane.

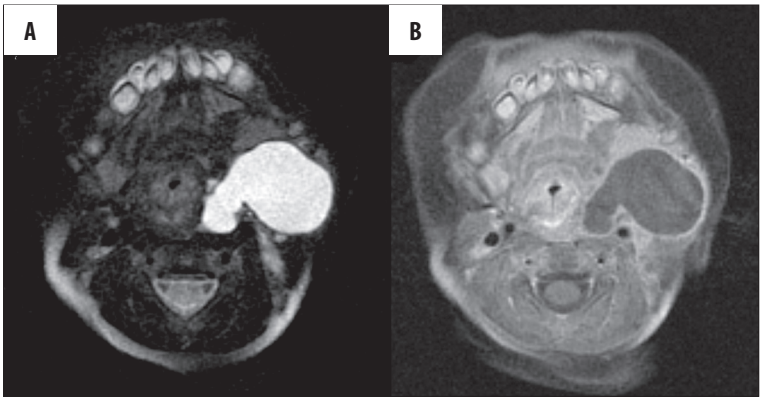

Figure 2. 2-month-old girl with a lymphatic malformation of the left parapharyngeal space. (**A**) T2-weighted image with fat suppression, transverse plane. (**B**) T1-weighted image with fat suppression after contrast administration, transverse plane.

precisely assess the size and extent of lesions [21] and their relationship to adjacent structures such as skin, subcutaneous tissue, muscles, nerves and bones [22–24], as shown by our results. The extent of pathology often remains underestimated in clinical [25] and ultrasound [10] examination; however, MRI technique allows for its precise visualization [26,27]. Moreover, imaging in several planes enables precise determination of topographic relationships [19].

Spin-echo, fat saturation sequences: T2-weighted and T1-weighted with contrast, are of the greatest value in the diagnostics of vascular malformations [19]. T2-weighted sequences with fat saturation can precisely evaluate of lesions located superficially and differentiation from subcutaneous fat tissue [28] and T1-weighted sequences after contrast administration enable determining their full extent due to characteristic enhancement. Gradient T2-weighted sequences that enable hemosiderin detection and assessment of rapid vascular flow are also useful [22].

Specific types of vascular anomalies present with characteristic picture in MRI examination and thus it is possible to differentiate them using this technique [7,29].

In the analyzed group lesions were of hypo- or isointense character in T1-weighted images and hyperintense in T2-weighted images, which is in accordance with literature data [19,30].

Hemangiomas are usually well-demarcated tumors, not infiltrating, hypo- or isointense compared to muscles in T1-weighted images [3,31]. They are hyperintense, often heterogeneous in T2-weighted images and exhibit strong contrast enhancement. Loss of signal in spin-echo sequences corresponds to rapid-flow vessels, often visible in the central part and on the peripheries of lesions [21]. In the examined group we found 9 hemangiomas (Figure 1) visualized as solid lesions, rather well-demarcated from the surroundings – 8/9 lesions, isointense in T1-weighted images – 7/9, hyperintense in T2-weighted images – 9/9, with homogeneous signal – 8/9 and strong contrast enhancement 7/9. Similar results were acquired in other studies – five cases of hemangiomas in a group of 23 vascular anomalies in a study by Meyer et al. are solid lesions of medium signal in T1-weighted images, high signal in T2-weighted images, enhanced following administration of contrast medium, with signs of rapid flow [29].

Lymphatic malformations often appear as heterogeneous, cystic structures of low or medium signal in T1-weighted and high in T2-weighted images and exhibit contrast enhancement within walls and septa [22]. Fluid levels are seen in case of large multicystic lesions [3,22]. Among studied patients there were two lymphatic malformations presenting with characteristic, multicystic image in MRI examination with enhancement of cystic walls (Figure 2). Similar results were described in the literature [32,33].

In MRI examination venous malformations are visible as lobulated, serpentine, poorly demarcated lesion with septa, hypo- or isointense in T1-weighted images. Due to the presence of venous sinuses they are hyperintense in T2-weighted images and do not exhibit signs of flow [1,3]. Rarely occurring but characteristic loss of signal on T2-weighted images or phlebolites, be caused by the presence of clots in veins (Figure 3A) [3,9,22,23].

