Diagnostic imaging of cervical vascular malformations

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Learning objectives

Show typical radiological findings of cervical vascular malformations.
Background

Vascular lesions were classified according to the presence of cell proliferation in hemangiomas or vascular malformations (Fig. 1).

Hemangiomas are benign tumors that mostly appear in the first weeks of life. They grow progressively until the first year and then regress spontaneously.

Vascular malformations are localized or diffuse abnormalities of the vascular system. The 40% is located in the head and neck. They are always present at birth, but sometimes are not visible until weeks or months later. They are less common than hemangiomas and not show a clear preference for either sex.

Not regress spontaneously, not have cell proliferation and they are slow growing and progressive in relation to trauma, infections, hormonal changes or pressure in the blood or lymph.

They are classified according to the predominant conduit: in arterial, venous, capillary, lymphatic or combinations. They can be single or multiple lesions can be subdivided into high and low flow. The pathogenesis of vascular malformations is not entirely clear, but there is some evidence of genetic alterations that alter angiogenesis and lymphangiogenesis.

The size varies from small lesions to large well circumscribed infiltrative lesions involving more than one anatomical compartment.

In recent years the incorporation of precise diagnostic methods has allowed change the diagnostic and therapeutic approach improving outcomes.

Radiography has been largely superseded by other imaging techniques and has limited value even to demonstrate the extent of bone involvement and the presence of calcifications.

Doppler ultrasound provides anatomical information in addition to hemodynamic data, such as speed and direction of flow, useful both high-flow malformations (arteriovenous) and in low flow (venous).
Computed tomography (CT) is significantly more sensitive and provides a much more accurate anatomical information with excellent visualization of bony structures and calcifications, but the best radiological investigation is magnetic resonance imaging (MRI), allowing demonstrate anatomical relationships, explore the surrounding tissues in contact with vascular malformations and also provides hemodynamic data.

**Classification:**

Mulliken and Glowacki in 1982 described a classification based on the different biological behavior, clinical course and histological features of vascular anomalies. Subsequently modified by Mulliken and Young was accepted by the International Society for the Study of Vascular Anomalies in Rome in 1996 and is now used with some modifications. It includes two main groups:

I. Classic infantile hemangiomas and other vascular tumors: Includes the classic infantile hemangioma most common and some rarer forms, comprising different behavior: rapidly involuting congenital hemangiomas and non-involuting, kaposiform hemangioendothelioma, and tusadas cells, granuloma pyogenic and other vascular tumors.

II. Vascular malformations are classified according to the type of vessel and the flow pattern consists in:

*High flow*: Includes the malformation and arteriovenous fistula

*Low flow*: Includes venular malformations, venous and lymphatic.

There are also some specific classifications for vascular malformations, excluding lymph, considering the predominant vascular component and timing of embryonic development in which the defect occurred. Have also been proposed morphological-anatomical classifications.

**Venous malformations**

They are the most frequent. They are formed by low flow ectatic vessels and are morphologically and histologically similar to veins.
They are divided into superficial or deep, localized or diffuse multicentric.

There may be a familial predisposition in some series has been detected more frequently in women.

We present as a spongy mass sometimes bluish as bluish, but may also have normal skin. They are soft, compressible, non-pulsatile generally expand after compression and Valsalva maneuvers, without increasing local heat.

In pharynx, larynx and palate can cause obstructive sleep apnea and the periorbital, eye involvement.

They may bleed or trombosarse causing pain and joint arthropathy. In large lesions may have hypertrophy of soft tissue and adjacent bone.

They have shaped calcifications or phleboliths dystrophic mineralization seen between 20 to 67% of cases, considered by some as a pathognomonic marker.

**Radiological findings** (Figures 2, 3, 4, 5, 6, 7)

**Radiography.** Calcifications can be observed corresponding to the lesion phleboliths. It can be seen hypoplasia, bone demineralization and sclerosis with periosteal reaction times.

**Ultrasound.** Venous malformations is usually hypoechoic, looking similar to cysts, confirmed with Doppler venous flow, especially after compression maneuvers, unlike other vascular malformations. Hipercogénicos foci can be displayed with acoustic shadowing in conjunction with phleboliths.

**CT scan.** Low attenuation lesion sometimes heterogeneous. It can be seen phleboliths or other calcifications. They have a slow uptake after contrast administration. We can identify the involvement of adjacent bone structures.

