Learning objectives

Hemangiomas are the most frequently encountered vascular soft tissue abnormalities and also the most common tumors of infancy, affecting 2%-3% of neonates and 10%-12% of children after 1 year. They are more common in Caucasians, with a female-to-male ratio ranging from 3:1 to 5:1 and the incidence increases with prematurity, especially in those infants weighing less than 1500g [1]. They can be found in all regions of the body, but there is an anatomic predilection in the head and neck region (60%), followed by the trunk (25%) and the extremities (15%), that is not yet explained [2]. Almost 80% of all hemangiomas are single lesions, but 20% of affected infants have multiple tumors [3]. This increases the risk of having visceral tumors involving the liver, spleen, lung, brain and intestines [4]. Although the majority of hemangiomas occur sporadically, a family history of hemangioma has been reported to be present in approximately 10% of patients [5].

The diagnosis of hemangioma is normally based on patient history and clinical findings. However, in some cases, there are difficulties in distinguishing hemangiomas from other soft-tissue tumors of the head and neck and imaging is necessary to characterize the lesion and determine its anatomic extent [6].

Background

Hemangiomas are the result of a derangement in angiogenesis that allows the uncontrolled proliferation of vascular elements [7]. According to the classification system proposed by Mulliken and Glowacki [8], hemangiomas are identified as vascular tumors that exhibit an active growth phase characterized by endothelial proliferation and hyper-cellularity, followed by a period of gradual involution that occurs over several years. In contrast, vascular malformations were characterized as lesions derived from dysplastic capillaries, veins, lymphatic vessels, arteries or a combination of these.

This classification system was modified in 1997 to include kaposiform hemangioendothelioma, pyogenic granuloma, tufted angioma and hemangiopericytoma in the category of vascular tumors [9]. These lesions were previously classified as hemangiomas but were shown to have distinct histopathologic and clinical features (see Table below).

A. Vascular Tumors
Hemangiomas

Proliferating

Involuting

Kaposiform hemagioma

Tufted angioma (angioblastoma)

Angiosarcoma

Congenital hemangiopericytoma / infantile myofibromatosis

Eccrine angiomatous hamartoma

B. Vascular Malformations

High flow

Arterial malformation-aneurysm, ectasia, stenosis, etc.

Arteriovenous fistula

Arteriovenous malformation

Low flow

Capillary malformation- Port-wine stain

Telangiectasias

Venous malformation

Lymphatic malformation

Macrocytic

Microcystic

Complex combined

Slow flow: Klippel-Trenaunay syndrome (capillary lymphatic venous malformation)

Fast flow: Parkes Weber syndrome (capillary arterial venous malformation)

Hemangiomas usually appear a few weeks after birth and grow more rapidly than the infant does. The maximum size of the hemangioma is reached at age 6 to 12 months and
the clinical appearance varies with the degree of cutaneous involvement and the depth of the lesions, from an area of simple erythema to a bruise-like macule, a pale patch or a telangiectasia with a pale halo [6]. Involution of the hemangioma generally begins at about 18 months of age and is characterized by a decrease in cellularity and tumor size, formation of larger vascular channels, fibro-fatty replacement and development of a lobular architecture with septa. Involution and proliferation may be happen at the same time within different parts of a given lesion or among hemangiomas in children with multiple lesions. An early onset of involution is the only factor associated with improved outcome [10].

Although most hemangiomas resolve completely without any sequelae, several potential complications are associated with hemangiomas, most commonly in the proliferative phase of growth of the lesion. An estimated 10-20% of hemangiomas may be so called alarming hemangiomas that may threaten life or an important visceral function (Fig.1). Important complications include ulceration, bleeding, infection, obstruction (visual axis, auditory canal, airway), congestive heart failure, skeletal distortion, and aesthetic deformity [11].
Fig.: 1. A large hemangioma in the left nasal cavity that obstructs the airway of this 11-month-old boy is seen in axial MR images (a, b, c). The coronal plane is useful for understanding the extent of the lesion (d).

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE

The diagnosis of most hemangiomas is based on clinical presentation, history, and physical examination and do not require any investigation or any treatment for they will subside spontaneously. However diagnostic imaging is needed in cases of clinically atypical soft-tissue masses, questionable superficial lesions, deeper lesions with normal overlying skin, vascular like lesions presenting in later life, and cases of alarming hemangiomas. Imaging is also required to demonstrate possible visceral involvement or other associated anomalies, guide therapy and assess treatment efficacy [12]. Magnetic resonance imaging is considered the modality of choice for the evaluation of soft-tissue masses and hemangiomas are no exception to this rule [13].

The MR images of 23 patients, (ranging from 1 month to 12 years old) old with soft-tissue hemangiomas in the head and neck were retrospectively studied. The subjects included 13 females and 10 males, ranging from 2 months to 12 years of age (median 2.1 years of age). The patients were imaged consecutively at our institution between 2005 and 2009. All patients were imaged before biopsy or surgery, when performed, and the following features were evaluated: detectability of the lesion, location, size, margins, signal intensity on T2- and T1- weighted images, enhancement of contrast medium and the presence of central low-intensity dots in the lesions.

