Pediatr Radiol (2010) 40:895–905
DOI 10.1007/s00247-010-1621-y

REVIEW

Vascular anomalies: what a radiologist needs to know

Josée Dubois · Marianne Alison

Received: 5 February 2010 / Accepted: 8 February 2010

Abstract Most haemangiomas and vascular malformations are identified according to clinical criteria. A good knowledge of the classification and clinical characteristics of the vascular anomalies is necessary when managing these patients. However, some cases are challenging either because of an atypical presentation (e.g., soft-tissue mass with normal overlying skin) or because of classification difficulties. Doppler US and MRI are the two main imaging modalities that allow classification of the vascular anomalies and are useful in those clinically uncertain cases to establish the correct diagnosis. This aids the choice of the most appropriate treatment and to inform the parents of the prognosis. High-resolution grey-scale and Doppler US allow excellent visualization of most superficial masses. Doppler US is the easiest way to assess the haemodynamics of a vascular lesion and to clarify a doubtful diagnosis between a haemangioma and vascular malformation. MRI is the best technique for evaluating the extent of the lesions and their relationship to adjacent structures. While newly developed drugs from angiogenesis research labs are awaited, radiologists have an important role in the treatment of haemangiomas and vascular malformations. Intervention remains crucial in cases of alarming haemangiomas and venous malformations (VM), lymphatic malformations (LM) and arteriovenous malformations (AVM). A multidisciplinary team, including paediatricians, haematologists, surgeons and radiologists, must manage the problem cases both in terms of diagnostic work-up and therapeutic options. This paper will briefly discuss the imaging findings and treatment of vascular anomalies.

Keywords Vascular anomalies · US · MRI · Children

J. Dubois (✉) · M. Alison
Department of Medical Imaging, Sainte-Justine Hospital,
3175 Cote Ste-Catherine,
Montreal, QC H3T 1C5, Canada
e-mail: josee-dubois@ssss.gouv.qc.ca

Introduction

Vascular anomalies comprise a wide spectrum of lesions involving all parts of the body. In the past, diagnosis and treatment of vascular anomalies was hampered by a considerable confusion due to the use of improper terminology. A biological classification has helped resolve the confusion regarding terminology in the field of vascular anomalies. In 1982, on the basis of cellular kinetics and clinical behaviour, Mulliken and Glowacki [1] proposed the most helpful classification for vascular anomalies [2]. In this classification, vascular anomalies are divided into vascular tumours (cellular proliferation and hyperplasia) and vascular malformations (lesions that arise by dysmorphogenesis and exhibit normal endothelial turnover) (Table 1). In 1996, this classification was adopted by the International Society for the Study of Vascular Anomalies (ISSVA).

The most frequent vascular tumours in infancy are infantile haemangiomas. Congenital haemangiomas (NICH: non-involuting congenital haemangiomas, or RICH: rapidly involuting congenital haemangiomas), haemangioendotheliomas, tufted angiomas and sarcomas are other vascular tumours seen in children.

Vascular malformations are classified into slow-flow malformations including capillary malformations (CM), venous malformations (VM), lymphatic malformations (LM), capillary and venous malformations (CVM), capillary lymphatic and venous malformations (CLVM), and high-flow malformations including arteriovenous fistula (AVF) and arteriovenous malformations (AVM).

Complex-combined malformations are found in some syndromes: Klippel-Trenaunay, Parkes-Weber, Blue Rubber Bleb, Proteus and Maffucci.

Most haemangiomas and vascular malformations are recognized on clinical grounds. Clinical evaluation and genetics are extensively discussed in the literature. We will

Table 1 Classification of vascular anomalies

Vascular tumours	Vascular malformations	
Haemangioma	Simple	Combined
Proliferative phase	Capillary malformation	AVF, AVM, CVM, CLVM
Involutive phase	Lymphatic malformation (macro, micro, mixed)	LVM, CAVM, CLAVM
Others	Venous malformation	

AVF, arteriovenous fistula; AVM, arteriovenous malformation; CAVM, capillary—arteriovenous malformation (Parkes Weber syndrome); CLAVM, capillary—lymphatic—arteriovenous malformation; CLVM, capillary—lymphatic—venous malformation (Klippel-Trénaunay syndrome); CVM, capillary—venous malformation; LVM, lymphatic—venous malformation

focus on the implications for the radiologist in assessing vascular anomalies. The radiologist has to establish the diagnosis using colour Doppler US and MR imaging and should be aware of associated syndromes or the possibility of multiple organ involvement to be able to recommend appropriate additional imaging investigations. Lastly, interventional radiologists play a major role in the treatment of vascular malformations with the increasing efficacy of sclerotherapy and embolization therapies.

Vascular tumours

Infantile haemangiomas (in proliferative phase)

The best diagnostic clue for infantile hemangioma is the presence of high flow soft tissue mass.

US findings Hyperechoic and/or hypoechoic lesions can be seen. A single or a few vessels may be visible at Grey-scale US that most often correspond to arteries. The lesion displays increased colour flow due to numerous arteries and veins. The spectral analysis displays low resistance. Direct arteriovenous shunting is rare but may be seen and is frequently misinterpreted as an AVM (Figs. 1 and 2) [3].

MR findings MRI typically shows a well-defined, non-infiltrating lesion, with an intermediate signal intensity on T1-weighted sequences and increased signal intensity on T2-weighted sequences. Fast-flow vessels are identified by the presence of flow voids within and around the soft-tissue mass on spin-echo (SE) sequences and as high signal intensity on gradient-recalled echo (GRE) sequences. Perilesional oedema should not be seen. Vessels with fast-flowing blood are often at the periphery of the mass [4]. After gadolinium injection, strong enhancement is observed (Fig. 3).

Associations with infantile hemangiomas Multiple haemangiomas of the skin have traditionally been linked with potential visceral haemangiomas [5]. However, segmental haemangiomas are more frequently associated with anomalies like PHACE syndrome (posterior fossa anomalies, haemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects and eye anomalies) [6]. Most of these segmental haemangiomas are telangiectatic or reticular and do not present a high-flow pattern on Doppler US. Nevertheless, in our experience, internal organ involvement can be seen including the liver, gastrointestinal tract, pancreas and lung with imaging findings similar to soft tissue infantile haemangiomas.

