Imaging features of pediatric vascular malformations of the skin and subcutaneous tissue.

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Learning objectives

To review and illustrate the radiological features of pediatric vascular lesions settled on the skin and subcutaneous tissues by pictures obtained at our Institution.

Background

Superficial vascular lesions (SVLs) are a heterogeneous group of entities characterized by the presence of vascular anomalies arising in the skin and subcutaneous fatty tissue. They are often presented in the childhood because most of them are congenital or arise during infancy, although some cases can be acquired later in life [1, 2].

Clinical diagnosis is usually easy because of its visual presentation and natural history, however sometimes, especially in the presence of palpable masses with normal skin, additional tests may be necessary because of the importance of an accurate diagnosis when planning treatment and expecting prognosis [1, 2].

In 1996 the International Society for the Study of Vascular Anomalies (ISSVA) approved a classification system modified from the original schema proposed by Mullikan and Glowacki [2, 3]. Their aim was to establish a common language for all the different medical specialists involved in the management of these lesions. Two groups of vascular anomalies were established:

- **Vascular tumours**: this group includes the classic hemangioma of the infancy (the most common type of vascular tumor) and other less frequent entities such as the rapidly involuting congenital hemangioma, the non-involuting congenital hemangioma, the Kaposiform hemangioendothelioma, the tufted angioma, the pyogenic granuloma or the glomangioma.
- **Vascular malformations**: subclassified according to the type of vessels that constitutes the lesion into high flowed (arteriovenous malformation and arteriovenous fistula) and slow flowed (including venular, venous and lymphatic malformations).

This classification is based on differences in the cellular kinetics and natural history of these lesions. Vascular tumors demonstrate endothelial cell hyperplasia, and the most common of these in children, hemangioma of infancy, spontaneously involutes. In contrast, malformations have flattened endothelial cells with normal endothelial cell turnover and do not involute spontaneously [2].
Although there are uncommon examples of either clinical or histologic coexistence of vascular malformations and tumors, this classification is still in force because of its simplicity and clinical relevance. The precise etiology of most of these conditions is not well known [2, 4].

Doppler sonography in most cases can provide important and definitive information with the advantages of its accessibility, safety and real-time evaluation of flow. Magnetic Resonance Imaging (MRI) or, in the fewer cases, Computerized Tomography (CT) are especially useful in the evaluation of large anomalies, with suspicion of other tissues involvement [1].

**Imaging findings OR Procedure details**

**A) VASCULAR TUMORS**

1. Hemangioma of the infancy

Hemangiomas of infancy are the most common benign tumors in children and have a distinctive natural history consisting of a proliferative phase in early infancy followed by an involutional phase, leading to complete and spontaneous regression in most patients.

These common vascular tumors occur in approximately 2.5% of all neonates and are seen in up to 5 to 10% of Caucasian infants by 1 year of age. They can occur in any race, been more frequent in girls three times as often as boys without a clear reason for this gender difference. Genetics have been proved its influence because approximately 10% of patients have a history of affected family members. Hemangiomas are also more common in preterm infants weighing less than 1500 g, and in infants whose mothers have undergone chorionic villus sampling [1, 5].

Anatomic location plays a critical role in determining whether complications could occur and if extra vigilance during the growth phase is necessary. Although any area of the body may be affected, approximately 60% occur on the head and neck. Less common anatomic sites include the trunk (25%) and extremities (15%). Hemangiomas can also develop in almost any internal organ [1, 2, 5].

**Imaging features**
In most cases the diagnosis of hemangioma is a clinical one, based on visual appearance and typical evolution. In a small minority of cases, specially largest and deepest ones, diagnosis is less obvious and imaging tests are necessary. Imaging studies can also be helpful in differentiating hemangiomas from many other entities, but biopsy may be necessary in very atypical presentations [1, 2].

**Doppler ultrasound**

Doppler ultrasound is the first imaging test to be performed when further analysis is indicated. The imaging behaviour of hemangioma of the infancy depends on the phase of its natural history.

