Ultrasound of Vascular Tumors

16

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Introduction

This chapter is focused on common vascular tumors that present published patterns in the literature. For academic purposes, the conditions are divided into benign and malignant.

Benign Tumors

Hemangiomas

There are several types of hemangiomas, but the most common types are infantile and congenital. These entities are reviewed in extent in the pediatric dermatologic chapter (Chap. 23). Thus, we will comment essential points in this chapter.

Infantile Hemangiomas

These are the most common soft-tissue tumor in infants [1, 2]. They present a fast growth after birth, and then, they show a slow regression period in the next 2 years. These are true endothelial proliferations that can be in the superficial or deep layers and test positive for the endothelial tissue marker glucose transporter-1 protein (GLUT-1) [1–4].

They could be a sign of dysmorphic conditions such as the PHACES syndrome (posterior fossa malformations, hemangiomas of the cervicofacial region, arterial anomalies, cardiac anomalies, eye anomalies, and sometimes sternal defects). Infantile hemangiomas can be associated with vascular anomalies of the major cerebral arteries, optic nerve hypoplasia, and abnormalities of the retinal vessels. Midline hemangiomas are more prone to the association with other abnormalities, particularly in the brain and spine. Among the complications of infantile hemangiomas are ulceration, bleeding, infection, and scarring [1–4]. Clinically, they could be categorized into superficial, deep, mixed, reticular, and abortive, with minimal growth or others, and their patterns can be focal, multifocal, segmental, or indeterminate (International Society for the Study of Vascular Anomalies Classification—ISSVA, 2018).

On ultrasound, they show as ill-defined masses in the dermis and/or hypodermis that modify their echogenicity and degree of vascularity over time (months). One of the key characteristics of the hemangiomas is their mass effect on the neighboring tissues, which allows the differential diagnosis with vascular malformations. The important role of ultrasound, besides the support

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of the diagnosis and extent, is the discrimination of the phase of the hemangioma [1, 3–13].

In the **proliferative phase**, the hemangioma is mainly hypoechoic and hypervascular. The vessels demonstrate arterial and venous flow and sometimes arteriovenous shunts. The peak systolic velocity of the arterial vessels may be variable, but it could be as high as the carotid artery velocity in these cases (Fig. 16.1) [3, 4].

In the **partial involution phase**, the hemangioma presents a mixed echogenicity with hypoechoic and hyperechoic areas and an intermediate degree of vascularity [3, 4].

In the **total involution phase**, the hemangioma shows a fully hyperechoic appearance with hypovascularity. Still, there may be some residual low-flow vessels in the region. In some cases, there are residual areas of lipodystrophy with