MRI examinations were performed at 1T with a head, neck or surface coil according to the size of the patient and the structure to be studied. The scan protocol included axial, coronal or sagittal spin-echo or fast spin-echo T1- and T2- weighted images. T2-weighted images with fat suppression technique were acquired in some patients. T1-weighted sequences after intravenous administration of paramagnetic contrast agent were also obtained in most patients and in at least one orthogonal plane.

Imaging findings OR Procedure details

All lesions in our patients were solitary and unilateral. Hemangiomas were located at- in descending order of frequency- the buccal space, the orbit and periocular area, the parotid and masticator spaces, the soft tissues of the anterior neck, the upper lip and the nasal cavity. However, in 6 patients, hemangiomas were not localized in a single compartment, but involved more than one spaces (Fig.1).
Fig.: 1. A 1-year-old boy with a large hemangioma that infiltrates surrounding tissues and occupies multiple contiguous spaces of the extracranial head and neck, is seen in axial T2WI (a), axial T1WI (b) and axial (c) and coronal (d) T1WI after contrast administration.

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE

MR imaging of hemangiomas in the proliferative phase demonstrated a well-defined, lobulated and heterogeneous mass with a high signal intensity relative to the muscle on T2-weighted images. On T2- weighted MR images with fat suppression technique, hemangiomas tended to show well defined borders, heterogeneity, and high contrast compared to the surrounding tissues (Fig.2).
Fig.: 2. Hemangioma in the left parotid space of a 2-month-old boy. The mass is hyperintense with lobules and septa on the T2-weighted images (a) and the lesion's contrast to the surrounding tissues is even higher on the T2-weighted images with the FS technique (b). There is marked enhancement after contrast medium administration (d).

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE

On T1-weighted images, the signal intensity of hemangiomas was intermediate between that of muscle and fat and the lesions had mostly irregular borders and showed heterogeneity (Fig.3).
Fig.: 3. A 9-month-old boy with an hemangioma of the left orbit, that is hyper-intense on a T2-weighted image (a). On the T1-weighted image the borders of the lesion are ill-defined (b). After contrast medium administration, the lesion is markedly enhanced on a T1WI (c). The lesion also involves the upper lid and extends to the periocular area as seen on coronal (d) and sagittal (e) MR images. The central low intensity dots in this case corresponded to small feeding vessels, that were clearly depicted with US imaging (f).

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE

Punctate or reticular low signal intensity areas were present in T2-weighted images in most cases and were attributed to fibrous tissue, fast flow within blood vessels, calcified or ossified foci (Fig.3,4) The presence of those low-intensity dots is an important feature in the differential diagnosis between hemangiomas and lipomas or intramuscular hematomas (Fig.5) [14].
Fig.: 4. Axial MR images in a 1-year-old girl with an hemangioma above the left eyebrow, that is hyper-intense on T2-weighted (a,b) and low-to-iso intense on T1-weighted (c) MR imaging. Paramagnetic contrast agent enhanced T1WI MR shows enhancement of the lesion (d). A low signal intensity are seen on unenhanced images is still visible after contrast administration.

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE

T1-weighted images after the intravenous administration of paramagnetic contrast agent showed a moderate to strong centripetal enhancement of all hemangiomas, with progressive accumulation of contrast material on delayed images (Fig.5,6).
Fig.: 5. An hemangioma is seen in the left masticator space of a 9-year-old girl on axial T2WI (a) and T1WI (b) MR images. The lesion shows early centripetal (c) and delayed strong (d) enhancement on T1-weighted images after contrast medium administration. There are also prominent vascular signal voids within the lesion.

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE
Fig.: 6. A 1-year-old boy with an hemangioma in the left upper lip, that is hyper-intense on T2WI (a) and hypo-intense on T1WI (b) MR. After contrast administration the lesion shows mild centripetal enhancement on T1WI axial (c) and coronal (d) MR imaging.

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE

Clinically involuting hemangiomas were observed in 3 patients and exhibited areas of high signal intensity on T1-weighted sequences and less contrast enhancement than proliferating hemangiomas (Fig.7). These findings probably corresponded to fibro-fatty replacement and the development of a lobular architecture with septa [15].
Fig.: 7. A 7-year-old boy with a right malar hemangioma in clinical regression is seen on axial images (a,b,c). T1WI MR reveals a low-to-intermediate signal of the tumor with interspersed areas of high signal intensity due to fatty infiltration (b). There is still considerable enhancement of the lesion after contrast agent administration on T1-weighted imaging (c).

References: V. Dimarelos; Radiology Department, Papageorgiou General Hospital, Thessaloniki, GREECE

Conclusion

In conclusion, an understanding of the nature and imaging characteristics of hemangioma of the head and neck soft tissues is important to enable the correct diagnosis, which is usually based on clinical presentation, history and physical examination. However, in many cases, additional investigation is required and MRI is the preferred modality because of the wealth of information provided, by using a synthesis of morphologic
features, the pattern of enhancement after contrast administration and the signal intensity of the lesion.

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**References**


