Cutaneous Vascular Tumors

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Contents

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8.1 Introduction

There are many types of vascular tumors that can affect the skin. The most common vascular tumors are hemangiomas of infancy that were reviewed in the previous chapter. Additionally, congenital hemangiomas [rapidly involuting congenital hemangioma (RICH) and non-involuting congenital hemangioma (NICH)] are discussed in Chap. 3 (Congenital Conditions). In this chapter we will discuss other types of cutaneous vascular entities that can be referred for a sonographic examination.

8.2 Vascular Tumors

8.2.1 Benign

8.2.1.1 Angiokeratoma

Angiokeratoma is conformed by dilated vessels and hyperkeratosis. Current classification distinguishes between widespread forms (angiokeratoma corporis diffusum), which are usually associated with an inborn error of metabolism, and localized forms, that include solitary angiokeratoma, Fordyce's angiokeratoma, angiokeratoma circumscriptum naeviforme, and angiokeratoma of Mibelli [1]. Thus, there are several variants that predominantly affect the trunk and limbs with reddish papulae that present a wart-like appearance clinically. Histologically, these lesions present a similar appearance in all variants showing multiple dilated capillaries in the papillary dermis that can extend to the epidermis, with acanthosis and hyperkeratosis also being detected. On sonography, angiokeratomas presents as a hypoechoic band or region that affects the dermis with thickening of the epidermis. The keratotic component can generate a tiny posterior acoustic shadowing artifact, and frequently, they present hypovascularity on color Doppler imaging because of their slow flow capillary component (Fig. 8.1).

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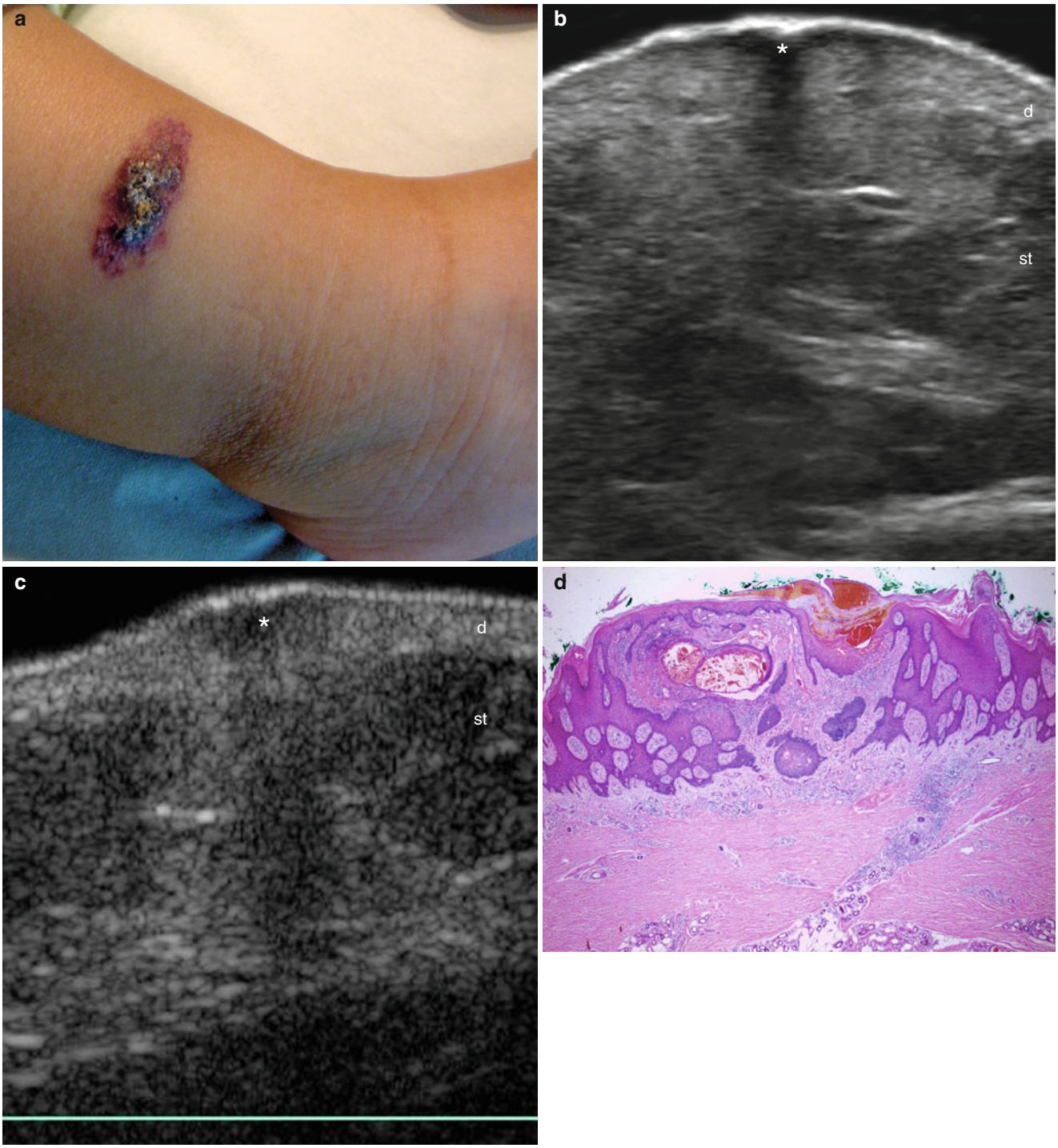


Fig. 8.1 (**a**–**d**) Angiokeratoma. (**a**) Clinical reddish lesion in the anterior aspect of the right leg. (**b**) Grey scale ultrasound (transverse view) shows hypoechoic region (*) in the dermis that generates a tiny posterior acoustic shadowing artifact. Notice the thickening of the epidermal layer. (**c**) Color Doppler ultrasound image (transverse view) demonstrates lack of detectable vascularity within the lesion. (**d**) Histology (HE × 40 zoom): proliferation of dermal vessels with epidermal acanthosis and hyperkeratosis

8.2.1.2 Verrucous Hemangioma

Verrucous hemangioma (VH) is a confusing name because it shows similar clinical and histological characteristics to angiokeratoma, with the exception that VH also involves the subcutaneous tissue. Clinically, it affects children with warty dark blue or reddish nodules predominantly on the limbs [2]. On sonography, VHs present as hypoechoic dermal bands with thickening of the epidermis and hyperechogenicity of