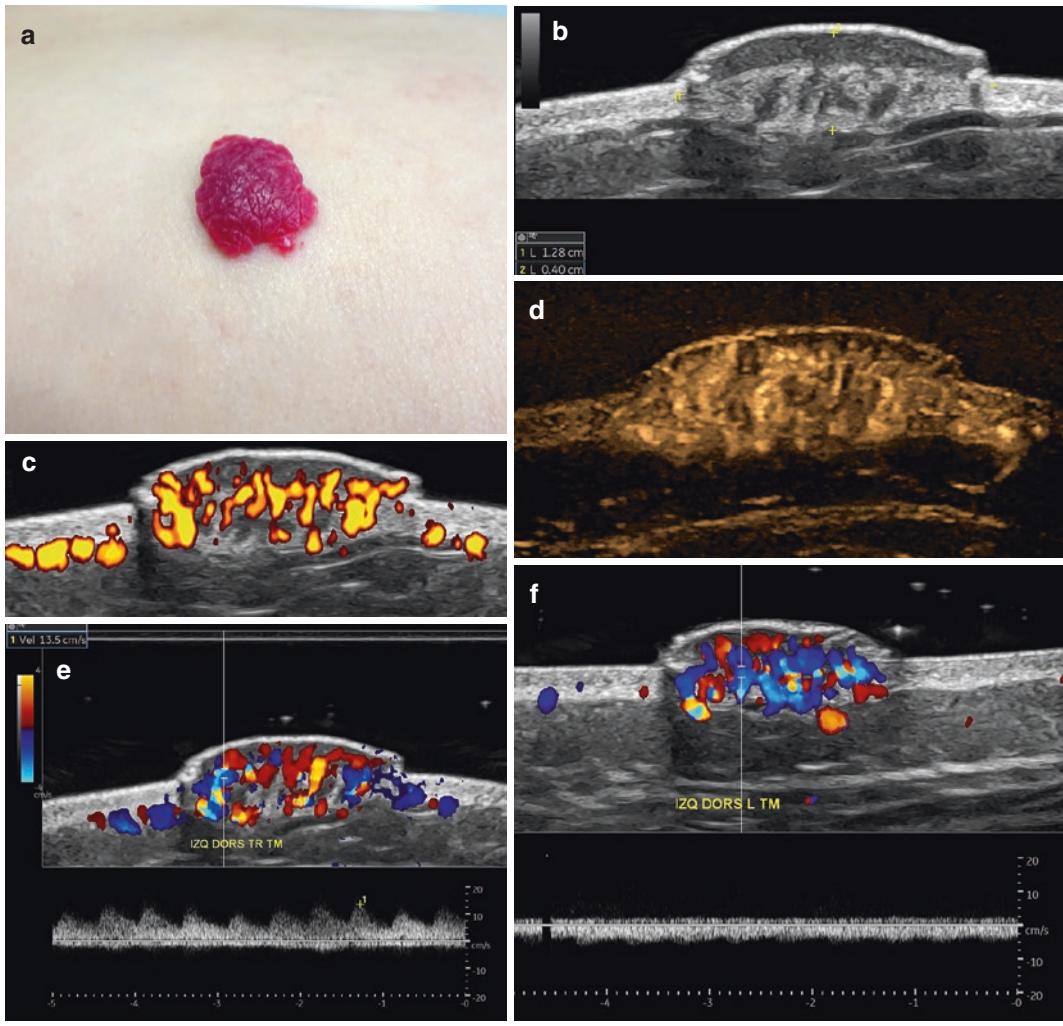

Fig. 16.1 Infantile hemangioma (proliferative phase). (**a**) Clinical image. Ultrasound images (**b**, grayscale; **c**, power Doppler; **d**, echoangio; spectral curve analysis; transverse views) demonstrate hypoechoic dermal lesion with prominent blood flow that displaces the epidermis upward. Notice that there is arterial (**e**) and venous (**f**) flow within this structure

hypertrophic or atrophic parts of the fat in the hypodermis [3, 4].

Commonly, the size, peak systolic velocity of the arterial vessels, and presence of arteriovenous shunts tend to decrease over time within the infantile hemangioma [3, 4].

Ultrasound can support the management of infantile hemangiomas, for example the decision to start, suspend, maintain, or increase a propranolol or timolol treatment [1, 3, 4, 7, 11, 12, 14–17].

Besides, it is possible to detect associated anomalies that can difficult or slow the involution of hemangioma such as the presence of direct afferent branches from medium-size arteries [18].

These anatomical details are not possible to detect with the naked-eye examination.

Ultrasound can support the diagnosis of infantile hemangioma in cases with ulceration, due to their fast growth [19].

Children presenting ≥5 cutaneous hemangiomas should be screened for ruling out hepatic hemangiomas [3, 4, 20].

Congenital Hemangiomas

These hemangiomas are fully developed at birth and test negative for the endothelial tissue marker glucose transporter-1 protein (GLUT-1) [3, 4]. According to ISSVA, they can be classified into rapidly involuting congenital hemangioma (RICH), non-involuting congenital hemangioma (NICH), or partially involuting congenital hemangioma (PICH). RICH presents a fast involution in months or the first year. NICH does not modify its size or features significantly; nevertheless, it could grow due to internal thrombosis or vascular vasodilation. PICH is in the middle with some degree of slow regression [3, 4].

On ultrasound, they can show as hypoechoic, hyperechoic, or heterogenous masses in the dermis and/or hypodermis. In contrast with infantile hemangiomas, they tend to present prominent tortuous and ingurgitated venous vessels; however, they also present arterial flow (Fig. 16.2) [3, 4, 21, 22].

RICH are commonly most hypervascular lesions compared to NICH and PICH, which tend to present a lower degree of vascularity [3, 4]. RICH may be diagnosed in the prenatal period [23].

RICH with venous lakes on ultrasound has been suggested to be more prone to develop bleeding, cardiac failure, and ulceration; however, the association was only significant for cardiac failure [22, 24].

In contrast with infantile hemangiomas, congenital hemangiomas may present calcifications [22, 24].

Angiokeratoma-Verrucous Hemangioma

These benign vascular entities are histologically similar with vascular spaces and hyperkeratosis; however, they involve different layers [25–27].

Angiokeratomas involve epidermis and dermis, and the name changes to verrucous hemangiomas when there is an additional involvement of the hypodermis [4, 25–27].

Clinically, they appear as a reddish-like papule or plaque with a warty appearance. Angiokeratomas could be single (solitary angiokeratoma) or multiple (angiokeratoma corporis diffusum) [4, 25–27].

On ultrasound, angiokeratomas present thickening and irregularities of the epidermis with decreased echogenicity of the dermis. Sometimes, there are areas with posterior acoustic shadowing artifact due to the strong hyperkeratosis (Fig. 16.3). Verrucous hemangiomas show additional ill-defined areas of hypodermal hyperechogenicity, besides the other features.

On color Doppler, these entities are hypovascular due to their predominant capillary flow [3, 4].

Glomus Tumor-Glomangioma-Glomuvenous Malformation

Glomus Tumor

It is derived from the neuromyoarterial plexus, and its main clinical symptoms are exquisite pain and sensitivity to cold. The most common loca-