- **Proliferative phase**: usually presented as a well-circumscribed solid mass, often hypoechoic to the surrounding soft tissue and containing anechoic channels because of its increased vascular density compared with the number of vessels per skin area.

![Imaging features of pediatric vascular lesions of the skin and subcutaneous tissue.](image)

**Fig. 1**

**References:** R. F. Ocete Pérez; Sevilla, SPAIN
Hemangiomas are fast-flow lesions with numerous high flow vessels around and within the soft tissue mass.

**Fig. 2**

*References:* R. F. Ocete Pérez; Sevilla, SPAIN

Spectral pulsed Doppler analysis shows decreased arterial resistance with increased systolic peak speed and increased venous flow with monophasic curve.
Fig. 3

References: R. F. Ocete Pérez; Sevilla, SPAIN
Some arterial signals with high diastolic component, pulsatile venous flow and signs of arteriovenous communications in small vessels can also be seen in this phase [1, 6].

- Involutional phase: in this stage the features gradually change to a low flow lesion ones. It is appreciated a decrease in the lesion volume, with loss of definition of the contours and heterogeneous appearance due to the presence of different types of tissue. There is also a progressive reduction of vascular density although systolic low-resistance high flow in the remaining vessels is conserved [1, 6].
CT and MRI

CT and MR both demonstrate well-circumscribed densely lobulated uniformly enhancing lesions with dilated feeding and draining vessels either at the centre or periphery [1].

Although the need of avoiding ionizing radiations in the pediatric age, CT-angiography may be useful for adequate representation of the vascular anatomy when treatment is necessary [1].

MRI is especially useful for assessing the lesion extension and involvement of adjacent tissues, being usually preferred to CT because of the lack of ionizing radiation. On MR hemangiomas behave as isointense or hypointense to muscle on T1-weighted images, and hyperintense on T2-weighted images. Flow voids are seen within and around the mass. The presence of hemorrhage or fatty replacement within the lesion in the
involutional phase can result in a heterogeneous signal on both T1 and T2 weighted sequences. A decreased vascular density is also evinced in this stage [1].

![Image of MRI scans](image.png)

**Fig. 5**

*References:* R. F. Ocete Pérez; Sevilla, SPAIN

**Conventional arteriography**

At present time, conventional arteriography as been replaced by the latest imaging techniques in the diagnosis of vascular tumors, being barely used for therapeutic purposes in the few complicated or life threatening cases, due to the spontaneous regression of hemangiomas. The findings described are single or multiple well-defined masses with intense and persistent contrast enhancement. The adjacent systemic feeding arteries may be increased in size and some early draining veins may be visible occasionally [1, 2, 7].
2. Other vascular tumors

- **Rapidly involuting congenital hemangioma**: typically present as raised violaceous tumors with ectatic veins, raised grayish tumors with overlying telangiectasias surrounded by a pale rim of vasoconstriction or as flat infiltrative tumors with violaceous overlying skin. They vary in size but are often several centimeters in diameter. They are frequently warm and occasionally have bruits or even a palpable thrill. In addition to their distinctive appearance, their behaviour also differs from hemangiomas of infancy, do not grow rapidly after birth and many involute extremely rapidly, completing involution by 12-18 months of age [2].

- **Non-involuting congenital hemangiomas**: also fully formed at birth always appearing as a solitary tumor. Typical lesions are round-to-ovoid in shape, plaque-like or bossed, with central or peripheral pallor as well as coarse, overlying telangiectasias. Lesions vary in size, from a few centimeters to 10-15 cm, with an average diameter of 5 cm. Most
have palpable warmth with a component of fast arterial flow that can be demonstrated by Doppler ultrasonography. Pathology typically reveals lobular collections of small, thin-walled vessels with a large, often stellate, central vessel. Interlobular areas contain predominantly dilated veins; arteries are also increased in number. These lesions do not involute over time but are usually easily excised without risk of recurrence [2].