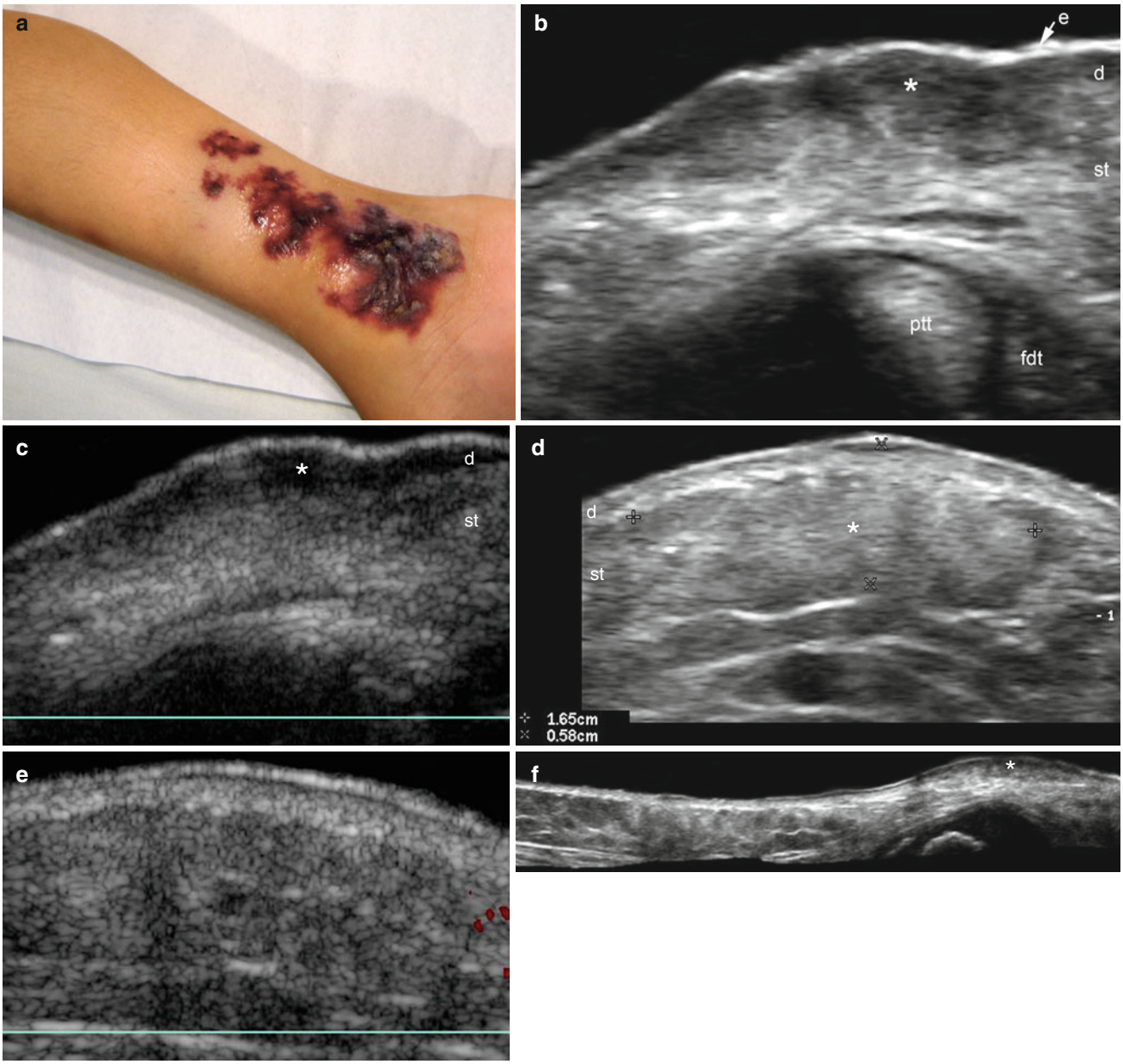


Fig. 8.2 (**a**–**f**) Verrucous Hemangioma. (**a**) Clinical reddish lesion in the medial aspect of the right leg and ankle. (**b**) Grey scale ultrasound image (transverse view, ankle region) shows the lesional area (*) with hypoechogenicity and thickening of the dermis and upper subcutaneous tissue. Also notice the thickening of the epidermis (arrow). (**c**) Color Doppler ultrasound (transverse view, ankle region) demonstrates lack of detectable vascularity within the tumor (*). (**d**) Grey scale ultrasound (transverse view, leg region) shows hyperechogenicity of the subcutaneous tissue. (**e**) Color Doppler ultrasound (transverse view, leg region) demonstrates hypovacularity of the subcutaneous tissue in the lesional area. (**f**) Grey scale (extended field of view in longitudinal axis) shows the heterogeneous echostructure in the dermis (mostly hypoechoic) and subcutaneous tissue (mostly hyperechoic) in the medial aspect of the leg and ankle. *Abbreviations*: *e* epidermis, *d* dermis, *st* subcutaneous tissue, *ptt* posterior tibial tendon, *fdt* flexor digitorum tendon

the subcutaneous tissue. The lesions are usually hypovascular on color Doppler ultrasound that can be related to the slow capillary flow component (Fig. 8.2).

8.2.1.3 Kaposiform Hemangioendothelioma

Kaposiform hemangioendothelioma (KH) are tumors that are common in children and young adults often manifesting later than infantile hemangioma. KH can be associated with lymphangiomatosis and has no known association with Kaposi sarcoma related to human immunodeficiency virus infection, and demonstrates aggressive local behavior with invasion but not distant metastasis. KH shows predilection for the limbs, head, and neck and is characterized by its common association with Kasabach–Merrit syndrome (i.e., consumptive coagulopathy)

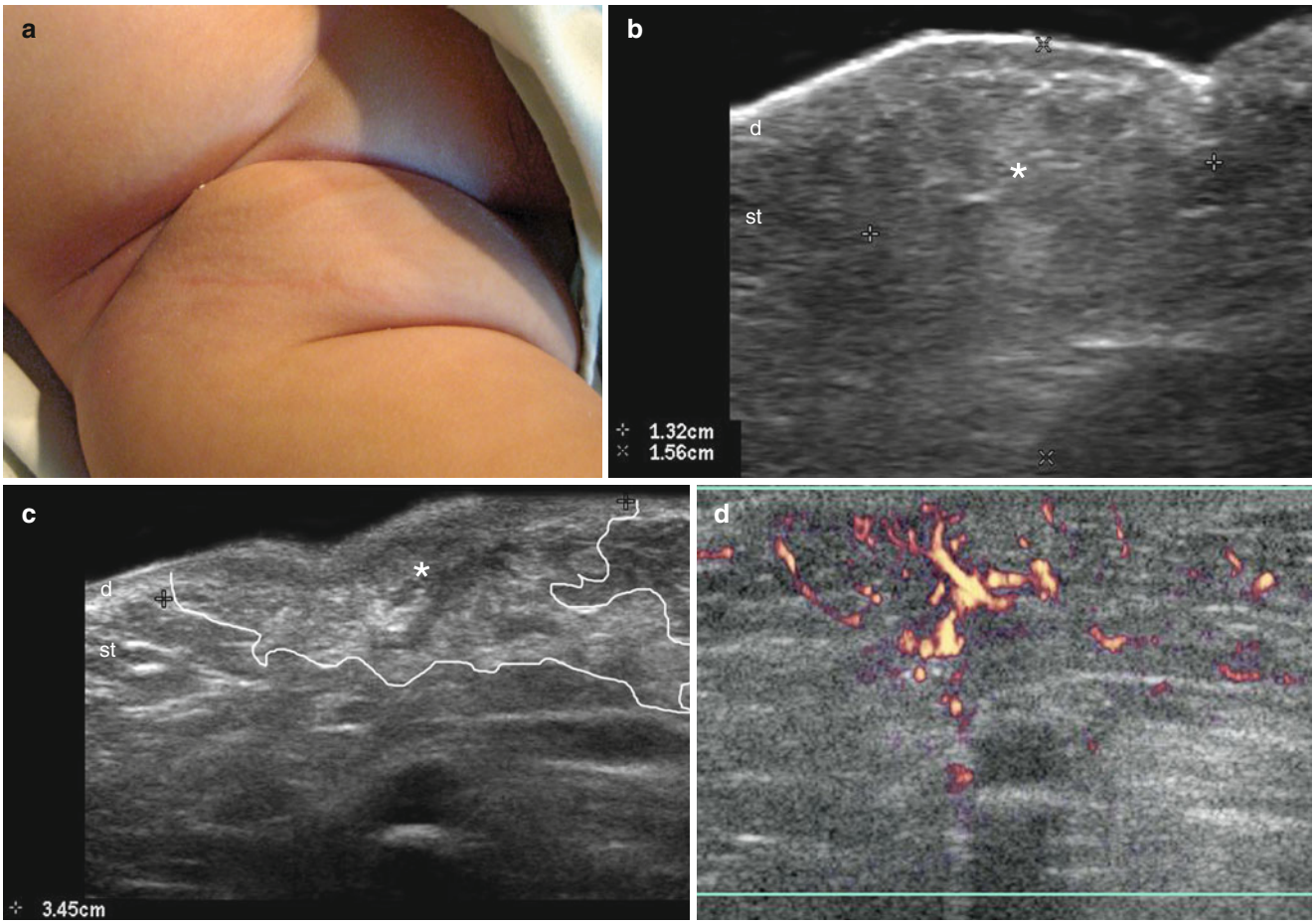


Fig. 8.3 (**a**–**d**) Kaposiform hemangioendothelioma. (**a**) Clinical swelling in the right groin. (**b**) Grey scale ultrasound (transverse view) shows ill-defined hyperechoic and heterogeneous region (*, measuring 1.32 × 1.56 cm, between markers; transverse and depth axis respectively) that affects mostly the subcutaneous tissue and to a lesser extent the dermal layer. (**c**) Grey scale ultrasound (longitudinal view) demonstrates 3.45 cm long ill-defined hyperechoic and heterogeneous lesion (*, outlined, between markers). (**d**) Color Doppler ultrasound (transverse view) shows irregular hypervascularity within the lesional area. *Abbreviations*: *d* dermis, *st* subcutaneous tissue

that can cause a high mortality rate among patients. Clinically, KH shows as reddish nodules or swelling that rapidly grows and infiltrates the soft tissues. Histologically, the tumors are lobular, infiltrative, and composed of spindle cells, congested capillaries, slit-like vascular spaces, and occasional pale epithelial cells. Thrombotic phenomena can be detected in parts of the lesion. Endothelial cells test positive for CD31, CD34, and FLI1 but negative for GLUT1 and LeY in inmunohistochemistry [3–5]. On sonography, KH tend to show an ill-defined heterogeneous echogenicity that commonly affects the dermis and subcutaneous tissue and easily involves deeper structures such as muscle. On color Doppler ultrasound, tumor vascularity may present a variable grading from hyper- to hypovascular (Fig. 8.3).