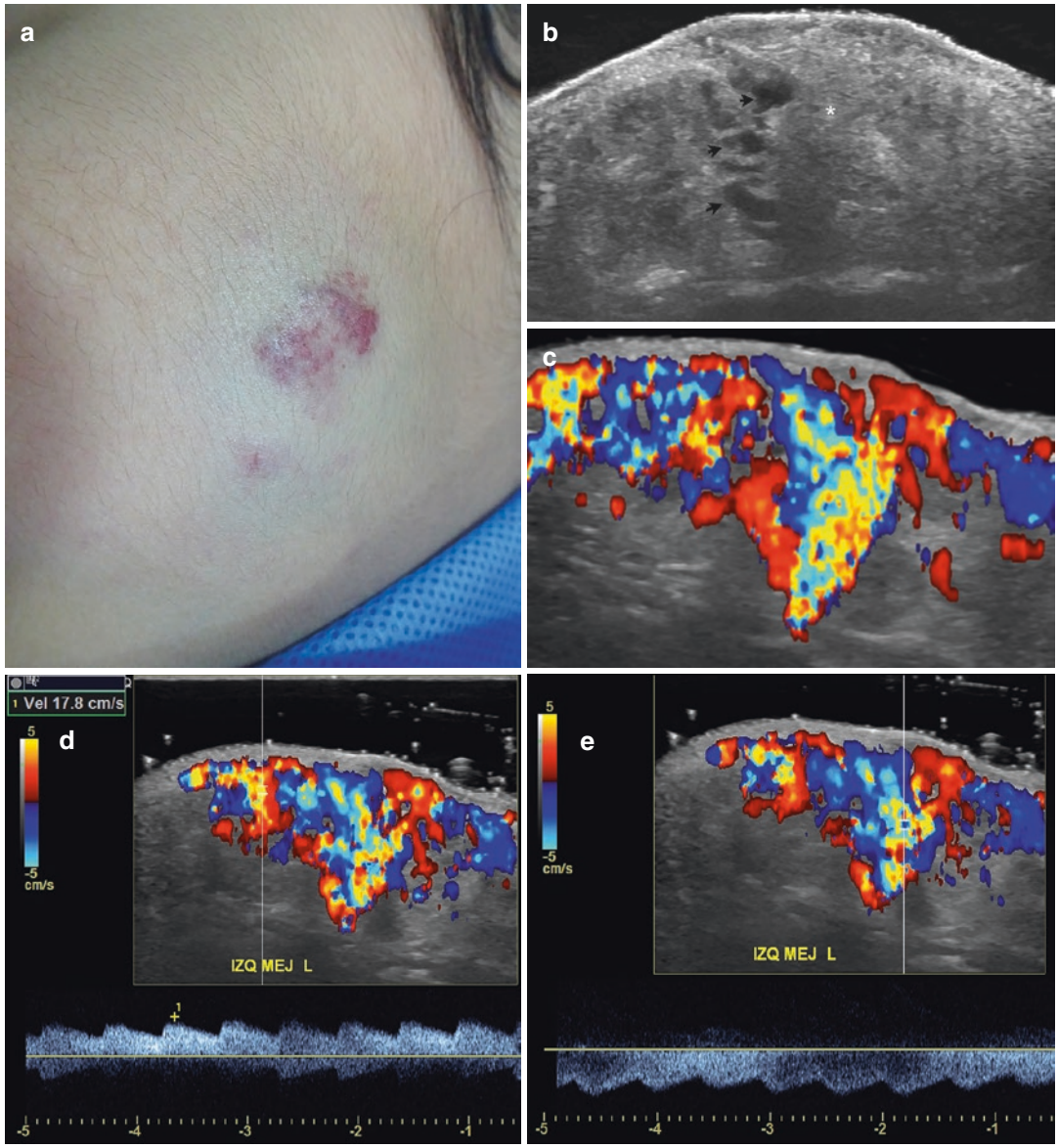

Fig. 16.2 Congenital hemangioma-type NICH. (**a**) Clinical photograph. Ultrasound images (**b**, grayscale; **c**, color Doppler; spectral curve analysis) show hyperechoic structure (*) with anechoic dilated spaces (arrows). On color Doppler, there is prominent vascularity within this structure (**c**) with arterial (**d**) and venous (**e**) flow

tion of glomus tumor is the nail bed, which has been reviewed in the corresponding nail chapter. Nevertheless, **extradigital glomus tumors** are not uncommon [3, 4, 28–33].

Usually, on ultrasound, extradigital glomus tumors present as a hypoechoic hypodermal oval-shaped dermal and/or hypodermal nodule with prominent vascularity that shows low-velocity arterial and venous vessels (Fig. 16.4). When glomus tumor is in the nail, there is additional scalloping of the underlying bony margin of the distal phalanx and sometimes an upward displacement of the nail plate [3, 4, 28–33].

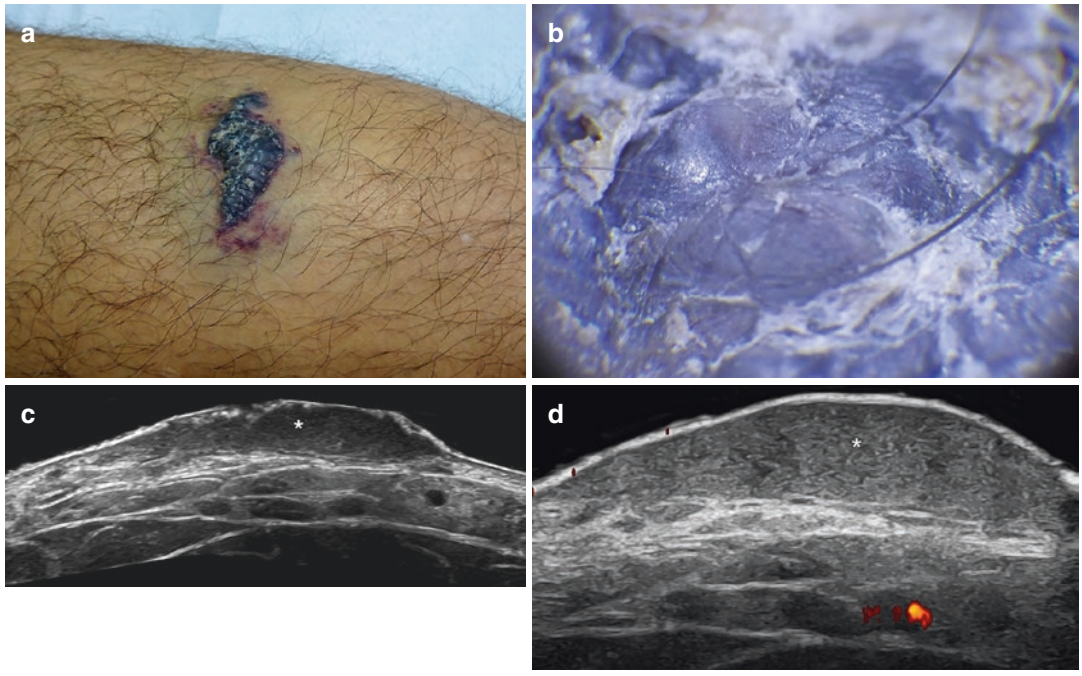

Fig. 16.3 Angiokeratoma. (**a**) Clinical image. (**b**) Dermoscopy. Ultrasound images (**c**, grayscale; **d**, color Doppler; transverse view; left calf) show epidermal thickening and upward displacement with hypoechoic thickening of the dermis (*). Notice some irregularities in the epidermis better displayed in **c**. The underlying hypodermis is unremarkable. No hypervascularity is detected within the lesion (**d**)