- **Pyogenic granuloma:** one of the most common vascular tumors in children, second in frequency to hemangioma of infancy. Lesions can be seen at any age but the majority occur during childhood. Pyogenic granulomas usually present as rapidly growing, bright red papules varying in size from a few millimetres up to 2 cm. The head and neck are the most common locations, but lesions can occur at any site including mucosal surfaces. Pyogenic granulomas often bleed repeatedly and profusely, even with very superficial ulceration. The pathology of pyogenic granuloma is neither pyogenic nor granulomatous but rather consists of a lobular capillary hemangioma [2].

- **Tufted angioma:** also known as angioblastoma of Nakagawa, is an uncommon vascular tumor that usually has its onset during infancy or early childhood. Various presentations have been described, including solitary tumors, large infiltrated plaques, sometimes having increased lanugo hair and "port-wine stain-like" areas. The characteristic histology is of vascular tufts of tightly packed capillaries dispersed throughout the dermis. Some cases resolve, leaving only minor cutaneous changes whereas others persist and expand over time [2].

- **Kaposiform hemangioendothelioma:** is a rare, distinctive neoplasm that can occur in the skin but has also been reported as a retroperitoneal tumor. Affected infants have either a congenital tumor or develop a lesion soon after birth. In rare cases, the tumor arises within a pre-existing lymphatic malformation. Histologic examination reveals densely infiltrating nodules composed of spindled cells with minimal atypical and infrequent mitoses [2].

**B) VASCULAR MALFORMATIONS**

Vascular malformations are composed of anomalous blood vessels and/or lymphatics lined by a quiescent endothelium without cellular hyperplasia. Although it is believed that all vascular malformations are present at birth, most are evident later in infancy. A minority appear in childhood or later, after a dormant phase, as "acquired" vascular anomalies [2, 8].

Depending on the flow characteristics, vascular malformations are defined as either slow flow or fast flow. The slow-flow anomalies include capillary, venous, and lymphatic
malformations and combinations thereof. Arteriovenous malformations (AVMs) and arteriovenous fistula with arteriovenous shunting are the fast-flow anomalies [1, 2].

Vascular malformations persist lifelong, unchanged, growing proportionate to the child's growth or, in some cases, worsening and expanding. They never regress spontaneously but only rarely go through a rapid growth phase such as is seen with hemangiomas of infancy. Growth in vascular malformations may be stimulated by trauma, clotting, the effects of hormones during puberty and/or pregnancy, or may occur in the absence of any known triggering factor [2].

1. Capillary malformations

Capillary malformations are the most common vascular malformations. They involve vessels of the capillary network in skin and mucous membranes. Capillary malformations may be isolated and innocuous, may cause disfigurement and stigmatization and, in some cases, may herald the presence of an extracutaneous disease [2].

The term capillary malformations include different kind of lesions:

- **Salmon patches**: consist of ectatic capillaries that have been thought to represent the persistence of fetal circulatory patterns in the skin. Their disappearance may be based on maturation of the autonomic innervation of these vessels in early infancy. The salmon patch is present at birth in about 40% of infants and appears as a pink to red. In contrast to the port-wine stain, the salmon patch tends to be located in the central portion of the face and does not follow a dermatomal distribution [2].

- **Port-wine stains**: also called nevus flammeus, are present at birth and do not undergo spontaneous resolution. These well-demarcated vascular stains grow in proportion to the growth of the child. They are usually unilateral and segmental, generally respecting the midline. The stain may appear on any area of the body. Histologically, the port-wine stain is composed of normal numbers of ectatic mature capillaries in the superficial dermis with no evidence of cellular proliferation. These vessels become more dilated over time and are found in the deep dermis and subcutaneous tissue when the clinical lesion is raised or nodular [1, 2].

Because its clinical diagnosis, based on the visual examination of the involved skin, no further imaging investigation is needed.

2. Venular malformations
Initially called capillary malformations by Mulligan, this term has been replaced because of the histological features of these lesions, showing anomalous small postcapillary venules surrounded by disorganized collagen fibers arising in papillar and upper reticular dermis [1, 2].