8.2.1.4 Glomus Tumor-Glomangioma

Glomus tumor or glomangioma are conformed by rare neoplasms arising from the subcutaneous glomus apparatus (i.e., neuromyoarterial component). They account for 1–5 % of soft-tissue tumors of the upper extremity, occurring in most cases in the nail bed. When located on the nail, the typical clinical presentation includes reddish spots, paroxysmal pain, and hypersensitivity. Nevertheless, occasionally painless subungual glomus tumors can be detected. Recently, the tumors comprised of glomus cells have been categorized into two major subtypes: the glomus tumor and the glomangioma. Glomangiomas differ clinically from glomus tumors in that they present in children and young adults, are usually asymptomatic, do not have a predilection for the subungual region, and often are multifocal. They can vary in color from pink to blue and often become darker with age; they can be plaque-like or nodular. Multiple glomangiomas are rare and comprise about 10 % of all glomus tumors. Because glomangiomas are not neoplastic and resemble venous malformations, it has been suggested that they should more precisely be called lineal glomuvenous malformations.

Histopathologic features of glomangiomas contain more-dilated venous channels than do glomus tumors and that is why they resemble venous malformations. Unlike venous malformations, they demonstrate single-to-multiple rows of surrounding cuboidal glomus cells. These cells stain positively for vimentin and α-smooth-muscle actin but are negative for desmin, von Willibrand factor, and S-100. Another

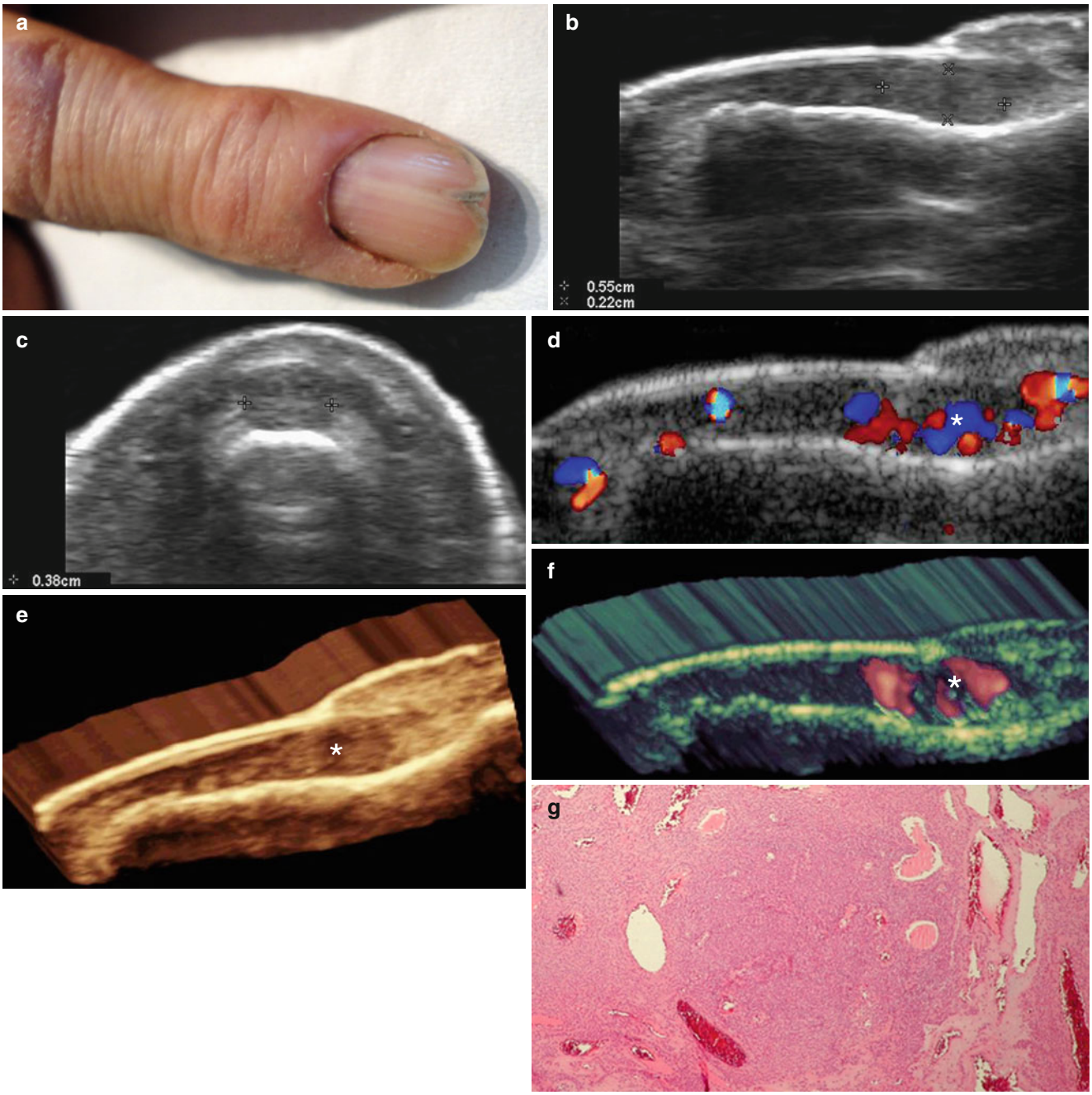


Fig. 8.4 (**a**–**g**) Glomus tumor of the nail. (**a**) Clinical dystrophy of the nail of the left index finger. (**b**) Grey scale ultrasound (longitudinal view) shows 0.55 × 0.22 cm (long and depth axis, respectively), well-defined, oval -shaped, hypoechoic nodule in the proximal nail bed. Notice the slight upward displacement of the nail plates and the remodeling of the bony margin of the distal phalanx. (**c**) Grey scale ultrasound (transverse view) also demonstrates the 0.38 cm hypoechoic nodule in the proximal nail bed. (**d**) Color Doppler ultrasound (longitudinal view) shows increased blood flow within the tumor (*). (**e**) (grey scale) and (**f**) (power angio) are different 3 D reconstructions of the tumor (*) (5–8 s sweep). (**g**). Histology (HE × 40 zoom): intradermal vascular proliferation surrounded by cuboidal cells that conform a solid pattern

difference is that histologically glomangiomas are less likely to have a capsule than glomus tumors. Glomus tumors lye in the deep reticular dermis where there are dilated vascular spaces that are lined by cuboidal cells with round nuclei and eosinophilc cytoplasm.

On ultrasound, these entities tend to present as well-defined hypoechoic nodules slightly heterogeneous and occasionally with anechoic areas. In the nail bed they tend to be centrally located (without involvement of the periungual region) and usually remodel the bony margin of the distal phalanx underneath the tumor. On color Doppler ultrasound, glomus tumors often show hypervascularity with low flow arterial and venous vessels, although there are some hypovascular subtypes (Fig. 8.4). In contrast, glomangiomas may show a variable morphology that goes from being well-defined, hypervascular, and nodular to ill-defined and