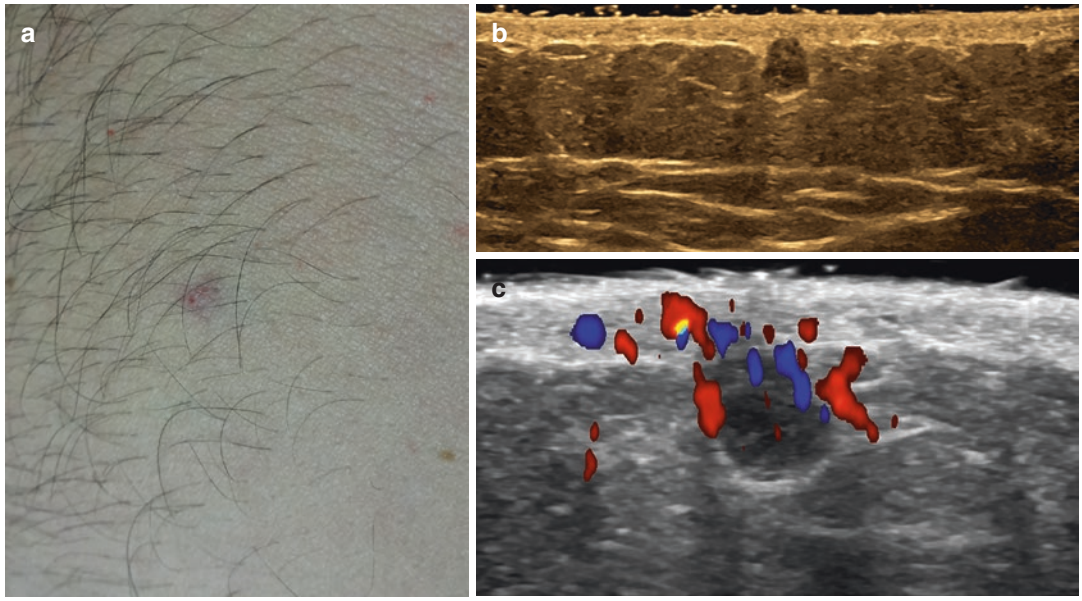

Fig. 16.4 Glomus tumor (extradigital) (**a**) Clinical photograph. The patient presented exquisite pain in the lesional area (left flank). Ultrasound images (**b**, grayscale with color filter, and **c**, color Doppler; transverse views) demonstrate hypoechoic nodule located in the upper hypodermis that protrudes into the dermis. On color Doppler, there is prominent vascularity within the lesion

Glomangiomas

These are variants of glomus tumors, and on histology they contain more venous vessels and are less likely to present a capsule in comparison with glomus tumors. They could be located outside the ungual region and are more frequent in the young population [4].Some of these cases are multiple and painless or with slight tenderness. On ultrasound, they tend to show a nodular or pseudonodular hypoechoic dermal and/or hypodermal appearance, and their degree of vascularity is variable and goes from hypovascular to hypervascular [4].

Glomuvenous Malformations

These are hamartomatous lesions conformed of glomus cells in the vascular smooth muscle [34].

On ultrasound, they show as delimited clusters of superficial dermal and/or hypodermal pseudonodular structures of mixed echogenicity, with hypoechoic and heterogeneous areas and anechoic, pseudocystic tubular and lacunar zones. On color Doppler, there is low-flow arterial and venous vessels; therefore, the name glomus venous malformation is incorrect (Fig. 16.5) [34].