**Imaging features**

No imaging assessment is needed in these cases. When performed, sonography may show thickening of the subcutaneous tissue and presence of few prominent veins. Doppler evaluation demonstrates no increase in vascular density [1].

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**Fig. 7**

References: R. F. Ocete Pérez; Sevilla, SPAIN

3. **Venous malformations**

Venous malformations are usually evident at birth and may occur with associated abnormalities as a part of a complex syndrome.
These lesions comprise anomalous dilated veins with irregularly thickened walls, which often have focal regions lacking smooth muscle cells. They extent by developing interconnected channels, which dissect through normal tissues. Thrombosis secondary to its slow flow gives rise to calcifications, which can become progressively more evident over time as phleboliths [1].

When arising in the limbs, they extent oriented parallel to the long axis of them and also to the planes of the fascia. The comparative images show adjacent muscle atrophy and hypertrophy of subcutaneous tissue [2].

The clinical presentation, evolution and management of these vascular anomalies differ depending on the age of the patient, the severity and the anatomic location, but they always share some common features such as a blue hue to the involved skin and mucous membranes, and its volume fluctuation dependent on changes in venous pressure (crying, exercise, Valsalva maneuver). Pain is variable but fairly common. Slow enlargement is commonly observed over time [2].

**Imaging features**

Radiologic approach is usually helpful in assessing the extent of involvement and in differentiating venous malformations from other vascular anomalies and soft tissue tumors.

**Conventional radiographies**

Calcifications due to the presence of phleboliths may be seen within the lesions. Adjacent bone changes such as developmental hypoplasia or sclerotic involvement with benign-appearing periostitis can be also present [1].

**US Doppler**

Venous malformations are presented as heterogeneous poorly delineated masses with multiple compressible vascular channels (98%). Most cases (82%) they are hypoechoic (82%) compared to adjacent subcutaneous tissue, although they can be hyperechoic (10%) or isoechoic (8%).
Fig. 8

References: R. F. Ocete Pérez; Sevilla, SPAIN
When present, rounded hyperechoic formations with posterior acoustic shadowing can be identified corresponding to phleboliths.
On Doppler US analysis, venous malformations generally show low vascular density with slow and turbulent flow, being more frequently monophasic (78%) than biphasic (6%) or absent (16%) [1, 2].

CT

There is rarely a need for CT scans when imaging assessment is indicated, but it can be performed to look for associated bony defects [1].

Venous malformations behave as heterogeneously low-density poorly circumscribed masses, with or without phleboliths within, showing a periphery slow progressive enhancement after contrast media administration [1].
MRI imaging is considered to be the best single imaging modality to characterize the lesion as distinct from the high flowed ones and for assessing its extent and relationship to adjacent structures, especially in deep locations. Although it may not require intravenous contrast, however, a better characterization of flow can be achieved with dynamic study [1, 2].

Venous malformations are showed as heterogeneous lesions composed of multiple vertiginous structures. It gives a bright hypersignal on T2-weighed spin-echo sequences, because of slow flow, and this clearly indicates the extent of the lesions throughout the involved tissues. On T1-weighted sequences they are often hypointense to fat but hyperintense to muscle. VM demonstrate multiple hyperintense fibroadipose septa on both T1 and T2. They may present areas of hemorrhage or thrombosis. No afferent artery or venous drainage is seen [1, 2].
**Conventional angiography**

Sometimes limb phlebography by direct puncture of the lesion is still used to plan sclerotherapy procedures. In addition to demonstrating the size and scope, it is useful to identify the drainage into the venous system [1].

![Image](https://example.com/image1.png)

**Fig. 11**

**References:** R. F. Ocete Pérez; Sevilla, SPAIN

4. **Arteriovenous malformations (ACMs)**

AVMs are fast-flow vascular malformations with direct arteriovenous communication without an intervening normal capillary bed. They are considered to be one of the most dangerous types of the vascular anomalies. No racial or gender predominance has been described [2].