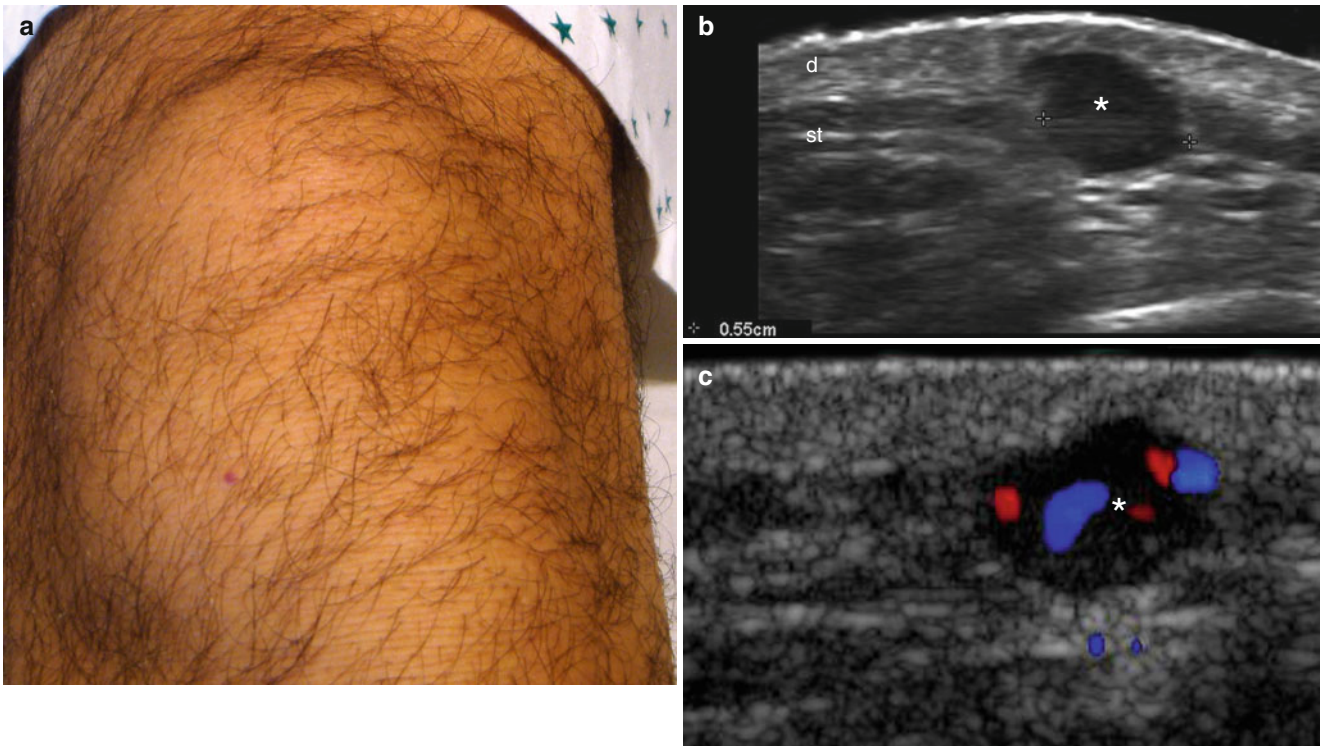


Fig. 8.5 (**a**–**c**) Glomangioma. (**a**) Unremarkable clinical view of the anterior aspect of the knee. Nevertheless, the patient presented with tenderness in correlation with the sonographic findings. (**b**) Grey scale ultrasound (transverse view) demonstrates 0.55 cm well-defined hypoechoic nodule in the dermis and upper subcutaneous tissue. (**c**) Color Doppler ultrasound (longitudinal view) demonstrates increased vascularity within the nodule (*)

hypovascular depending on the amount and size of the vessels. These benign entities are usually composed of slow flow and capillary structures (Fig. 8.5). Thus, glomangiomas are commonly located in the dermis and subcutaneous tissue and may present clinically as multiple linear bluish and serpiginous lesions that demonstrate a nodular appearance on ultrasound as previously described [6–12].

8.2.1.5 Pyogenic Granuloma

Pyogenic granuloma (PG), also called telangiectatic granuloma and lobular capillary hemangioma, is a lesion that is a reactive and lobular proliferation of capillary vessels that commonly affects the skin of the head, neck and limbs, the nail, and the oral mucosa. PG appears more frequently in children, young adults, and pregnant women and shows a fast initial growth. Clinically, it appears as erythematous or bluish swelling or polypoid lesions that easily ulcerates and bleeds. Histologically, these entities are composed of lobules with multiple capillary vessels within a loose collagenous. Mitosis is frequent and the lesion may also show inflammatory cells and fibrosis during late stages. On sonography, PG presents as hypoechoic pseudonodules or structures usually located in the dermis and subcutaneous tissue. On color Doppler imaging, prominent vascularity is commonly detected with slow flow arterial and venous vessels [13, 14] (Fig. 8.6).

8.2.1.6 Epithelioid Hemangioma

Epithelioid hemangioma, also called angiolymphoid hyperplasia with eosinophilia, atypical pyogenic granuloma, pseudopyogenic granuloma, inflammatory angiomatous nodule, papular angioplasia, and intravenous atypical vascular proliferation, is a benign, reactive vaso-proliferative disease, presenting with painless, reddish or purple, single or multiple nodules in the dermal and subcutaneous tissues of the head and neck, particularly around the ear. Blood eosinophilia is present in up to 15 % of the cases and a male predominance has been noted in selected Asian studies and presents most commonly in patients 20–50 years old, with a mean onset of 30–33 years [15].

Histology shows nodules or masses conformed by multiple vascular spaces lined with rounded endothelial cells with prominent eosinophilic cytoplasm. Blood vessels lined with cuboidal endothelial cells and numerous eosinophils are also detected.

On sonography, hypoechoic and slightly heterogeneous nodules or pseudonodules are located in the dermis and occasionally in the subcutaneous tissue. Color Doppler imaging demonstrates increased vascularity within the nodules usually with slow flow arterial vessels. When this entity affects the ear, it may cause a thinning of the auricular cartilage (Fig. 8.7).