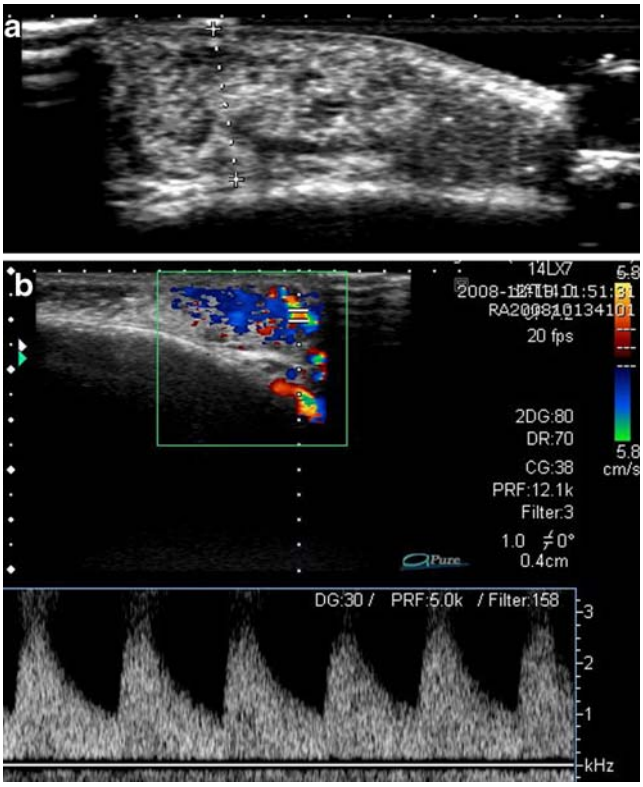

Fig. 1 Three-month-old girl. **a** *Grey-scale US* shows a hypervascular small soft-tissue mass located in the frontal region. **b** Numerous vessels and high velocity arterial flow with low resistance are typical of this classical haemangioma proven by imaging and clinical evolution

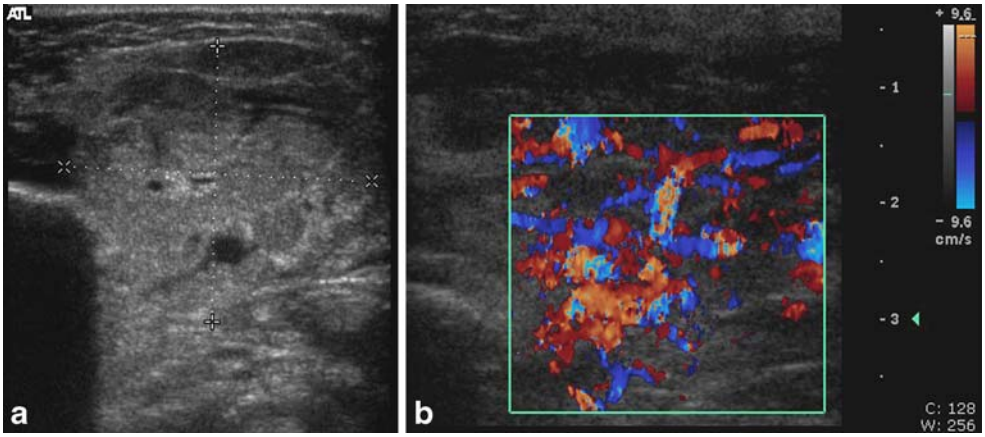

Fig. 2 One-month-old girl with a soft-tissue mass in her right cheek with normal overlying skin. **a** *Grey-scale US* shows a heterogeneous mass. **b** Doppler US demonstrates numerous vessels classical of infantile haemangioma

Take home point If the soft-tissue mass shows few arteries or veins, the spectral analysis shows a high resistance index and, if you notice perilesional oedema on T2-W MR imaging, rule out another tumoral lesion, e.g., sarcoma, neuroblastoma, myofibromatosis, tufted angioma, haemangiopericytoma, infantile myofibromatosis, fibrosarcoma, rhabdomyosarcoma, metastatic neuroblastoma or other tumours (Fig. 4).

Treatment In the majority of cases, no treatment is required because of spontaneous involution in cases of infantile haemangiomas and haemangioendotheliomas. Approximately 10–20% of all haemangiomas need to be treated. The major indications for treatment are periocular location with vision compromise, high-output cardiac failure, ulceration, compression of the airway, facial haemangiomas with rapid growth and distortion (presumed to result in important cosmetic sequelae) and symptomatic muscular haemangiomas. Medical treatment is usually attempted first with most of the vascular anomaly groups using propanolol as a first-line therapy with excellent results [7]. Other treatments include steroids, interferon, vincristine and laser. Embolization and/or surgery are required when medical alternatives are ineffective, mostly in cases of liver haemangiomas with cardiac failure that does not respond to pharmacologic treatment.

Congenital haemangiomas

The notion of congenital haemangiomas was first introduced in 1996 by Boon et al. [8]. Among this group, two sub-sets were subsequently identified: NICH that undergo a proportional growth with the child but no regression, and RICH in which regression is complete within 14 months after birth. It is noteworthy to mention that sometimes it may be clinically difficult to distinguish a common infantile haemangioma (CIH) from a RICH when the CIH is present and prominent at birth and undergoes quick but moderate proliferative phase. The GLUT1 marker is negative compared to infantile haemangiomas. However, in segmental haemangiomas the GLUT1 is positive [9].

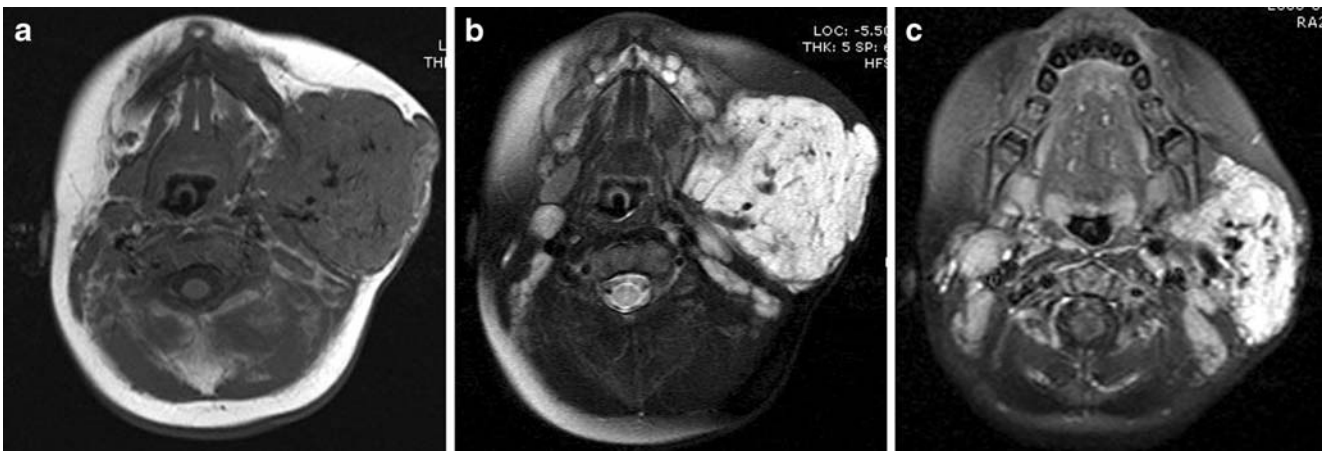

Fig. 3 Three-month-old girl. **a** Axial T1-W scan of the head shows a subcutaneous hypointense mass located in the parotid. Fast flow vessels are also identified by the presence of flow voids within the mass. **b** On axial T2-W FS scan the mass is hyperintense. Flow voids are still detected. No perilesional oedema was identified. **c** One month after propanolol treatment significant regression was noticed. Axial T1-W contrast-enhanced FS scan shows strong enhancement of the haemangioma