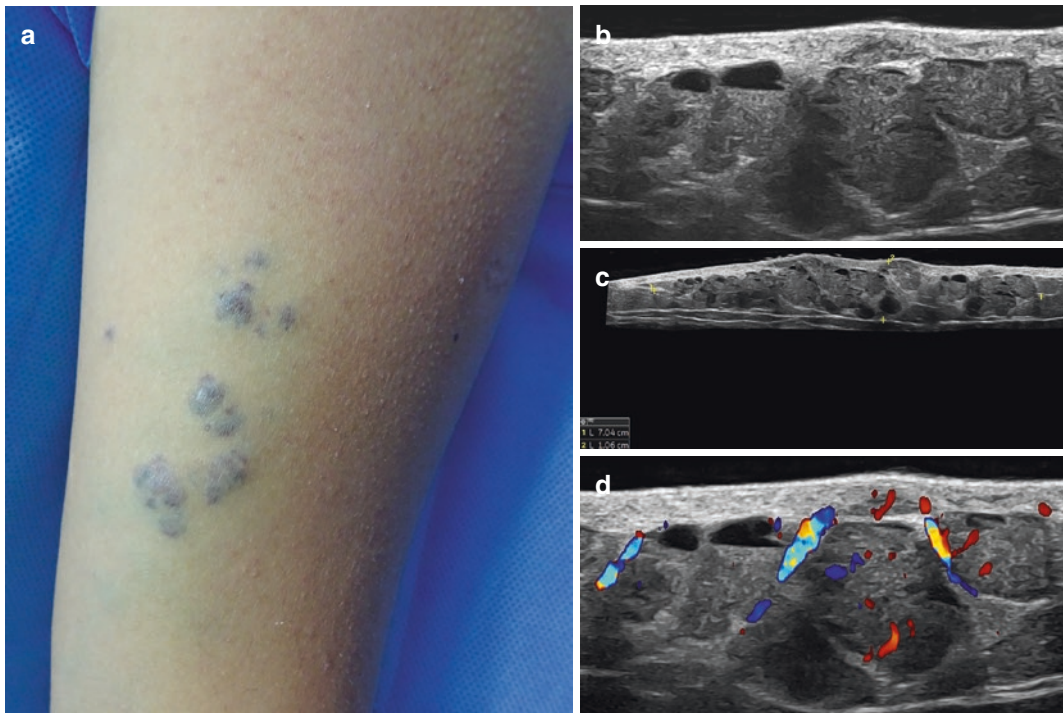

Fig. 16.5 Glomuvenous malformation. (**a**) Clinical image. Ultrasound images (**b** and **c**, grayscale; **c**, panoramic view; **d**, color Doppler; longitudinal views; left arm) show multiple lacunar anechoic and hypoechoic structures located in the dermis and hypodermis, which predominate in the hypodermis. On color Doppler, there are some low-velocity arterial vessels in the periphery. At the spectral curve analysis of the lesions, there were some areas with slow venous flow and others without detectable blood flow, which corresponded to the detection threshold of the flow which is usually 2 cm/s

Pyogenic Granuloma

Also called telangiectatic granuloma and lobular capillary hemangioma, this vascular entity is supposedly reactive and more frequent in children and pregnant women. It appears as a fast-growth erythematous swelling or polypoid lesion that commonly bleeds and tends to ulcerate [4, 35].

On ultrasound, it shows as an epidermal, dermal, and/or hypodermal, well-defined and hypoechoic polypoid, or ill-defined tissue, commonly exophytic. This lesion usually displaces the epidermis upward. On color Doppler, it presents high vascularity with intermediate- or low-velocity arterial and venous vessels (Fig. 16.6) [4, 36].

Angiolymphoid Hyperplasia with Eosinophilia

Angiolymphoid hyperplasia with eosinophilia (ALHE), also called epithelioid hemangioma, is an uncommon benign vasoproliferative disorder. Clinically, it presents solitary or multiple papules or nodules or plaque-like regions, commonly the head and neck region [37]. Frequent locations are

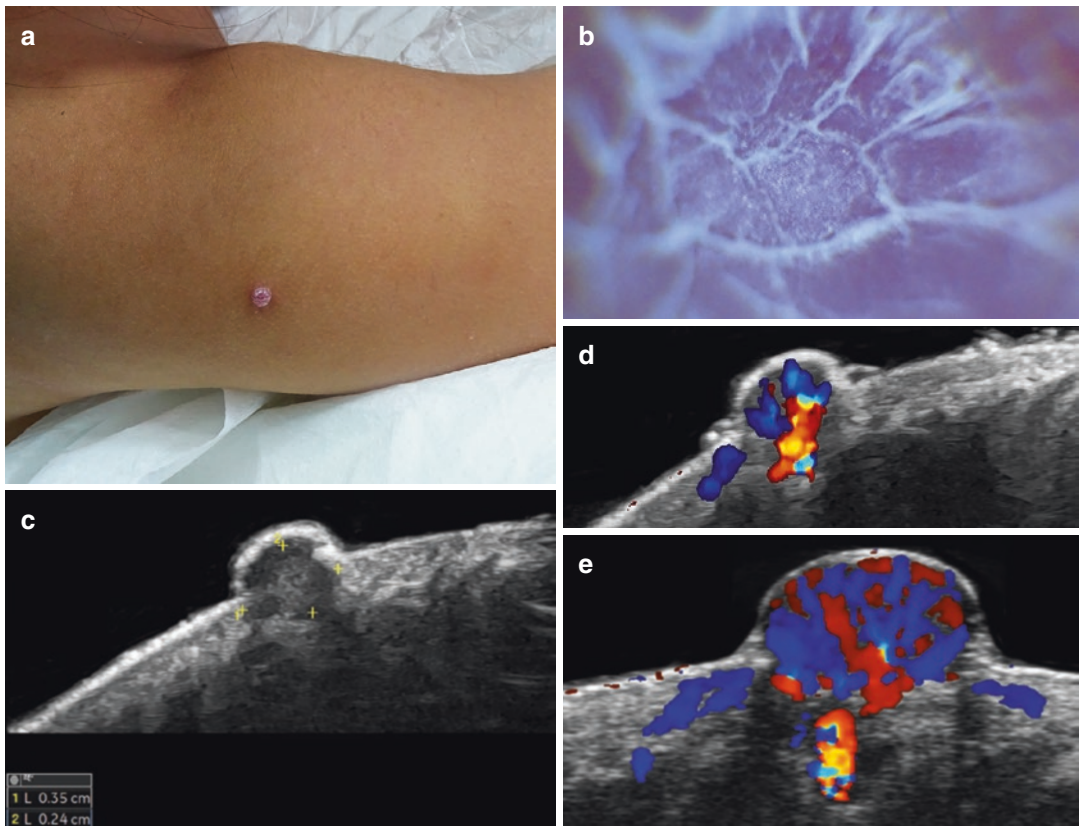

Fig. 16.6 Pyogenic granuloma (telangiectatic granuloma). (**a**) Clinical photograph. (**b**) Dermoscopy. Ultrasound images (**c**, grayscale; color Doppler; (**d**) at 24 MHz and (**e**) at 70 MHz) demonstrate hypoechoic exophytic polypoid dermal structure that displaces the epidermis upward. On color Doppler, there is prominent vascularity within the lesion

the auricular and periauricular region as well as the scalp [38].