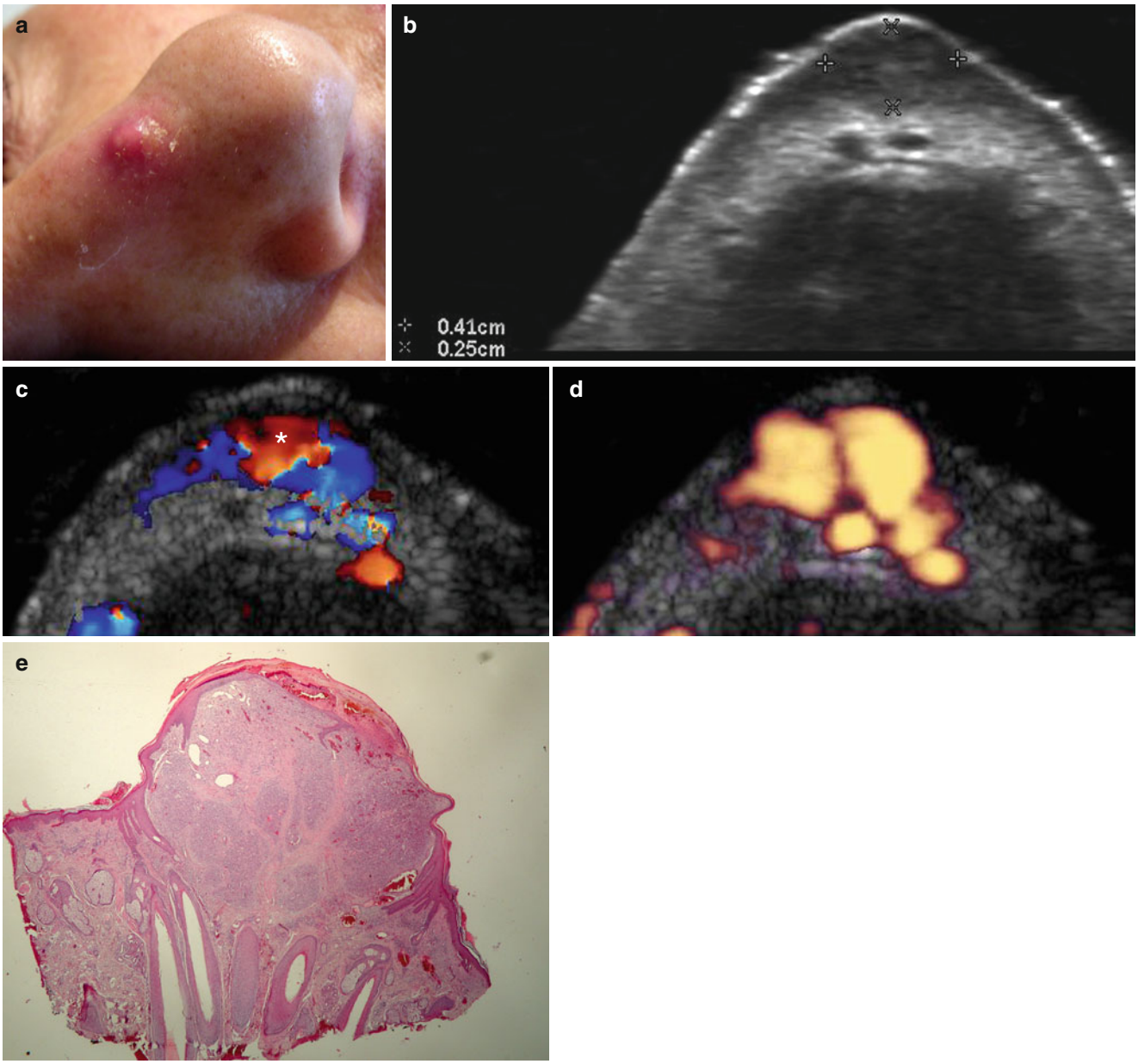


Fig. 8.6 (**a**–**e**) Pyogenic Granuloma. (**a**) Clinical image shows erythematous swelling in the dorsum of the nose. (**b**) Grey scale ultrasound (transverse view): 0.41 × 0.25 cm hypoechoic nodule that affects dermis and upper subcutaneous tissue. (**c**) Color Doppler ultrasound and (**d**) Power Angio (transverse view) demonstrate increased vascularity within the nodule (*). (**e**) Histology (HE 20 × zoom): proliferation of capillary intradermal vessels with a lobulated pattern and covered by excoriated epidermis

8.2.1.7 Lymphangioma

Lymphangiomas are congenital malformations of the lymphatic system that can involve the skin. They account for 4 % of all vascular tumors, but comprise 25 % of benign vascular growths in children. They are hamartomatous in nature and can be grouped into cutaneous lymphangioma circumscriptum (CLC), cavernous lymphangiomas, and cystic hygromas. CLC appears localized to the dermis, although frequently extends deeper and laterally. Clinically, lymphangioma circumscriptum shows clusters of flesh-colored to translucent clear, pink, red, or black papules that measure 1 to 4 mm, overlying hyperpigmented and erythematous indurated plaques [16, 17]. On sonography, they may present as mixed echogenicity nodules with a variable degree of anechoic and hypoechoic areas in the cutaneous layers. On color Doppler imaging, lymphangiomas are predominantly hypovascular. However, hypervascularity can be found in part of the tumor because of stromal components (Fig. 8.8).

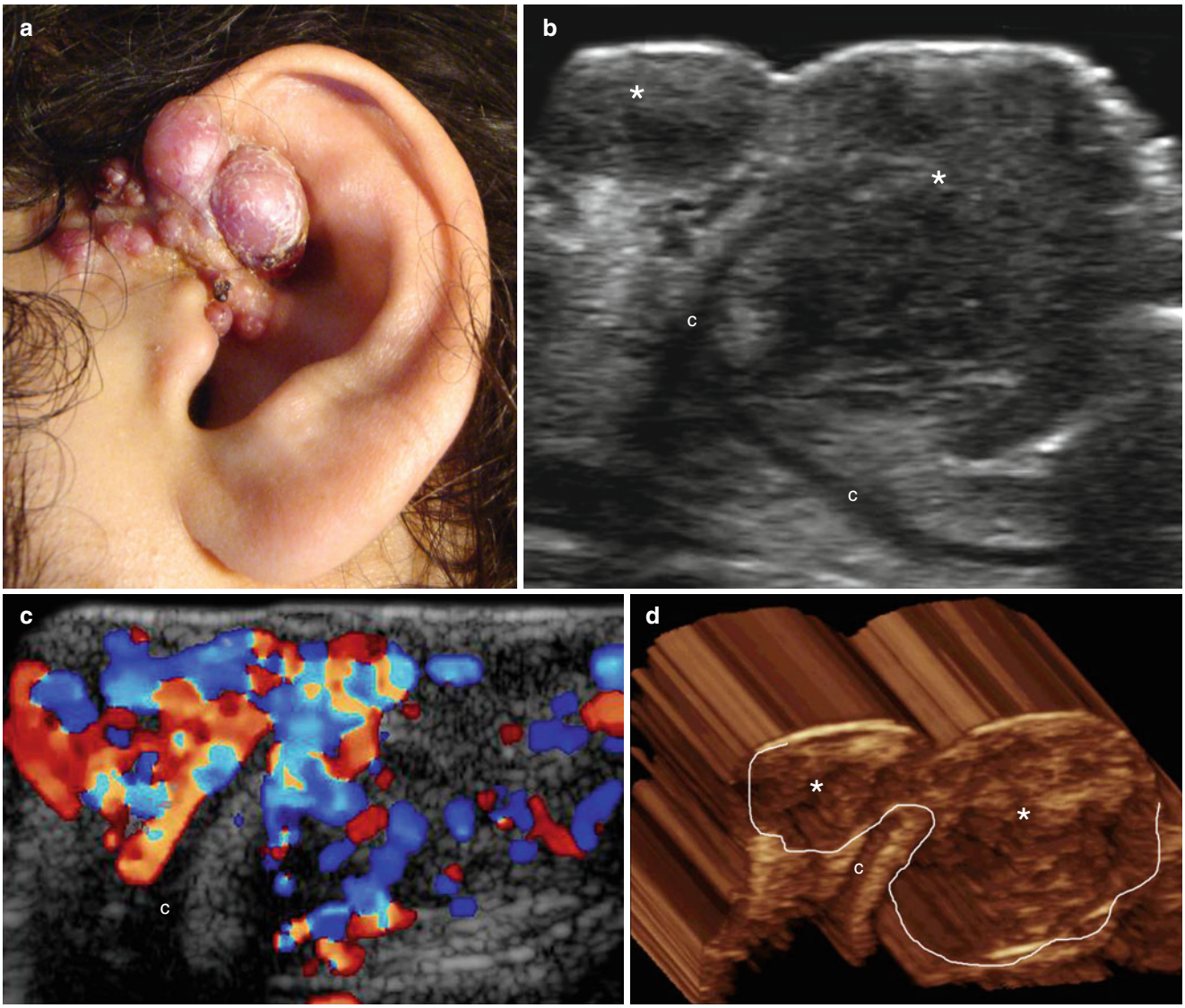


Fig. 8.7 (**a**–**d**) Epithelioid hemangioma (angiolymphoid hyperplasia with eosinophilia). (**a**) Clinical lesion shows multiple erythematous bumps that affect the left ear pinna and pre-auricular region. (**b**). Grey scale ultrasound (transverse view) demonstrates hypoechoic and confluent nodules and pseudonodules (*) that involve the cutaneous layers of the helix. Notice that there is thinning of the ear cartilage (*c*) that conserves its hypoechogenicity. (**c**) Color Doppler ultrasound (transverse view) shows prominent vascularity within the nodules. (**d**) 3D reconstruction of the lesion (*, and outlined; transverse axis, 5–8 s sweep). *Abbreviations*: *c* cartilage

8.2.1.8 Cutaneous Angiomyxoma

Cutaneous angiomyxoma (CA), also called superficial angiomyxoma, is a condition that affects all ages, with a peak incidence in the third and fourth decades and arises in the trunk, lower limb, head, and neck regions. Approximately one third of CAs recur locally, but there have been no metastases reported. Patients with multiple lesions may have the Carney complex (i.e., autosomal dominant condition conformed by myxomas of the heart and skin, hyperpigmentation of the skin [lentiginosis], and endocrine overactivity). CA affects females approximately seven times more often as it does males.

Clinically, CA presents as cutaneous papules, nodules, swellings or polypoid lesions. Tumors can show variable size and may measure 10 cm or more in diameter, demonstrate local infiltration of the surrounding tissues, and recur in approximately 50 % of cases [18].

Histology shows extensive myxoid stroma, numerous small blood vessels, varying cellularity, acellular mucin pools, stellate or bipolar fibroblastic cells, muciphages, a sparse and mixed inflammatory cell infiltrate with notable neutrophils, and occasional plumper cells with eosinophilic cytoplasm. Cytologic atypia is reported as mild. In approximately 20 % of cases, the primary lesion or its recurrence contained epithelial structures, including epidermoid cysts, thin strands of squamous epithelium, and small buds of basaloid cells. Immunohistochemically, tumor cells are negative for S-100 protein, smooth muscle actin, and pankeratin [19, 20].