AVMs are noted at birth in 40% of patients, being usual the progression during childhood related to some trigger factors as puberty, pregnancy or trauma (including iatrogenic
causes such as subtotal surgical resection, proximal artery ligation or partial arterial embolization) [2].

AVMs can be classified according to the Schrodinger staging system into four clinical stages: 1, dormancy; 2, expansion; 3, destruction; and 4, destruction plus congestive cardiac failure. In stage 1, AVMs mimic a port-wine stain or an involuting hemangioma, or they create a small pulsatile mass under normal skin. In stage 2, the expansion creates plaques or masses, which are red and warm, with local tenderness, pulsations, bruits and enlarged tortuous veins. In stage 3, skin necrosis, torpid ulcers, bleeding and hemorrhage become evident and lytic bone lesions may develop. Stage 4 is rare, occurring in approximately 2.5% and consists of increasing congestive cardiac failure from increased arterial pressure [9].

These kinds of vascular lesions often posed differential diagnosis with some vascularized tumors such as angiosarcoma, rhabdomyosarcoma and some others mesenchimal sarcomas. The presence of intralesional fat, muscle atrophy and the absence of peripheral edema should make us think more in AVMs. However, lesions with increased vascularity in the periphery or those with rapid growth and aggressive behaviour require a histological study [2, 9].

**Imaging features**

Once the diagnosis of AVM is considered, certain investigations are always necessary.

**Conventional radiography**

It is useful to assess adjacent bone changes such as hyperostosis or lytic destruction [1].

**Doppler US**

US evaluation can help to confirm the high-flow nature of the condition and will provide measurements of the comparative output between arteries on the affected and unaffected sides of the body [1].

AVMs usually appear as poorly delineated heterogeneous masses with high density of both arterial and venous vessels. Spectral pulsed Doppler evaluation reveals increased systolic arterial flow and arterialized biphasic venous curves. If the output is markedly increased on the affected side, cardiac evaluation and follow-up become mandatory [1].
Fig. 12

References: R. F. Ocete Pérez; Sevilla, SPAIN
Although MRI evaluation is considered a better alternative, CT-Angiographic studies with multislice technique can give a suitable map for surgical or interventional procedures. AVMs show huge enhancement in arterial vascular phase after contrast media administration; usually demonstrating numerous enlarged afferent vessels and early efferent veins. No persistent tissue enhancement is seen, giving the clue to differentiate these lesions from hemangiomas. Effects on the bone are best evaluated with CT scan, which can identify complications such as draining veins penetrating the skull and draining into an intracranial sinus [1, 2].

**MRI**

MRI demonstrates the presence of a vascular nidus rather than a dominant solid mass as multiple serpinginous hypointense channels on both T1 and T2 weighted sequences. Usually with the flow void phenomenon related to its high speed and turbulence. The flow
void phenomenon is not always present, but it is a reliable sign of high flow malformation. AVMs can be visualized as hyperintense lesions on gradient echo sequences due to its high flow. Afferent artery and efferent vein usually appear dilated and tortuous. Some hyperintense areas in T1 weighted sequences may be seen due to hemorrhage or thrombosis [1, 2].

Fig. 14

References: R. F. Ocete Pérez; Sevilla, SPAIN

Conventional angiography

It is used to delineate the vascular anomaly and to plan the treatment. Usually shows the dilated and elongated afferent arteries, the tortuous vascular nidus and early venous drainage by dilated efferent veins [1].

5. Lymphatic malformations (LMs)
Lymphatic malformations, also called lymphangiomas, are disorders of the lymphatic system and of the circulation of lymphatic fluid secondaries to lack of communication between the embryonic lymph sacs and the lymphatic draining vessels. Several subcategories appear in the medical literature based on clinical features, but it is probably more helpful to classify them according to the cyst volume into microcystic (cysts < 2 cc, corresponding to lymphangioma circumscriptum and cavernous lymphangioma), macrocystic (cysts > 2 cc, previously called cystic hygroma) or mixed malformations [1, 2].