On ultrasound, it appears as multiple hypoechoic dermal and upper hypodermal areas or plaques, commonly with a pseudonodular and exophytic appearance. Undulation of the epidermis and decreased echogenicity and increased thickness of the dermis can also be detected [38]. Some authors have reported a wooly pattern [39] and lesions with a hypoechoic rim and an echogenic central part [40].

On color Doppler, there is prominent arterial and venous vessels within the lesions, commonly with connecting vascularity (Fig. 16.7).

Kaposiform Hemangioendothelioma

It is a locally aggressive vascular neoplasm that mainly occurs in children. It originates on the skin but affects deeper tissues by infiltrative growth; nevertheless, these are not known to produce distant metastases [41, 42].

Clinically, it shows as single or multiple reddish nodules or a swelling that rapidly grows and infiltrates the soft tissues, and in most cases is associated with consumptive coagulopathy (Kasabach-Merritt syndrome) and lymphangiomatosis [4, 41, 42].

On ultrasound, they show as ill-defined heterogeneous dermal and/or hypodermal areas that may also involve the muscular layer.

On color Doppler ultrasound, they present a variable degree of vascularity that can go from hypovascular to hypervascular (Fig. 16.8) [4, 41, 42].

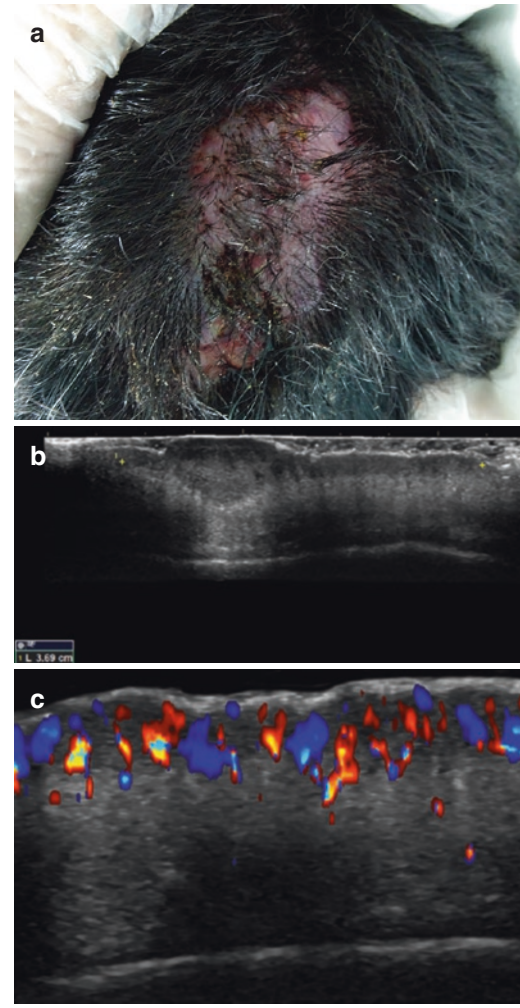

Fig. 16.7 Angiolymphoid hyperplasia with eosinophilia. (**a**) Clinical photograph. Ultrasound images (**b**, grayscale; **c**, color Doppler; longitudinal views) present hypoechoic dermal thickening (plaque-like) with some pseudonodular area that involves the upper hypodermis (**b**). On color Doppler, there is prominent vascularity within this region