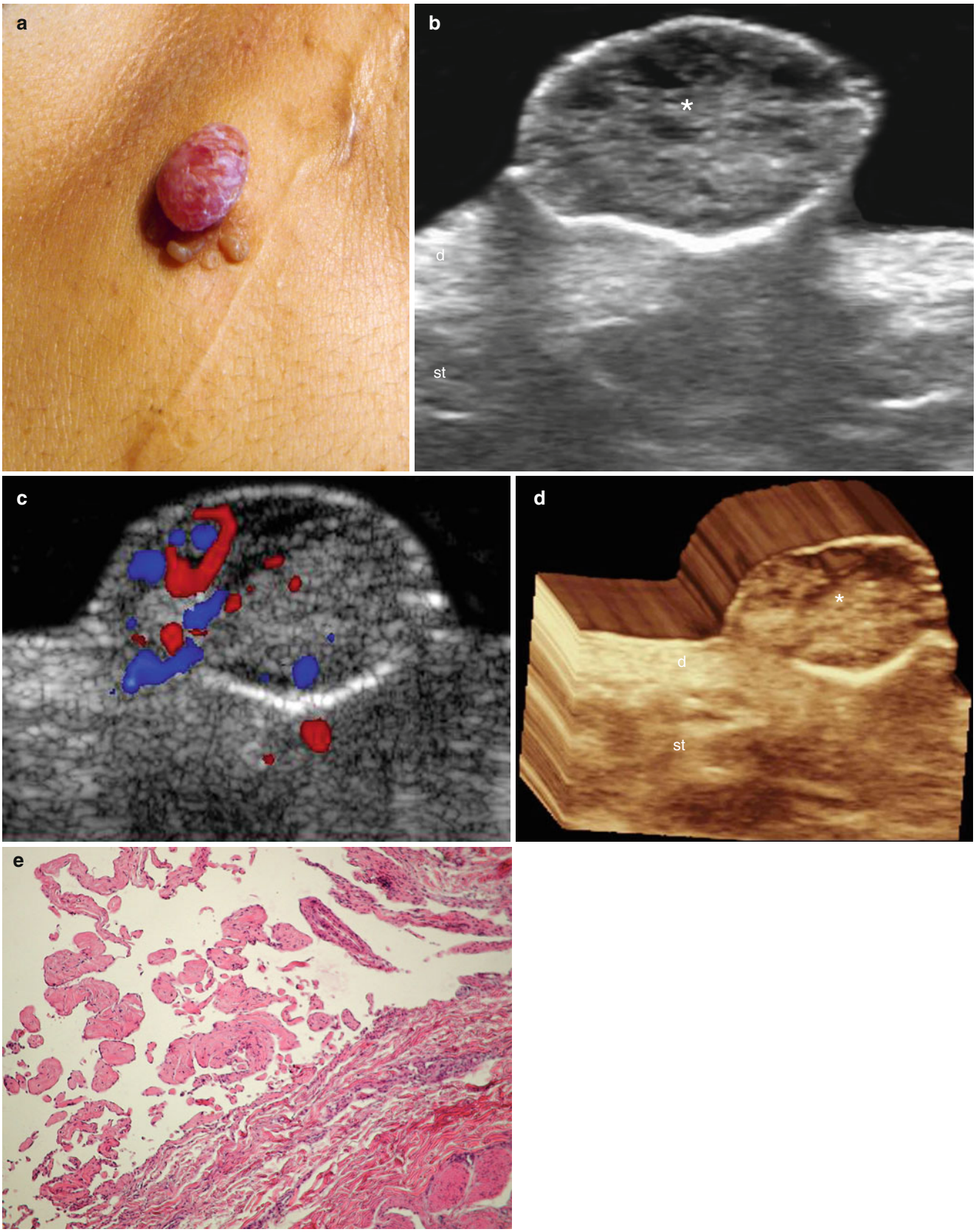


Fig. 8.8 (**a**–**e**) Recurrent lymphangioma circumscriptum. (**a**) Clinical image shows erythematous and translucent cutaneous lumps and papules that recurred 2 years after surgery. (**b**) Grey scale ultrasound (transverse view in the large nodule) shows a well-defined mixed echogenicity oval-shaped nodule with anechoic and hypoechoic areas that emerges from the dermis and protrudes into the epidermal region. (**c**) Color Doppler (transverse view) demonstrates increased vascularity in part of the lesion (mostly *left* border of the image) and hypovascularity in the remaining areas. (**d**) The lesion in 3D (5–8 s sweep). (**e**) Histology (HE 100 × zoom): vascular spaces without content and covered by a monolayer of flat endothelial cells. *Abbreviations: d* dermis, *st* subcutaneous tissue

On sonography, there are ill-defined areas with round or oval-shaped hypoechoic nodules and some anechoic pseudocystic structures that usually involve the subcutaneous tissue with additional increased echogenicity of the vicinity. On color Doppler imaging, these areas are mostly hypovascular or show slow flow vessels (Fig. 8.9).

8.2.2 Malignant

8.2.2.1 Cutaneous Angiosarcoma

Cutaneous angiosarcoma are rare soft-tissue sarcomas of the endothelial cells most commonly affecting the head and neck and particularly the scalp, but can also involve other corporal regions such as the limbs. A subset of patients presents with multifocal disease and/or positive regional nodes,