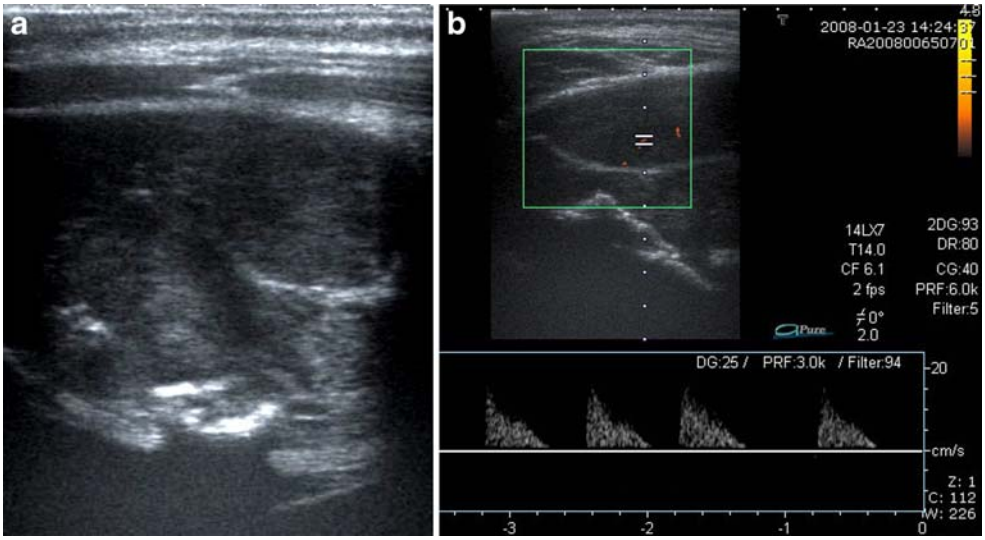

Fig. 4 One-year-old girl with a soft-tissue mass in the gluteal region. **a** *Grey-scale US* shows a soft-tissue mass in the gluteus maximus muscle. **b** Power Doppler US identifies a few vessels with high resistance index. Biopsy diagnosed high-grade undifferentiated sarcoma

US findings RICH and NICH share the same imaging findings. Most of these imaging findings are similar to those of infantile haemangiomas with some differences like various-sized vascular aneurysms, intravascular thrombi (never seen in infantile haemangiomas), increased venous component and arteriovenous shunting. It is important to be aware that some malignant tumours and AVMs have similar features (Fig. 5).

Treatment For NICH, since embolization is less effective, surgical resection is the best option.

Haemangioendotheliomas

US findings They are seen as ill-defined soft-tissue masses with variable echogenicity.

Calcifications can be present (never seen in infantile haemangiomas). Doppler US shows a high, moderate or low vessel density with high and low resistance index [10].

MR imaging T1-W MRI sequences show a heterogeneous soft-tissue mass that is isointense or hypointense compared to the muscle. T2-W sequences show a hyperintense lesion with subcutaneous stranding. Signal voids can be seen on GRE and represent haemosiderin or other blood products. Contrast-enhanced imaging displays diffuse, heterogeneous enhancement in the soft-tissue mass. Unlike infantile haemangiomas, destruction of the adjacent bones can be seen in haemangioendotheliomas (Fig. 6) [11].

Key facts Often associated with Kasabach-Merritt phenomenon, ill-defined lesion, calcifications.

Treatment Medical treatment is recommended. Steroids are the first-choice treatment. Interferon, vincristine and embolization are also useful in refractory steroids treatment. Embo-

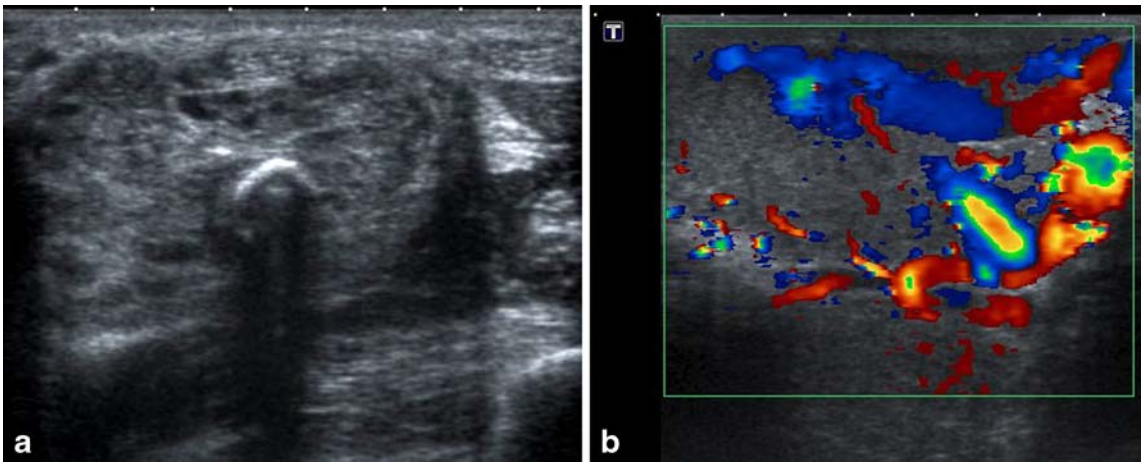

Fig. 5 Nine-year-old boy with a congenital mass of the forearm. **a** US shows a heterogeneous echogenic mass containing phleboliths. **b** Colour Doppler performed at the age of 3 years shows numerous arteries and veins. Biopsy confirmed the diagnosis of NICH with GLUT1 negative