LM arises more frequently in the soft tissues of head and neck (over 50% of cases) than those of trunk and limbs (over 40%). Visceral involvement is described in up to 10%, but no involvement has been described in central nervous system [1, 2].

Common complications of lymphatic malformations include disfigurement, infection and bleeding (superficial vesicles become hemorrhagic or there is a sudden hemorrhage in a large cyst) [2].

**Imaging features**

The diagnosis is often later in childhood and based on clinical examination, but is best confirmed with imaging techniques, which often demonstrates fluid levels to suggest cystic components and shows the extent of the malformation [1].

**Conventional radiography**

LMs may cause both hyperostosis and bone destruction in the adjacent skeletal structures [1].

**Doppler US**

Its monographic appearance varies from macrocystic to microcystic type.

- **Macrocystic**: presented as multilocular cystic masses with large thin-walled cysts containing variable echogenicity. Complicated cases (infected or bleeding ones) may show solid mass appearance. The internal septa may have variable thickness, sometimes showing some vessels [1].
Fig. 15

References: R. F. Ocete Pérez; Sevilla, SPAIN
Fig. 16

**References:** R. F. Ocete Pérez; Sevilla, SPAIN

- **Microcystic:** usually presented as hyperechoic masses as result of the multiple interfaces constituted by the small cysts walls. In these lesions, poorly arterial and venous flow is seen in the walls, allowing differentiating them from hemangiomas in proliferative phase [1].
Cysts are presented as hypodense masses that may show liquid levels in complicated cases. When contrast media is administrated, peripheric Wall enhancement may be seen [1].
Especially useful in the diagnosis of huge masses with deep extension, adjacent tissues involvement and mixed vascular-lymphatic origin.

Macrocystic malformations are multiseptated masses with low signal in T1-weighted sequences and hyperintense behaviour on T2-weighted ones. Internal signal may vary depending on the protein content or the presence of hemorrhage, showing liquid-liquid levels in cases of infection or bleeding [1, 2, 10].
Microcystic malformations are presented as solid homogeneous masses with intermediate signal intensity on T1 and high signal on T2 sequences, being characteristic the lack of significative enhancement after gadolinium administration. Lymphedema of the adjacent tissues may also be present [1, 10].

**Association of multiple vascular anomalies**

The association of different types of vascular anomalies is not unusual in the context of some clinical syndromes. The imaging features are the same as described in the isolated forms but combined [2].

The most relevant syndromes with vascular anomalies involvement are:
- **Klippel-Trenauny syndrome**: characterized by a triad of port-wine stain, varicose veins (venous malformation) and bony and/or soft tissue hypertrophy of the involved limb may also associate other low-flow malformations (venular or lymphatic ones) [2].

- **Parkes-Weber syndrome**: the presence of arteriovenous fistula distinguishes Parkes-Weber syndrome from KT syndrome. Other features of Parkes-Weber syndrome include warmth and tenderness overlying the malformation, dilated veins with a thrill on palpation, lengthening of the affected limb and hypertrophy in girth owing to both lipomatosis and lymphatic hyperplasia [2].

- **Mafucci syndrome**: characterized by the presence of multiple enchondromas associated with multiple hemangiomas. Also lymphangiomas may be present [2].

- **Sturge-Weber syndrome**: is the triad of a facial port-wine stain in the first trigeminal branch distribution, an ipsilateral leptomeningeal vascular malformation and a choroidal vascular malformation of the eye [2].

**Conclusion**

Most of vascular superficial anomalies are diagnosed by clinical criteria. Imaging tests can be useful to establish the diagnosis in cases of uncertain clinical appearance, to assess the extent and relationship of the lesion with the adjacent tissues and as a guide in monitoring therapy.

Doppler ultrasound and MRI are the main imaging techniques used in the pediatric age when further than clinical assessment is needed.

Pediatric radiologists should be familiar with the behavior and management of the various entities we review.

**Personal Information**

**References**