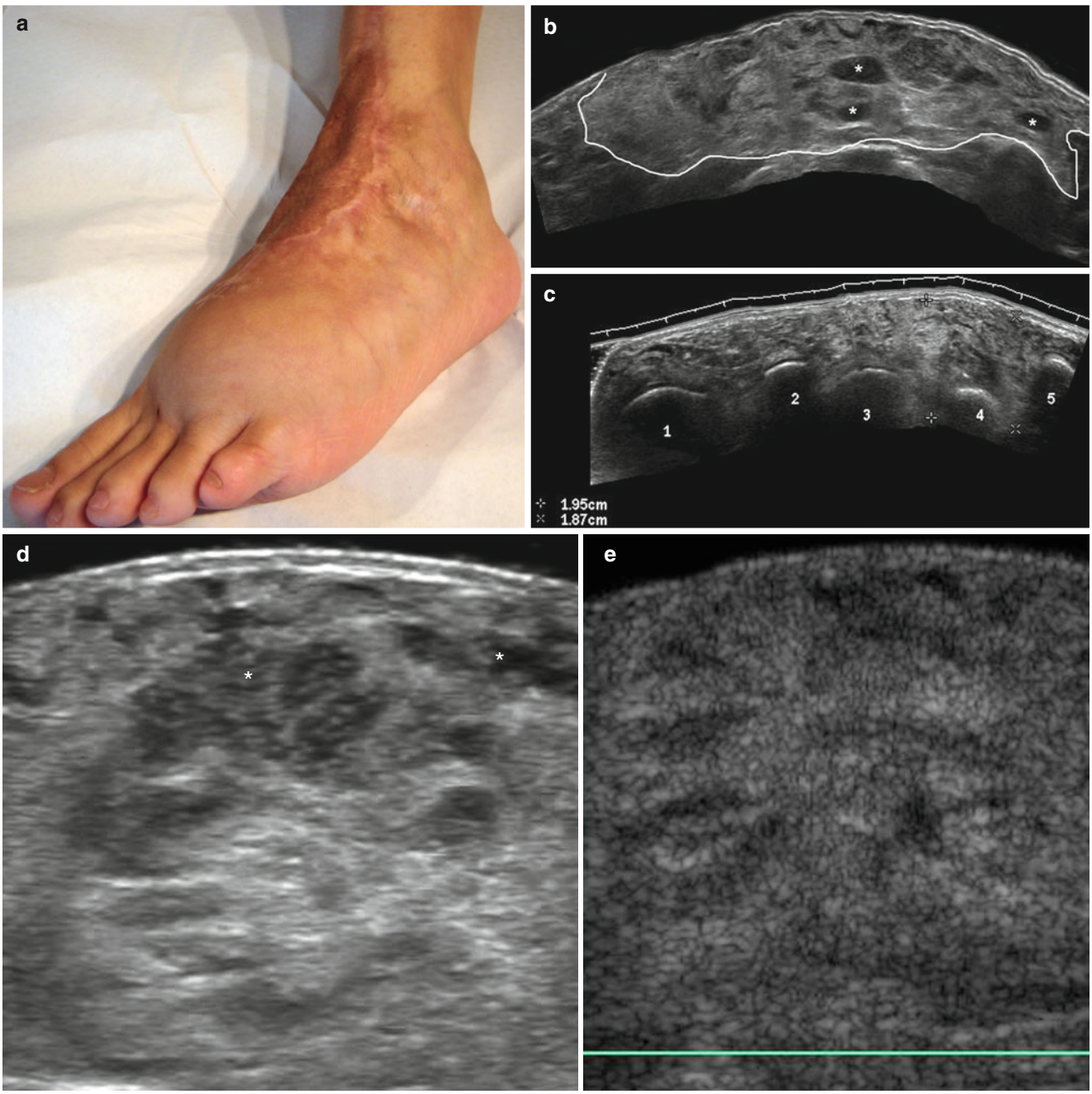


Fig. 8.9 (**a**–**g**) Recurrent angiomyxoma. (**a**) Clinical image shows swelling in the dorsum of the left foot. The patient was operated on 1 year previously (scar) and the biopsy was positive for angiomyxoma. (**b**–**d**) Grey scale ultrasound (transverse axis; b, ankle level; c, foot-metatarsal level (numbers); d, zoom dorsum of the foot level) demonstrates ill-defined heterogeneous structure with some oval-shaped hypoechoic areas (*) that mostly involve the subcutaneous tissue. (**e**) Color Doppler ultrasound (transverse view, dorsum of the foot) shows lack of detectable vascularity within the lesional area. (**f**) The lesion in 3D (5–8 s sweep). (**g**) Histology (HE × 100 zoom): proliferation of capillary vessels within a myxoid stroma

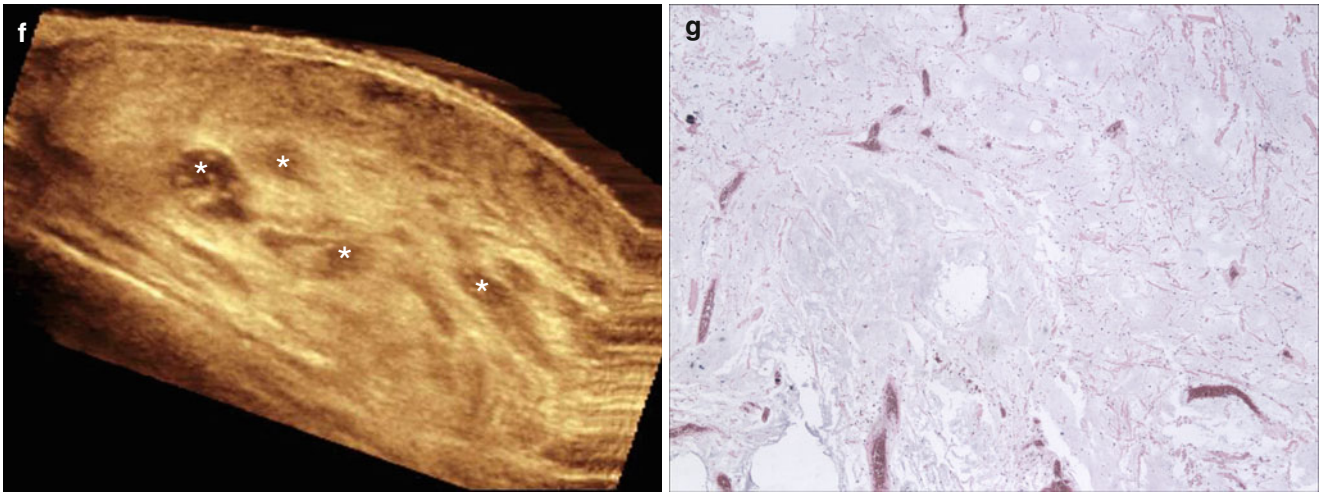


Fig. 8.9 (continued)

and the probability of metastasis is relatively high [21]. This condition is thought to be a collection of hemangiosarcoma, lymphangiosarcoma, tumors that cannot be classified as being of vascular and lymphatic origin, or mixed tumor of both. Thus, based on immunohistochemistry, cutaneous angiosarcoma can be divided into vascular, mixed, and lymphatic types [22].

Clinically, lesions show as rapidly growing, erythematous or purple, bruise-like swellings, sometimes with ulceration. Histology shows extensive infiltration with pleomorphic, multilayered endothelial cells that show a high rate of mitosis, embedded in a collagen matrix [21].

On sonography, cutaneous angiosarcomas present as hypoechoic and sometimes lobulated solid masses that affect the cutaneous layers and commonly deeper structures. Hyperechoic fibrous septa can be detected within the mass and on color Doppler imaging, increased vascularity, with asymmetric and irregular disposition of the vessels is usually found within the lesion (Fig. 8.10)

8.2.2.2 Kaposi's Sarcoma

Kaposi's sarcoma (KS) is a malignancy of endothelial skin cells with multifocal localization on the skin, lymph nodes, and visceral organs. KS comprises four clinical variants:

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

All variants are associated with human herpesvirus-8 (HHV-8), and they display very similar histopathological features, with the proliferation of spindle cells (considered as the KS tumor cells) associated with inflammation and neoangiogenesis.

Clinically, KS is characterized by violaceous and/or reddish macules and papules, which over the course of months or years tend to merge into plaques and nodules (in some cases ulcerated), commonly associated with edema, particularly evident in the lower limbs. However, definitive diagnosis is based on histopathological evidence of spindle cell and the presence of HHV-8 latency associated nuclear antigen, in spindle cells, and vascular or lymphatic endothelial cells.

The clinical progression of classic KS is generally slow and not very aggressive, although cases with rapidly growing lesions and with signs of local invasiveness, can be observed as well as forms that fail to respond to physical or systemic treatment. By contrast, the natural history of AIDS-KS, which can affect mucous membranes, lymph nodes, the gastrointestinal tract, and the lungs, is more aggressive, particularly in untreated HIV-infected individuals.

Preliminary results have suggested that small cutaneous KS lesions–in both classic KS and AIDS-KS patients–display similar B-mode ultrasound patterns conformed by hypoechoic and/or heterogeneous lesions, localized in the epidermis and dermis and sometimes affecting the subcutaneous tissue. On color Doppler imaging, vascularity is variable although lesional blood flow has been reported to be more prominent in cases with AIDS-KS [23–26] (Fig. 8.11).