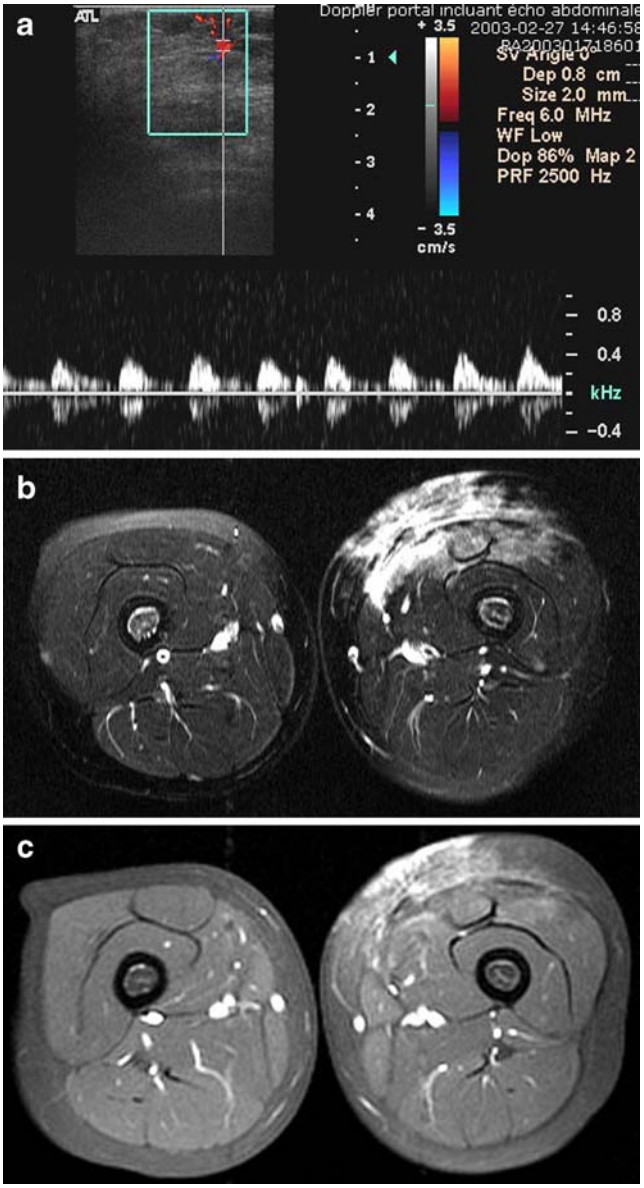

Fig. 6 Three-year-old boy with red/purple-coloured soft-tissue mass in the anterior thigh. **a** Pulsed-wave colour Doppler US shows a few vessels with high resistance index**. b** Axial T2-W FS image shows an ill-defined hyperintense subcutaneous and muscular lesion. Subcutaneous stranding is clearly seen. **c** Axial contrast-enhanced FS T1-W scan shows heterogeneous diffuse enhancement. Biopsy confirmed the diagnosis of haemangioendothelioma

lization is always associated with medical treatment. More studies are necessary in order to evaluate the effectiveness of propanolol in the treatment of haemangioendotheliomas.

Vascular malformations

Doppler US is the first examination to perform when a vascular malformation is suspected. This inexpensive and noninvasive examination allows differentiation between low-flow and fast-flow lesions.

Low flow vascular malformations

Venous malformations (VM)

Best diagnostic clue The presence of phleboliths.

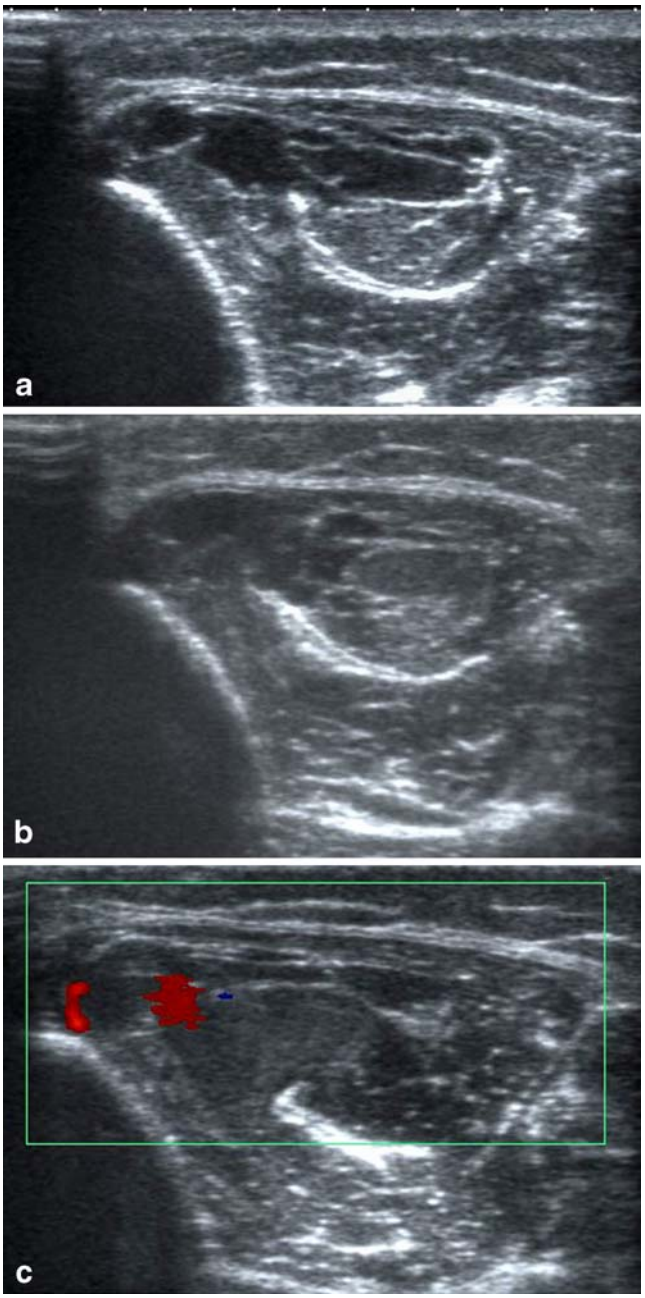

Fig. 7 Three-year-old boy with VM. **a** *Grey-scale US* shows a heterogeneous hypoechoic lesion with internal fluid component. The lesion was compressible (**b**) and after compression, filling of the cavities was seen with a few vessels on colour Doppler US (**c**)

US findings VMs can be classified in two types: cavitary and dysplastic. The cavitary VM is more common. Grey-scale US shows a compressible hypoechoic and heterogeneous infiltrative lesion [12, 13]. After applying compression, US shows the movement of blood into the cavities (Fig. 7) and phleboliths can be identified [12]. Doppler US typically shows no flow or monophasic low-velocity flow. Most of the time Doppler flow is difficult to obtain because of below-threshold flow or thrombosis. Dynamic manoeuvres, such as Valsalva or manual compression, are sometimes necessary to induce visible Doppler flow. Arterial flow can be observed in lesions with a capillary component. This is especially found in cases of CVM. Vessel density and Doppler shift are typically low. Sometimes, a high resistive index is present indicating a normal arterial vessel in the mass. Dysplastic VMs consist of multiple varicose veins. On B-mode imaging multiple anechoic tubular, tortuous channels infiltrating the subcutaneous fat, muscles, tendons or other tissues are observed. Doppler interrogation reveals slow venous flow.

MR findings MRI is an excellent modality to define the extension of the lesions and their relationship to adjacent structures. The examination protocol should begin with SE or fast SE T1-W sequence for basic anatomic evaluation. The extension of the malformation should be assessed with a T2-W sequence with fat suppression (FS). Fat suppression with short TI (inversion time) inversion recovery (STIR) T2-W sequence using a 512 matrix are well suited for this purpose. T2-W GRE sequences can also be used to demonstrate calcification or haemosiderin. On GRE sequences, the absence of signal in the blood vessel in the vicinity of the malformation suggests a slow-flow malformation [14]. FSE T1-W sequence with FS should be performed after gadolinium injection to evaluate the perfusion of the malformation. In our institution, we perform a dynamic perfusion study at 1, 2, 5 and 10 min after contrast infusion using a volumetric interpolated breath-hold examination (VIBE) sequence (Fig. 8). These contrast-enhanced 3-D acquisitions are also useful to appreciate the drainage of the malformation in the venous system [15]. Usually, VMs are hypo- or isointense on T1-W sequences. In cases of haemorrhage or thrombosis, a heterogeneous signal can be observed on T1-sequences. Abnormal veins can be observed in the area of the malformation. On T2-W sequences, VMs are of high signal. Areas of low signal can be observed related to thrombosis, sep-