**RM:** It is the technique of choice because it allows to identify the extent of injury in various anatomical planes. Serpinginosas structures are observed, hyperintense on T2 and hypointense on T1 with respect to adjacent fat and both sequences are hyperintense with respect to the muscle. Septa can be observed, hyperintense on T1 and T2 and areas of bleeding or thrombosis.
Lymphatic malformations

Although congenital, only 65-75% are diagnosed at birth, reaching 80-90% at the end of the second year of life.

The most common location is the head and especially the neck (90%).

They are classified by the size of the cysts in: macrocystic with cysts larger than 2 cm³ and microcystic with cysts less than 2 cm³, which are within a solid matrix and are later diagnosis.

The clinical appearance varies depending on the size, location and depth of the lesion.

Complications can occur such as infection, bleeding or rupture of the lesion and compression of adjacent structures for growth.

Radiological findings (Figures 8, 9, 10).

Radiografía. Cuando the lesion involves the bone can cause hypertrophy or bone destruction.

Ultrasound. Multilocular cystic mass thin wall, with different content of echogenicity. In macrocystic form can be displayed variable thickness septa, occasionally with cups in its thickness. Complicated cases may simulate a solid mass.

Computed tomography (CT). Mass of low attenuation (when complicated with hemorrhage shows areas of increased attenuation). Sometimes it can be observed liquid-liquid levels. Cysts can show peripheral uptake after intravenous contrast administration.

MRI: Very useful for the study of large, deep or mixed formations commitment. Septate mass with low signal intensity on T1 and hyperintense on T2. In some cases the signal strength may be variable by the presence of protein or hemorrhage. You can observe fluid-fluid levels due to hemorrhage or infection and peripheral uptake of cysts or septa after intravenous gadolinium administration.
The microcystic are diffusely infiltrating solid masses with intermediate signal on T1 and high T2. There may be Lymphedema in adjacent subcutaneous tissue. Do not show gadolinium enhancement.

**Arteriovenous malformations**

They are the group that is diagnosed late, sometimes during the fourth to fifth decade of life.

They are located anywhere in the body, but are most frequent intracranial.

Exhibit an abnormal communication between efferent arteries and draining veins, without the intervention of a normal capillary bed.

Preferred book entitled arteriovenous fistula for traumatic variant acquired, formed by a single fistula.

Present clinically as pulsatile masses, with skin discoloration, increasing local heat and thrills. The proximal can lead to heart failure and distal ischemic changes.

Unlike venous malformations, not emptied completely after compression, are filled quickly and are firmer on palpation.

Within the group of vascular malformations are the most active, with greater potential for expansion and growth and at the same time, the most difficult to treat.

Generally, your extension histological far exceeds the clinical appearance, appreciating microscopic infiltration of the underlying tissue which favors recurrence after partial removal.

**Radiological findings:**

**X-rays:** may show destructive bone changes, intraosseous changes and overgrowth.

**Ultrasound.** It can be seen as an ill-defined heterogeneous mass. With Doppler study confirms the presence of afferent and efferent vessels, enlarged caliber, both arterial
and venous. You can identify high maximum systolic arterial flows and biphasic pulsatile venous flow or arterialized. AV fistulas have vascular flow fast, continuous and irregular.

**MRI:** Presence of a vascular nest, rather than a dominant solid mass. Multiple channels in T1 and T2 hypointense on SE, by the flow void phenomenon, related to its speed and turbulence. Hyperintense lesion in GE. Dilated afferent artery and efferent vein dilated and tortuous. You can have small T1 hyperintense areas due to hemorrhage or thrombosis. The flow void phenomenon is not always present, but when it does indicate a high flow malformation.

**Angiography:** Has a special indication in this vascular malformation, as a diagnostic and therapeutic method. Identify feeding arteries were dilated and elongated, vascular nidus and dilated tortuous channels and early venous return with dilated venous outflow (vascular nest is absent in fistulas).

It may be difficult to make the differential diagnosis with some vascularized tumors such as angiosarcoma, rhabdomyosarcoma and some soft tissue sarcomas. The presence of intralesional fat, muscle atrophy and absence of peripheral edema should be suspected arteriovenous malformation. Lesions with peripheral increased vascularity, with rapid and aggressive growth require a pathological study.

**Mixed or combined malformations**

They are relatively common, venular in particular associated to malformations deep (the arteriovenous, lymphatic or combined).

Syndromes have been described presenting combined vascular malformations associated with musculoskeletal disorders generally hypertrophy. Many known with the name of who described.