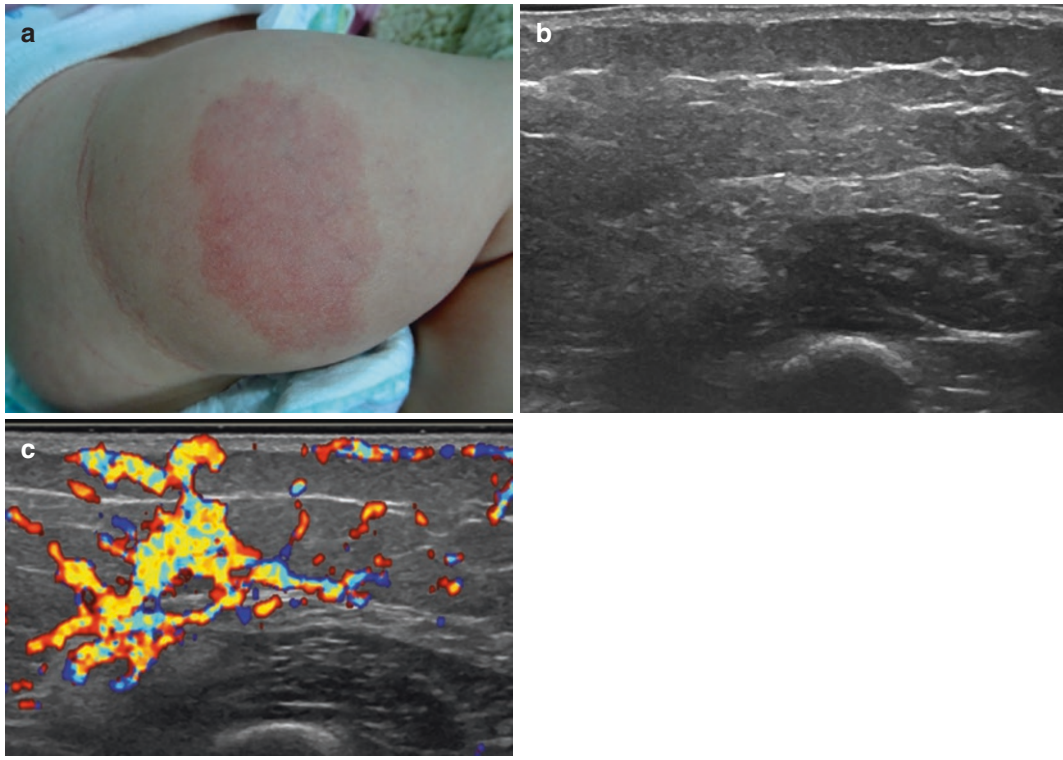

Fig. 16.8 Kaposiform hemangioendothelioma. (**a**) Clinical image. Ultrasound images (**b**, grayscale; **c**, color Doppler; transverse views; anterolateral aspect of the right thigh) show ill-defined hyperechoic hypodermal region. On color Doppler, there is prominent vascularity in the region

Malignant

Angiosarcoma

Angiosarcomas are rare, aggressive soft-tissue sarcomas originating from endothelial cells of lymphatic or vascular origin and associated with a poor prognosis.

Clinically, they show as a fast-growth erythematous-violaceous swelling, nodule, or plaque, sometimes with ulceration.

On ultrasound, they show as ill-defined and occasionally lobulated hypoechoic dermal and/or hypodermal tissue or masses with prominent and chaotic vascularity. It can involve deeper tissues such as the fascial or the muscular layers (Fig. 16.9) [4, 43, 44].

Kaposi's Sarcoma

This malignant angiomatous condition is commonly seen in immunosuppressive patients. It can involve the skin, lymph nodes, and visceral organs. It is associated with the human herpesvirus-8 (HHV-8), and on histology it presents a proliferation of spindle cells (also called the KS tumor cells), inflammatory cells, and neoangiogenesis [4, 45, 46].

Four clinical variants have been reported:

(a) Classic KS, also called Mediterranean KS that affects middle-aged men of Mediterranean and Jewish descent.
(b) Iatrogenic KS, in iatrogenically immunosuppressed patients (e.g., posttransplant).

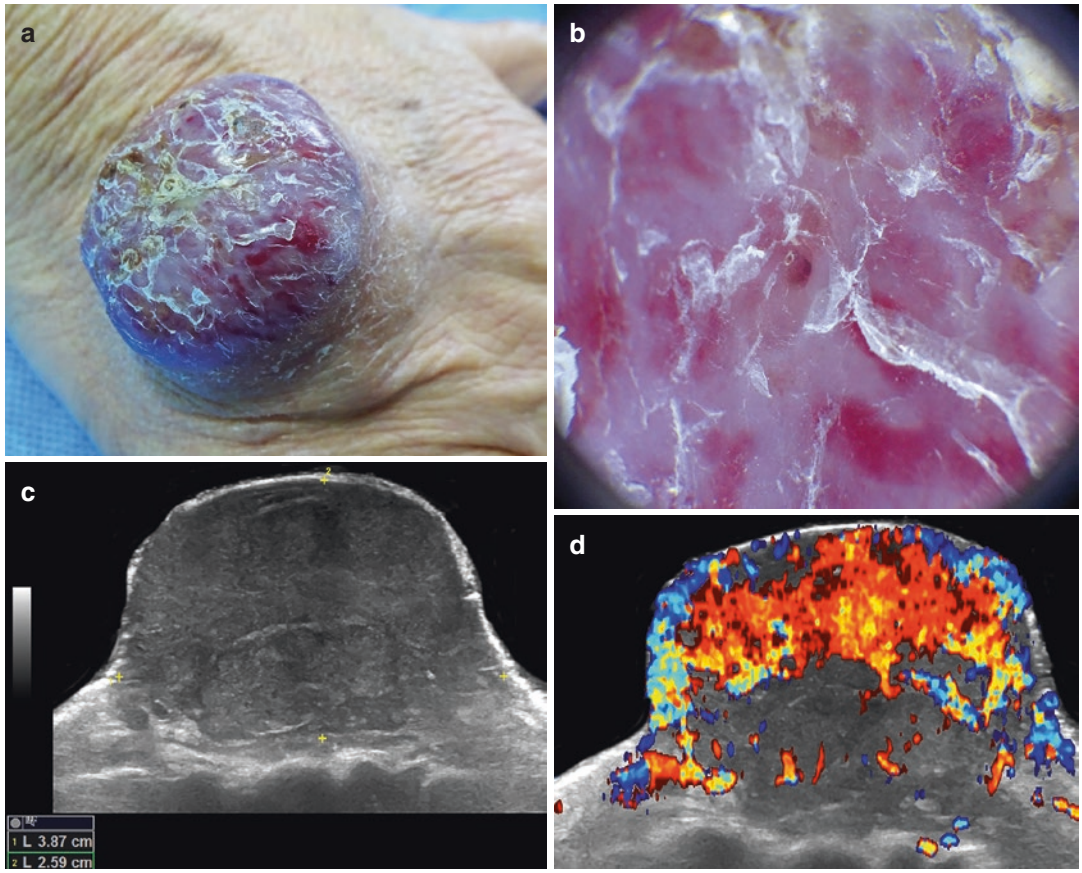

Fig. 16.9 Angiosarcoma. (**a**) Clinical image. (**b**) Dermoscopy. (**c**) and (**d**) Ultrasound images (**c**) grayscale; (**d**) color Doppler; transverse views; dorsum of the right hand) demonstrate hypoechoic dermal and hypodermal mass (between markers) that shows irregular borders at the bottom. On color Doppler, there is high vascularity within the mass

(c) African endemic KS.
(d) AIDS related, also called epidemic AIDS-KS.