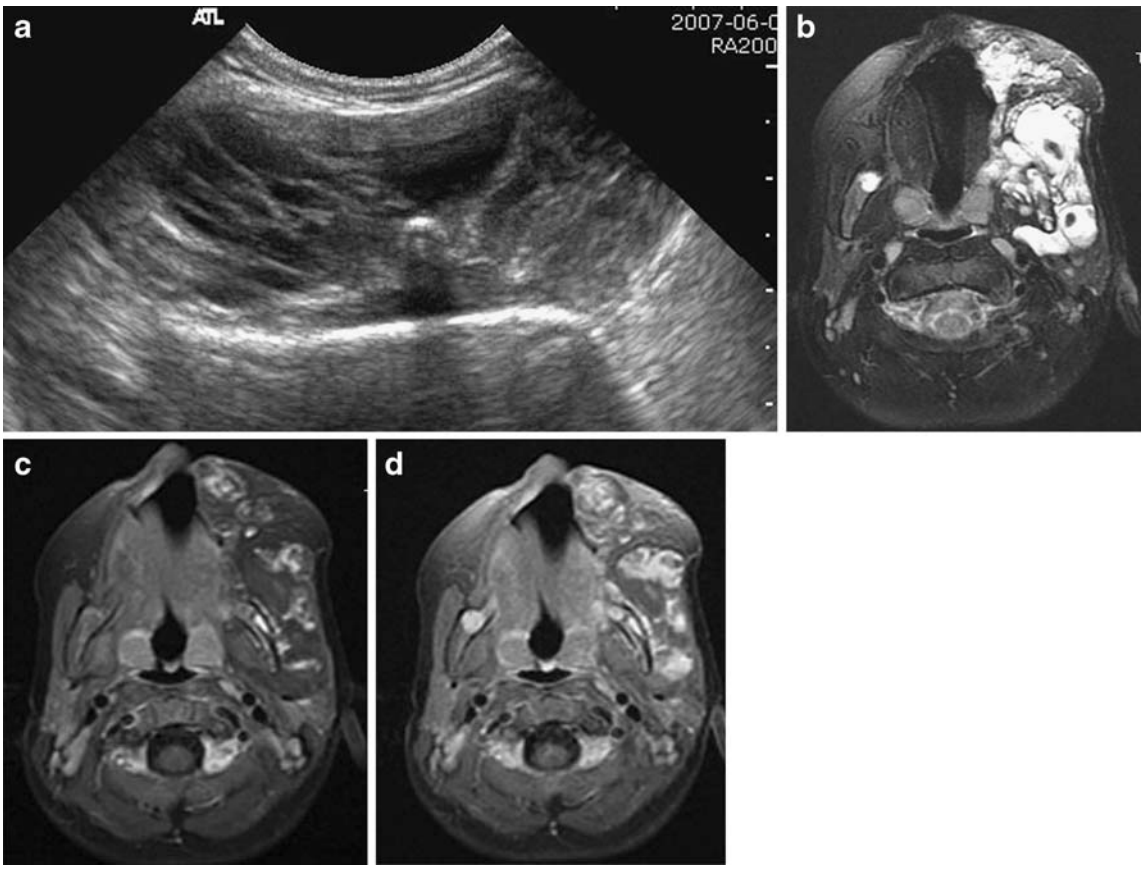

Fig. 8 Eight-year-old girl with a soft-tissue lesion of the cheek. **a** *Grey-scale US* shows a compressible soft-tissue hypoechoic lesion with phleboliths. **b** Axial T2-W FS scan shows the extension of the hyperintense VM containing phleboliths. **c**, **d** Axial contrast-enhanced T1-W FS images shows heterogeneous enhancement of the VM at 2 min (**c**) and 5 min (**d**) later

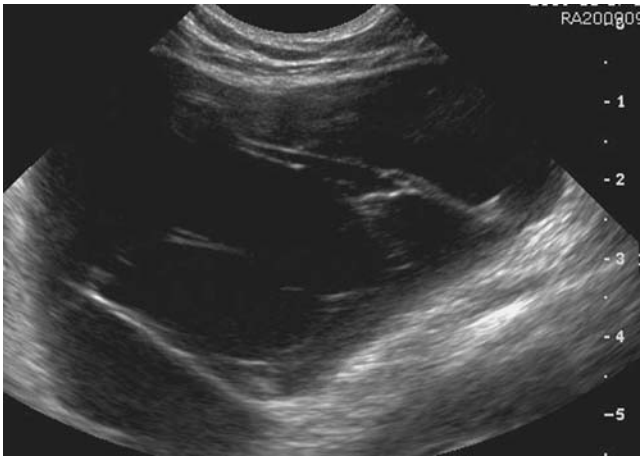

Fig. 9 Two-month-old boy. US shows a cystic lesion with multiple septa with a diagnosis of macrocystic LM

tation inside the malformation or phleboliths. On T2-W sequences, the extension of the malformation into adjacent structures is usually clearly delineated.

Treatment Most VMs are managed conservatively with compression bandage to the extremity. The indications for treatment are pain, articular involvement, disfigurement and gastrointestinal bleeding. The first-line treatment is sclerotherapy and can be followed by resection, laser and photodynamic therapy. Many sclerosing agents are used like dehydrated ethanol, sodium tetradecyl sulfate, polidocanol and bleomycin.

Lymphatic malformations (LM)

Best diagnostic clue Cutaneous angiokeratosis for microcystic LM and cystic lesions for macrocystic LM.

US findings Grey-scale US imaging is instrumental in characterizing the type of LM. Macrocystic LMs consist of multiloculated cystic lesions (Fig. 9) [13]. Pure microcystic lesions are ill-defined and hyperechoic due to numerous wall interfaces. Mixed lesions consist of cystic and solid lesion related to the size of the cyst. Colour Doppler reveals vascular channels in the septa, including veins and arteries as confirmed by spectral analysis.

MR findings On MRI the characteristic findings are the presence of a heterogeneous fluid-filled mass with an iso- to hyposignal on T1-W sequences and hypersignal on T2-W sequences (Fig. 10). Sometimes high signal on T1-W sequences or a fluid level can be observed in cases of a cyst with a high content of protein or haemorrhagic contents [14]. Pure LMs have absent or minimal enhancement of septa whereas combined lymphatic VMs show enhancement of the lymphatic space [14].