It should always be differentiation in high and low flow as it directly influences the treatment.
a) **Klippel-Trenaunay Syndrome.** Diffuse malformation that usually affects the lower extremities with venular vascular malformations, venous, lymphatic and hypertrophy of skeletal and soft tissue.

There is a lymphatic hypoplasia in 50% of cases with lymphedema and microcysts isolated. Thrombophlebitis complications are up to 45% of patients and pulmonary thromboembolism.

b) **Maffucci syndrome.** Familial status consisting venular malformations, venous malformations with multiple bony exostoses and enchondromas, similar to Ollier's disease.

c) **Proteus Syndrome.** Asymmetric body overgrowth, lipomatosis with both hands and / or feet, hemihypertrophy, macrocephaly or cranial abnormalities and vascular malformations, mainly venular.

d) **Sturge-Weber syndrome:** It is characterized by capillary malformations in the face with leptomeningeal vascular anomalies.

Association with high-flow vascular malformations.

a) **Parkes-Weber syndrome:** Similar findings with Klippel-Trenaunay, with venular malformations, absence of venous malformations and arteriovenous malformations. Upper extremities can affect up to 23% of cases.
Fig. 1

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Patient 1. B-mode ultrasound images with the use of color and pulsed Doppler. We observed a thickening of the left neck muscles splenius and levator scapula. They present inside vascular structures dilated discretely with venous flow. Diagnosis: venous malformation.

Fig. 2

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Patient 1. Images A, B, C: coronal reconstructions. 
C, D axial CT with civ. 
We observe an asymmetrical thickening of the left cervical musculature showing calcifications in relation to phleboliths and irregular enhancement of intravenous contrast. 
Diagnosis: Venous vascular malformation.

Fig. 3

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Fig. 4

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Patient 2.
We can see a left cervical homogeneous tumor, with calcifications, well-circumscribed with soft tissue attenuation. It is located behind the left mandibular angle in situation anteromedial to the sternocleidomastoid muscle and posterolateral carotid space.

Diagnosis: Venous malformation.

Fig. 5

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Pte 5. 24 year old woman diagnosed with cervical lymphangioma.
A) Ultrasound. We can see a well circumscribed cystic lesion with fine internal echoes.
B) Avascular at the Doppler study.
C) Cervical CT with contrast.
Right cervical tumor well defined, with low attenuation, located medial to the sternocleidomastoid muscle behind the carotid space causing minimal anterior displacement of this.

**Fig. 9**

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Patient 4. 27 year old woman. A and B) Ultrasound. C) cervical CT with contrast. We observed a polylobulated lesion affecting superficial and deep lobe of the right parotid with lower attenuation than the rest of the gland. Identified by ultrasound cystic lesions and tubular structures inside. The MRI shows hyperintense on T2 and contrast enhancement. Diagnosis of cervical lymphangioma.

Fig. 8

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Patient 3. RM. The lesion is hyperintense on T2 and hypointense on T1 with respect to the adjacent fat and it shows inhomogeneous enhancement.

Fig. 7

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Patient 3. A, B, C) Axial CT planes. D) Coronal reconstruction. 31 year old man. We observed a thickening on the left side of the soft palate, tonsillar region, pterygoid muscles, tongue base and sidewall pharyngolaryngeal to glottic region with obliteration of the parapharyngeal space, vallecula and pyriform sinus, and protrusion on light oropharyngeal and supraglottic larynx. No significant contrast enhancement observed. Associated calcifications (phleboliths) located in the masticator space, tonsillar region and anterior third of the floor of the mouth. Diagnosis: Venous vascular malformation.

Fig. 6

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A) Ultrasound: We can see a hypoechoic lesion, well-defined, with multiple septa and fine internal echoes. B) Avascular in the Doppler study.

C, D, E) Cervical MRI. Well circumscribed multilocular cyst lesion located in the floor of the mouth between the digastric muscles. It is hypointense on T1 (C) and hyperintense on T2 (D) with enhancement of the septa (E).

Fig. 10

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We reviewed the medical records of cervical vascular malformations diagnosed in our hospital from December 2002 to October 2011.

Patients were studied by ultrasound, CT and MRI.

We included a total of 25 cervical vascular malformations: 8 Type venous malformations and 17 lymphangiomasz
Conclusion

Vascular malformations are often located in the cervical region. We must be familiar with typical findings with different imaging techniques and be able to define the exact location in the cervical spaces for diagnosis at an early facilitating appropriate therapeutic strategy.
References