Clinically, KS is characterized by erythematous and/or violaceous macules, papules, plaques, or nodules with edema. These are more common in the lower limbs and sometimes show ulceration [4, 45, 46].

On ultrasound, they can show as hypoechoic dermal and/or hypodermal pseudonodules that may displace the epidermis upward. Some cases present as ill-defined hypoechoic dermal and/or hypodermal tissue (interstitial involvement). In cases with ulceration, a disruption of the epidermis can be detected.

On color Doppler, the vascularity is variable and can go from hypovascular to hypervascular with slow-flow vessels (Fig. 16.10) [4, 47, 48].

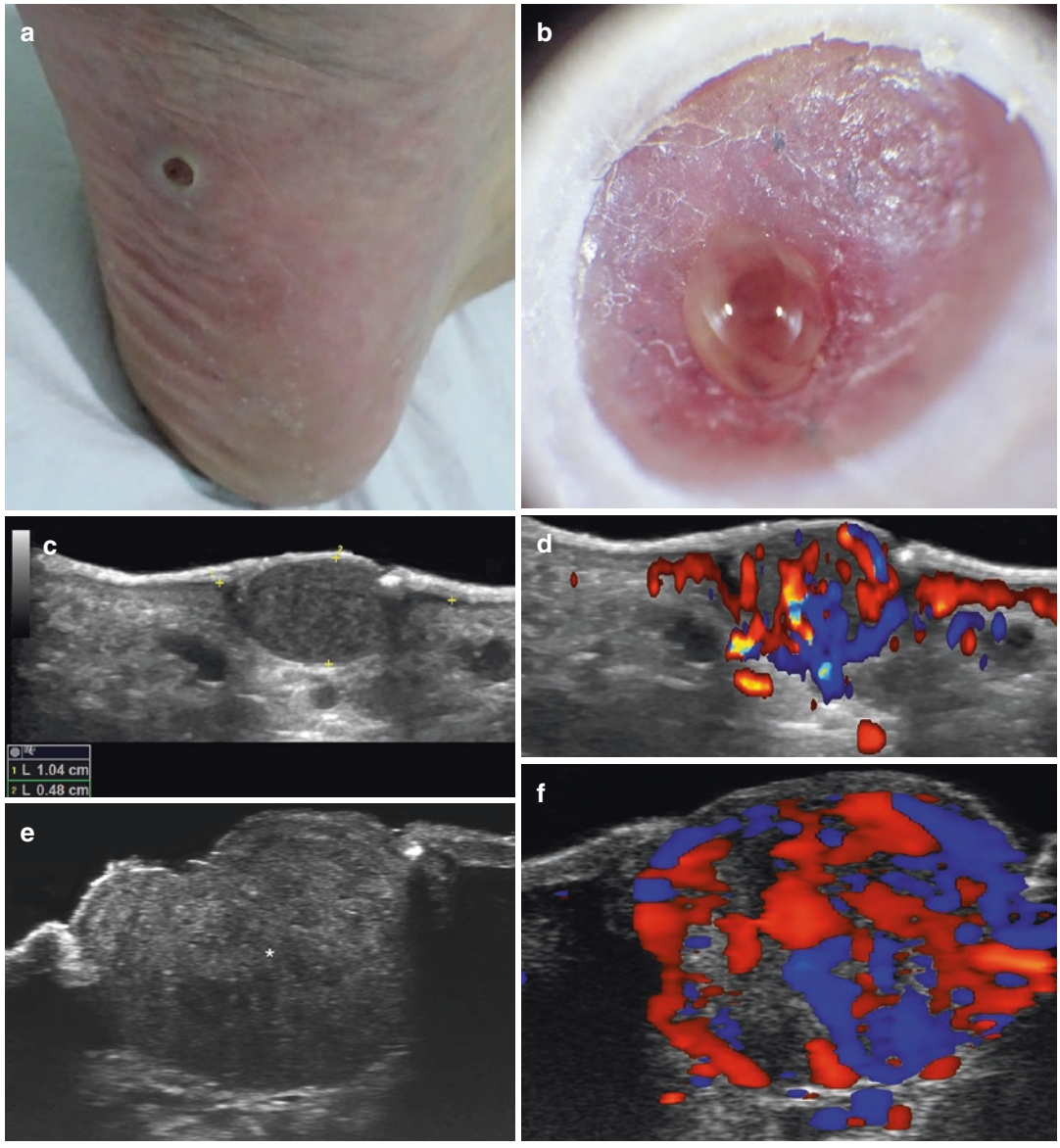

Fig. 16.10 Kaposi's sarcoma. (**a**) Clinical image. (**b**) Dermoscopy. Ultrasound images (**c** and **e**, grayscale ultrasound; **c** at 18 MHz and **e** at 70 MHz; color Doppler ultrasound; **d** at 18 MHz and **e** at 70 MHz; right plantar region) present hypoechoic dermal nodular structure (between markers in **c** and * in **e**) with epidermal disruption. On color Doppler (**d**, **f**), there is high vascularity within the lesion

Conclusion

Ultrasound can support the diagnosis and assessment of the extent and degree of vascularity in benign and malignant vascular cutaneous tumors.

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