Treatment Macrocystic LMs can be treated either by surgery or sclerotherapy. Usually macrocystic LMs respond well to sclerotherapy. Numerous sclerosing agents are used: ethanol, sodium tetradecyl sulfate and doxycycline in North America; alcoholic solution of zein (Ethibloc, Ethicon, Somerville, NJ) in Europe and Canada [16, 17]; OKT3 [(Centocor Ortho Biotech Inc, Horsham, PA) picibanil, that is a killed strain of group A Streptococcus pyogenes] in Japan [18]. More recently, bleomycin has been used in macrocystic and mixed malformations with a success rate varying between 70 and 95% [19–24]. Microcystic LMs do not respond well to sclerotherapy, although a good response using bleomycin and OKT3 has been reported by several authors [22, 25]. Microcystic lymphangiomas should be managed conservatively but, if a treatment is required, the surgical approach should be favoured. Recurrence rates of

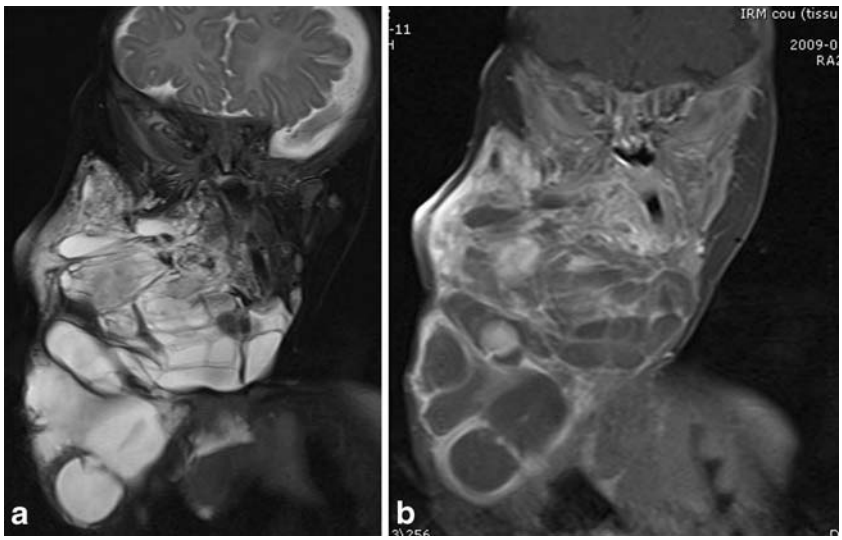

Fig. 10 One-month-old girl with macrocystic LM. **a** Coronal T2-W FS image shows a multiloculated hyperintense lesion. **b** Coronal contrast-enhanced T1-W FS scan shows a large macrocystic LM with enhancement of the septa

40% after incomplete excision and of 17% after macroscopically complete excision have been reported [26]. The role of bleomycin sclerotherapy combined with surgery is not yet certain.

Syndromes with vascular skin lesions and slow-flow malformations

Sturge-Weber syndrome is a nonheritable cutaneous disorder that consists of a unilateral facial port-wine stain in the trigeminal area; an ipsilateral leptomeningeal malformation and malformation of the choroid of the eye; atrophy and calcifications in the subjacent cerebral cortex; seizures; hemiparesis and visual field defects contralateral to the brain lesion; mental retardation of variable degree and sometimes buphthalmos or glaucoma [27].

Klippel-Trenaunay syndrome is a capillary venolymphatic malformation (CLVM) with limb overgrowth [28, 29]. Clinical presentation is variable and depends on the predominance of abnormal lymphatic or venous vessels. This syndrome includes a dermal capillary stain associated with venous varicosities. These anomalous veins have deformed, insufficient or absent valves. The pathognomonic marginal vein of Servelle is often identified in the subcutaneous fat of the lateral calf and thigh and can communicate with the deep venous system at various levels. Lymphoedema can be associated with malformations and hypoplasia of the lymphatic vessels. The lower limb is more frequently involved with possible extension of the malformation into the perineum or sometimes in the abdomen. The upper limb, the trunk or the neck are rarely involved [30]. Contrast venography is useful in selected patients to depict the route of drainage and the feasibility of resecting or sclerosing varicosities [2].

Blue rubber bleb nevus syndrome is characterized by multiple VMs of the skin with multiple gastrointestinal VMs. The gastrointestinal lesions can result in haemorrhage, intussuception and volvulus. It is a sporadic disease, but familial cases have been reported [31, 32].

Maffucci syndrome is a nonheritable syndrome that consists of diffuse enchondromatosis involving the metacarpal phalanges of the hands and feet associated with multiple venous or LMs [33].

Proteus syndrome consists of multiple subcutaneous hamartomatous tumours, hemihypertrophy, pigmented nevi, gigantism involving the extremities (hand, foot), intra-abdominal lipomatosis, pachydermia, macrocephaly, bony exostosis and lymphatic venous malformations (LVM) [34–37].

Bannayan-Riley-Ruvalcaba syndrome is an autosomal-dominant condition with a variable clinical phenotype. The disorder is associated with phosphatase and tensin homolog (PTEN) gene mutation on chromosome 10q. Clinical features include macrocephaly, pseudopapilloe-

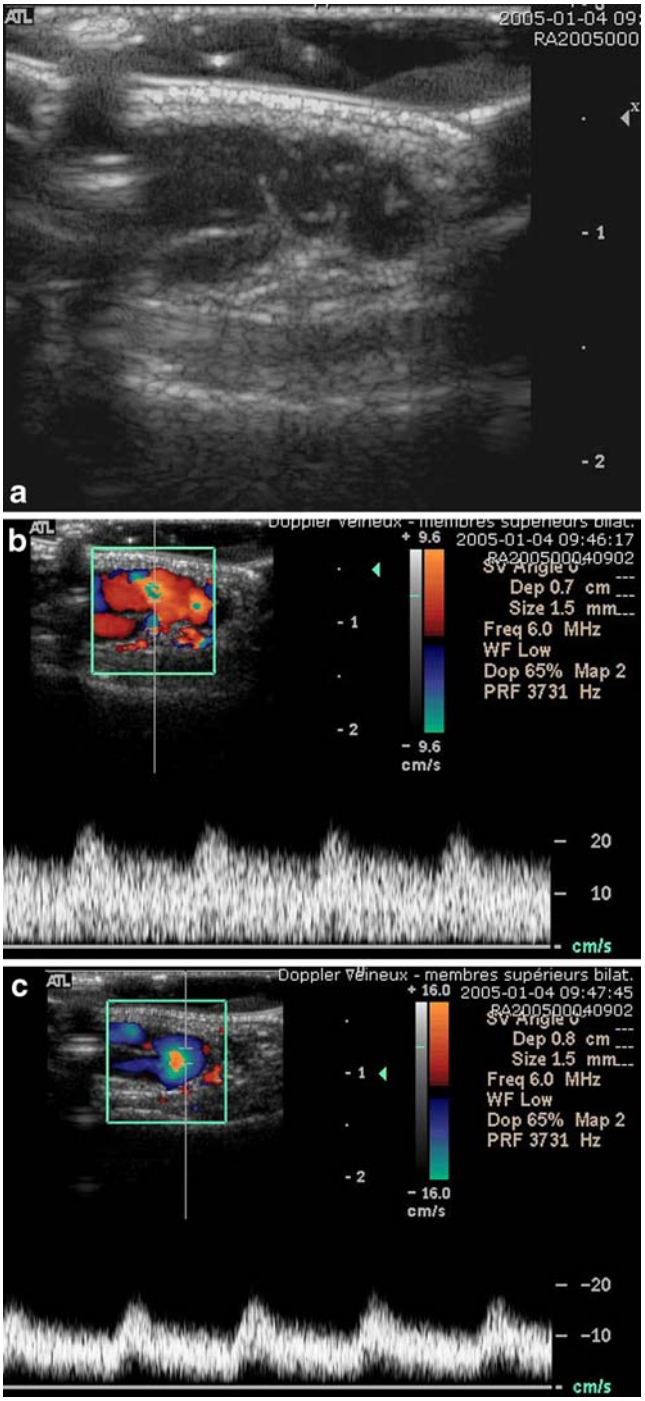

Fig. 11 Fifteen-year-old male with finger AVM. **a** *Grey-scale US* shows numerous tortuous vessels. **b** Doppler US of the pulsatile soft-tissue mass of the finger shows only high velocity arteries with a low resistance index. **c** Doppler US at another level shows pulsatile venous flow in a typical AVM

dema, pigmented maculas on the penis, gastrointestinal polyposis, visceral lipoma, thyroiditis, capillary and combined malformations.