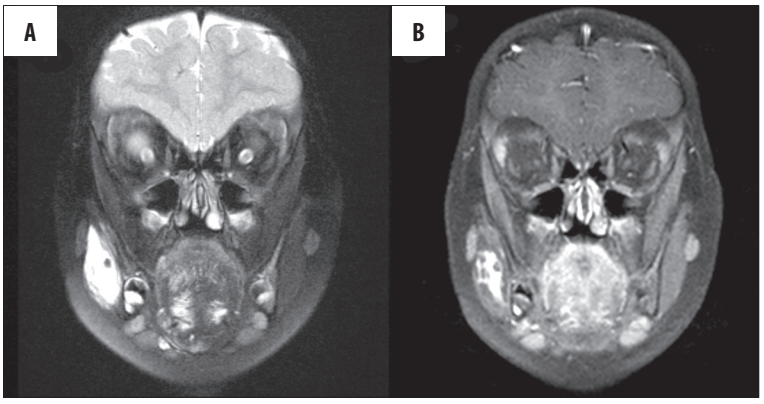

Figure 3. 2-year-old girl with a cystic lesion with marginal wall enhancement localized in the oral vestibule and the right cheek. Radiological and clinical picture argue for venous malformation(**A**) T2-weighted image with fat suppression, coronal plane. (**B**) T1-weighted image with fat suppression after contrast administration, coronal plane.

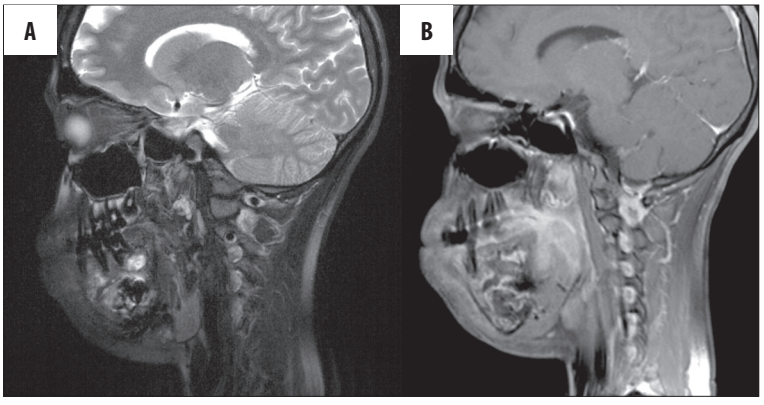

Figure 4. Multiple signal voids within expanded and deformed left part of mandible of 15-year-old girl. Radiological and clinical picture argue for arteriovenous malformation. (**A**) T2-weighted image with fat suppression, sagittal plane. (**B**) T1-weighted image with fat suppression after contrast administration, sagittal plane.

Diffuse contrast enhancement of the entire lesion, delayed in a dynamic study, is seen in venous malformations (Figure 3B). In the studied group venous malformations were usually poorly demarcated lesions (4 of 6 lesions). They were all characterized by medium signal in T1-weighted and high signal in T2-weighted images, usually involving neighboring muscles aside from subcutaneous tissue (4 of 6 lesions). All six patients underwent sclerotherapy, some of them several times, according to the methods used by other authors at other facilities [34].

In case of arteriovenous malformations MRI examination visualizes a lesion containing small component of tissue, consisting of dilated afferent and efferent vessels and numerous connecting vascular canals with visible loss of signal in spin-echo sequences, indicating high flow within them [23,35]. Early venous filling is visible following administration of contrast medium and magnetic resonance dynamic angiography with administration of contrast enables assessment of flow dynamics [36]. Differentiation from other high-flow malformations is possible due to characteristic serpentine loss of signal, lack of dominating tissue mass and frequent involvement of bone structures with attenuation of signal in T1-weighted sequences [4,23,29,36]. One arteriovenous malformation and its feeding vessel consisting of numerous vascular structures located within mandibular corpus was visualized in our study (Figure 4).

Capillary malformations usually do not require imaging except for cases of coexisting developmental abnormalities [3]. MRI examination may show nonspecific thickening of skin and subcutaneous tissue [37].

Specific types of malformations may occur within one vascular lesion as mixed malformations. Administration of contrast medium enables differentiation between solid and cystic parts of pathological mass, which is in accordance with our observations. Assessment of the venous and lymphatic component in slow-flow malformations consists of various patterns of contrast enhancement. On the other hand, in case of lymphatic malformations only cystic septa and walls undergo enhancement [38]. There were two mixed malformations with a venous and lymphatic component diagnosed in our study group.

Magnetic resonance technique is also appropriate for monitoring of treatment response and follow-up of possible recurrences [7,22].

Conclusions

Magnetic resonance examination preceded by a careful clinical examination is an effective and safe imaging modality. It aids in characterizing vascular anomalies in children. It enables thorough assessment of lesions and thus, proper planning of treatment. It may be useful in monitoring of therapeutic progress. MRI is advantageous compared to ultrasound in the evaluation of the extent of lesions and their topographic relationships.

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