Glomovenous malformation is an autosomal-dominant condition, also known as glomangioma. It is characterized by multiple, often tender, blue nodular dermal lesions in the skin. Glomovenous malformations are multifocal and painful to palpation. US imaging features are similar to those of VM with the exception that the lesion cannot be completely emptied by compression. The lesions are VMs with the presence of glomus cells.

High-flow vascular malformations

Arteriovenous malformations (AVM)

Best diagnostic clue Numerous visible arterial and venous vessels and high diastolic flow.

US findings AVMs are poorly defined with no or little tissue mass visible. Most of the time, fat tissue can be seen around the AVM. The lesion is made of multiple feeding arteries with increased diastolic flow and increased venous return with systolic/diastolic flow. Power or colour Doppler examination is helpful to delineate the network of the malformation (Fig. 11). Unlike haemangiomas, there is always arterialisation of all the draining veins (i.e. pulsatile flow) in AVMs.

MR findings MRI examination allows evaluation of the extension into adjacent structures, especially bone involvement. MR imaging findings include dilated feeding arteries and draining veins with little tissue matrix and no venous lakes [39]. Signal voids are typically observed in these vessels on both T1- and T2-W SE sequences, whereas hypersignal is observed on gradient-echo and angiographic sequence indicating a high-flow lesion [14]. Gadolinium-enhanced MR angiography is helpful to evaluate feeding arteries and draining veins. The presence of early venous filling is typically seen in AVMs. Using time-resolved MR angiography sequence it is now possible to evaluate the dynamic opacification of AVM (Fig. 12) [40–44]. Since these sequences have a high temporal resolution there is a compromise on spatial resolution that is lower than conventional 3-D MR.

Treatment AVMs are an important challenge for interventional angiographers. Some AVMs respond well to embolization while others progress despite embolotherapy. For this reason we recommend conservative management in quiescent AVMs. However, treatment is required in cases with severe cosmetic consequences, ulceration, pain associated with distal steal phenomenon and/or gangrene. Less commonly bleeding, compartment syndrome, severe overgrowth, congestive heart failure and failure to thrive can lead to intervention. Embolization is the first-choice treatment for AVMs. The procedure should be performed by well-trained angiographers and under general anaesthesia. To destroy the AVM and reduce the risk of recurrence, super-selective catheterization is necessary combined with a percutaneous direct puncture of the nidus when feasible. The best agent to destroy the nidus is dehydrated alcohol. Other agents can be used like onyx and histoacryl (N-butyl cyanoacrylate, not approved by the FDA).

Syndromes with high-flow malformations

Parkes-Weber syndrome involves a combination of AVFs, congenital varicose veins and a cutaneous capillary malformation associated with limb hypertrophy [45].

Rendu-Osler-Weber syndrome (hereditary haemorrhagic telangiectasia) manifests as diffuse mucosal telangiectasia

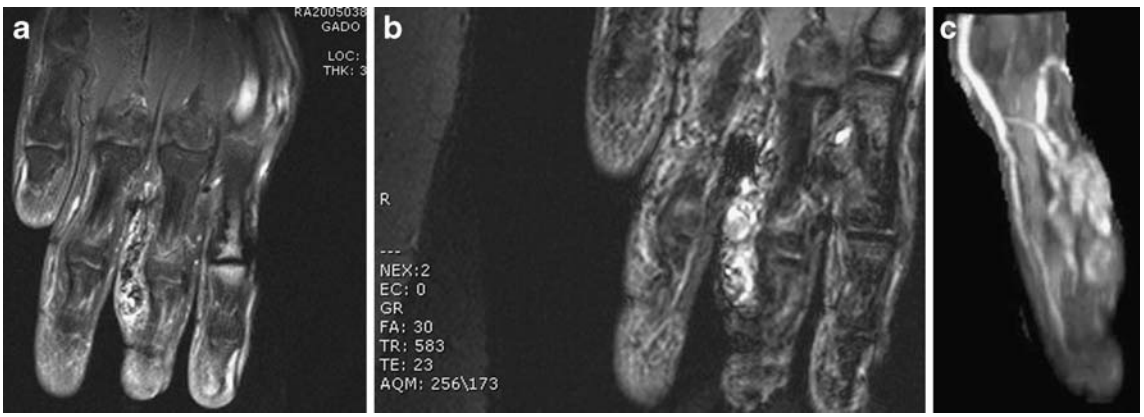

Fig. 12 MR imaging (same patient as Fig. 11). **a** Coronal T2-W FS scan of the hand shows flow voids. **b** Coronal gradient-echo T2-W image shows numerous tortuous bright vessels. **c** Contrast-enhanced 3-D acquisition illustrates the arterial feeding of the arteriovenous malformation

involving the nasopharynx, gastrointestinal tract and sometimes the urinary and genital mucosa. AVFs and arterial aneurysms involving the pulmonary, hepatic and digestive arteries are typically observed [46].

Capillary malformation–AVM syndrome is a hereditary disorder characterized by cutaneous capillary malformation associated with AVM or AVF. Mutations in the RASA1 gene are reported in this condition [38].

Cobb syndrome is a rare nonhereditary syndrome where a cutaneous capillary malformation is associated with AVM of the spinal cord [38, 47].

Conclusion

Diagnostic and interventional radiologists play a major role within a team dealing with vascular anomalies. They must be aware of the clinical history and findings to be able to make the right diagnosis and propose the best therapeutic options.

References

1. Mulliken JB, Glowacki J (1982) Classification of pediatric vascular lesions. Plast Reconstr Surg 70:120–121
2. Mulliken JB, Fishman SJ, Burrows PE (2000) Vascular anomalies. Curr Probl Surg 37:517–584
3. Dubois J, Patriquin HB, Garel L et al (1998) Soft-tissue hemangiomas in infants and children: diagnosis using Doppler sonography. AJR 171:247–252
4. Konez O, Burrows PE (2002) Magnetic resonance of vascular anomalies. Magn Reson Imaging Clin N Am 10:363–388
5. Metry DW, Hawrot A, Altman C et al (2004) Association of solitary, segmental hemangiomas of the skin with visceral hemangiomatosis. Arch Dermatol 140:591–596
6. Metry DW, Dowd CF, Barkovich AJ et al (2001) The many faces of PHACE syndrome. J Pediatr 139:117–123
7. Léauté-Labrèze C, Taïeb A (2008) Efficacy of beta-blockers in infantile capillary haemangiomas: the physiopathological significance and therapeutic consequences. Ann Dermatol Venereol 135:860–862
8. Boon LM, Enjolras O, Mulliken JB (1996) Congenital hemangioma: evidence of accelerated involution. J Pediatr 128: 329–335
9. North PE, Waner M, James CA et al (2001) Congenital nonprogressive hemangioma: a distinct clinicopathologic entity unlike infantile hemangioma. Arch Dermatol 137:1607–1620
10. Dubois J, Garel L, David M et al (2002) Vascular soft-tissue tumors in infancy: distinguishing features on Doppler sonography. AJR 178:1541–1545
11. Robertson RL, Robson CD, Barnes PD et al (1999) Head and neck vascular anomalies of childhood. Neuroimaging Clin N Am 9:115–132
12. Trop I, Dubois J, Guibaud L et al (1999) Soft-tissue venous malformations in pediatric and young adult patients: diagnosis with Doppler US. Radiology 212:841–845
13. Paltiel HJ, Burrows PE, Kozakewich HP et al (2000) Soft-tissue vascular anomalies: utility of US for diagnosis. Radiology 214:747–754
14. Siegel MJ (2001) Magnetic resonance imaging of musculoskeletal soft tissue masses. Radiol Clin North Am 39:701–720
15. Li W, David V, Kaplan R et al (1998) Three-dimensional low dose gadolinium-enhanced peripheral MR venography. J Magn Reson Imaging 8:630–633
16. Dubois J, Garel L, Abela A et al (1997) Lymphangiomas in children: percutaneous sclerotherapy with an alcoholic solution of zein. Radiology 204:651–654
17. Molitch HI, Unger EC, Witte CL et al (1995) Percutaneous sclerotherapy of lymphangiomas. Radiology 194:343–347
18. Ogita S, Tsuto T, Nakamura K et al (1994) OK-432 therapy in 64 patients with lymphangioma. J Pediatr Surg 29:784–785
19. Baskin D, Tander B, Bankaoğlu M (2005) Local bleomycin injection in the treatment of lymphangioma. Eur J Pediatr Surg 15:383–386
20. Muir T, Kirsten M, Fourie P et al (2004) Intralesional bleomycin injection (IBI) treatment for haemangiomas and congenital vascular malformations. Pediatr Surg Int 19:766–773
21. Orford J, Barker A, Thonell S et al (1995) Bleomycin therapy for cystic hygroma. J Pediatr Surg 30:1282–1287
22. Zhong PQ, Zhi FX, Li R et al (1998) Long-term results of intratumorous bleomycin-A5 injection for head and neck lymphangioma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 86:139–144
23. Zulfiqar MA, Zaleha AM, Zakaria Z et al (1999) The treatment of neck lymphangioma with intralesional injection of bleomycin. Med J Malaysia 54:478–481
24. Sanlialp I, Karnak I, Tanyel FC et al (2003) Sclerotherapy for lymphangioma in children. Int J Pediatr Otorhinolaryngol 67: 795–800
25. Sung MW, Lee DW, Kim DY et al (2001) Sclerotherapy with picibanil (OK-432) for congenital lymphatic malformation in the head and neck. Laryngoscope 111:1430–1433
26. Alqahtani A, Nguyen LT, Flageole H et al (1999) 25 years' experience with lymphangiomas in children. J Pediatr Surg 34:1164–1168
27. Enjolras O, Riche MC, Merland JJ (1985) Facial port-wine stains and Sturge-Weber syndrome. Pediatrics 76:48–51
28. Jacob AG, Driscoll DJ, Shaughnessy WJ et al (1998) Klippel-Trénaunay syndrome: spectrum and management. Mayo Clin Proc 73:28–36
29. Gloviczki P, Driscoll DJ (2007) Klippel-Trenaunay syndrome: current management. Phlebology 22:291–298
30. Cohen MM Jr (2000) Klippel-Trenaunay syndrome. Am J Med Genet 93:171–175
31. Moodley M, Ramdial P (1993) Blue rubber bleb nevus syndrome: case report and review of the literature. Pediatrics 92:160–162
32. Fretzin DF, Potter B (1965) Blue rubber bleb nevus. Arch Intern Med 116:924–929
33. Esterly NB (1996) Cutaneous hemangiomas, vascular stains and malformations, and associated syndromes. Curr Probl Pediatr 26:3–39
34. Wiedemann HR, Burgio GR (1986) Encephalocraniocutaneous lipomatosis and Proteus syndrome. Am J Med Genet 25:403–404
35. Clark RD, Donnai D, Rogers J et al (1987) Proteus syndrome: an expanded phenotype. Am J Med Genet 27:99–117
36. Samlaska CP, Levin SW, James WD et al (1989) Proteus syndrome. Arch Dermatol 125:1109–1114
37. Darmstadt GL, Lane AT (1994) Proteus syndrome. Pediatr Dermatol 11:222–226

38. Garzon MC, Huang JT, Enjolras O et al (2007) Vascular malformations. Part II: associated syndromes. J Am Acad Dermatol 56:541–564
39. Hovius SE, Borg DH, Paans PR et al (1996) The diagnostic value of magnetic resonance imaging in combination with angiography in patients with vascular malformations: a prospective study. Ann Plast Surg 37:278–285
40. Taschner CA, Gieseke J, Le Thuc V et al (2008) Intracranial arteriovenous malformation: time-resolved contrast-enhanced MR angiography with combination of parallel imaging, keyhole acquisition, and k-space sampling techniques at 1.5 T. Radiology 246:871–879
41. Reinacher PC, Stracke P, Reinges MH et al (2007) Contrast-enhanced time-resolved 3-D MRA: applications in neurosurgery and interventional neuroradiology. Neuroradiology 49:S3–S13
42. Saleh RS, Lohan DG, Villablanca JP et al (2008) Assessment of craniospinal arteriovenous malformations at 3 T with highly temporally and highly spatially resolved contrast-enhanced MR angiography. AJNR 29:1024–1031
43. Wu Y, Kim N, Korosec FR et al (2007) 3D time-resolved contrast-enhanced cerebrovascular MR angiography with subsecond frame update times using radial k-space trajectories and highly constrained projection reconstruction. AJNR 28:2001–2004
44. Ziyeh S, Strecker R, Berlis A et al (2005) Dynamic 3D MR angiography of intra- and extracranial vascular malformations at 3 T: a technical note. AJNR 26:630–634
45. Enjolras O (1997) Classification and management of the various superficial vascular anomalies: hemangiomas and vascular malformations. J Dermatol 24:701–710
46. Guttmacher AE, Marchuk DA, White RI Jr (1995) Hereditary hemorrhagic telangiectasia. N Engl J Med 333:918–924
47. Jessen RT, Thompson S, Smith EB (1977) Cobb syndrome. Arch Dermatol 113:1587–1590