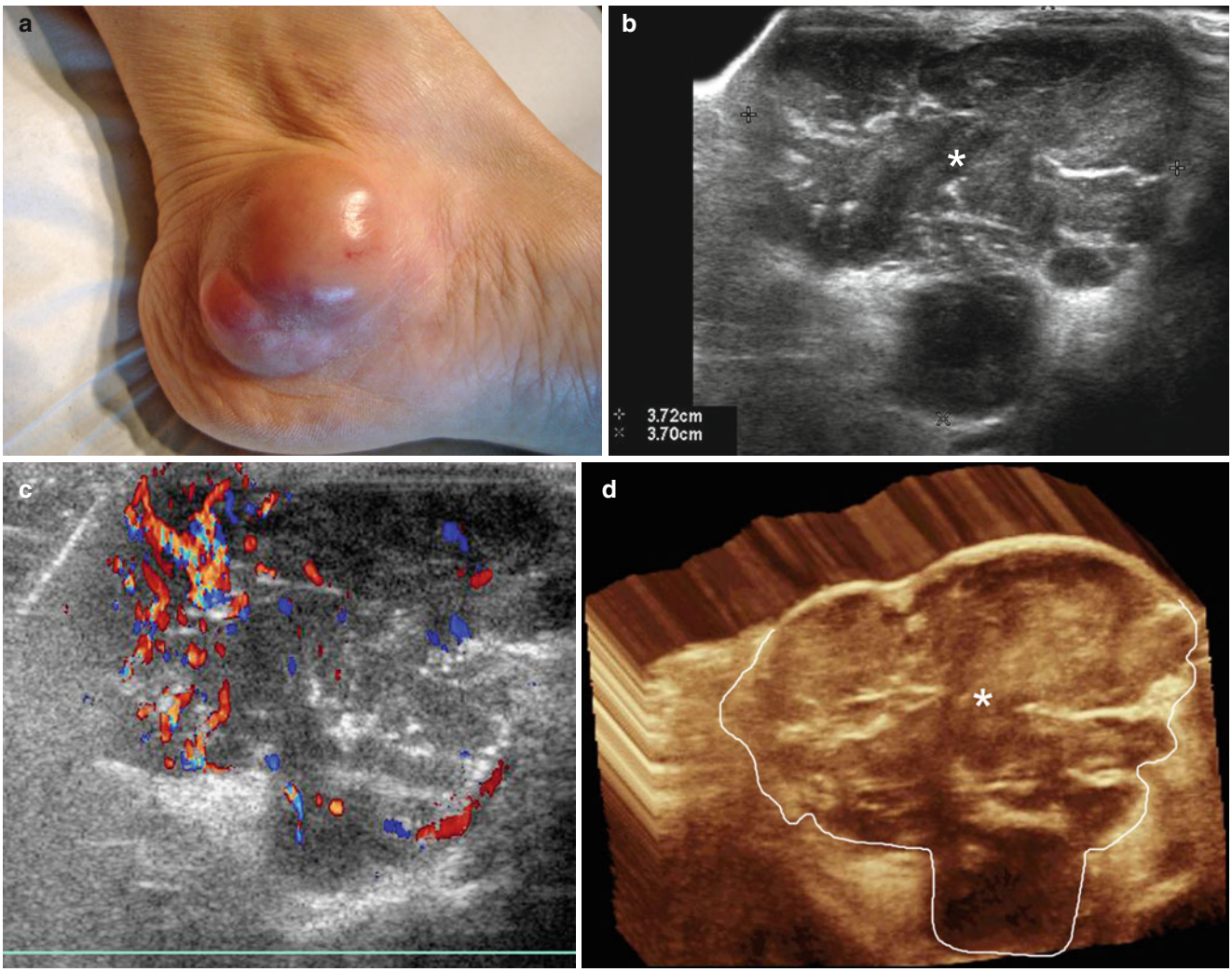


Fig. 8.10 (**a**–**d**) Cutaneous angiosarcoma. (**a**) Clinical image shows erythematous lump in the inner aspect of the left foot. (**b**) Grey scale ultrasound (transverse view) demonstrates 3.72×3.70 cm lobulated, hypoechoic and solid mass (*) that involves the subcutaneous tissue and also affects the dermis. (**c**) Color Doppler ultrasound (transverse view) shows irregular and asymmetric hypervascularity within the mass. (**d**) The tumor in 3D (*, and outlined, 5–8 s sweep)

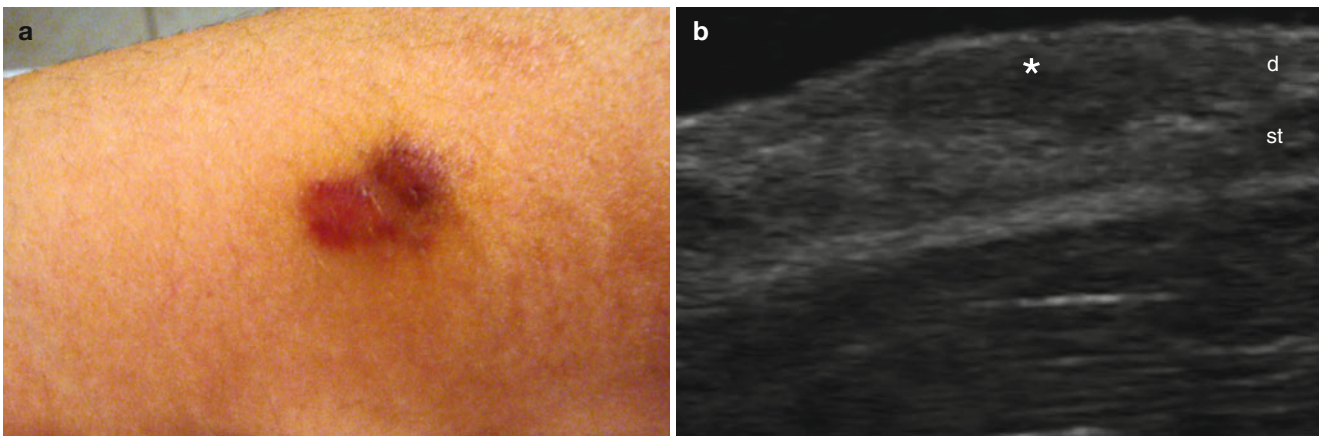
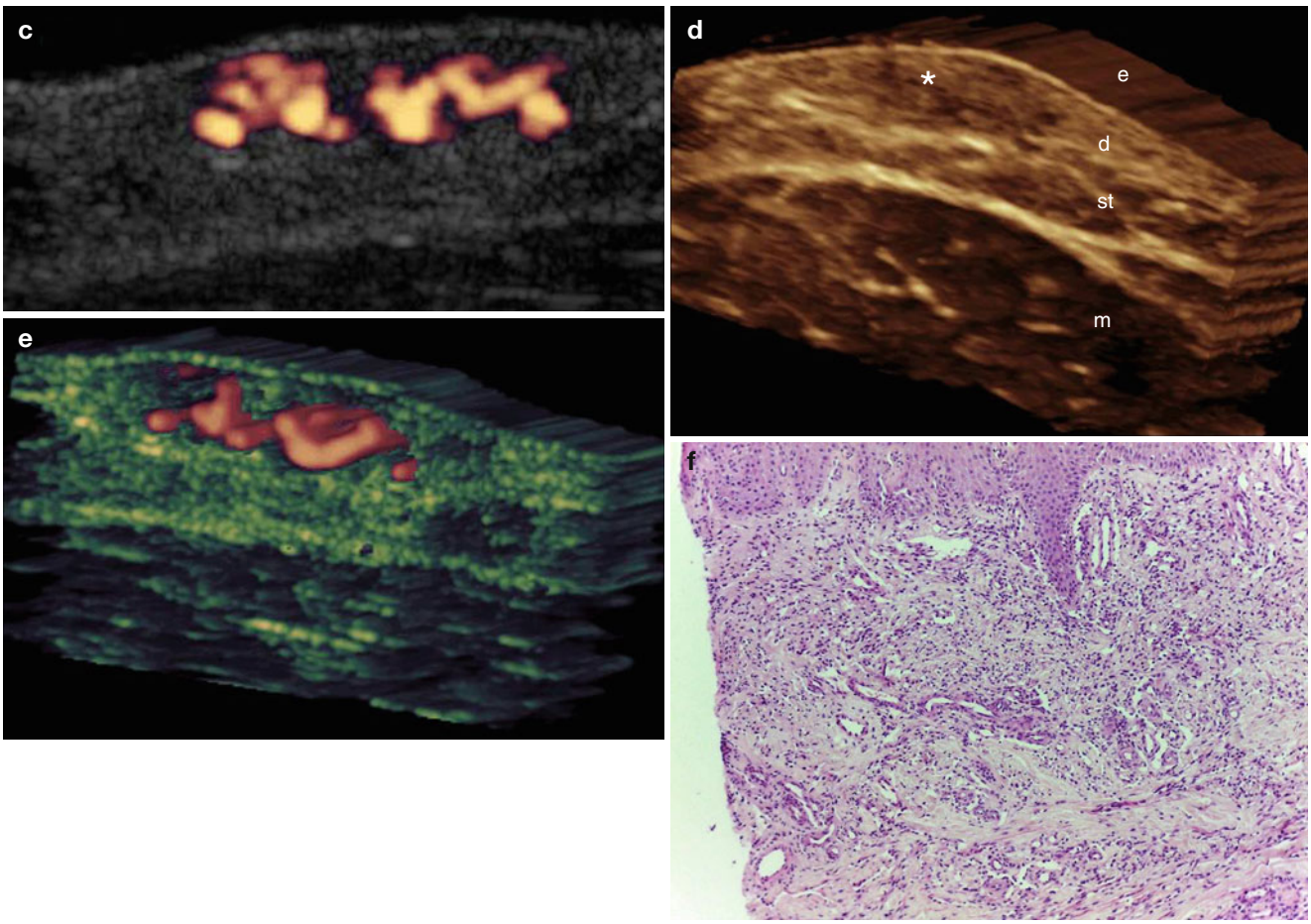


Fig. 8.11 (**a**–**f**) Kaposi's sarcoma. (**a**) Clinical reddish lesion in the inner aspect of the left arm (HIV positive patient). (**b**) Grey scale ultrasound (longitudinal view) shows ill-defined, focal hypoechogenic thickening (*) that involves the dermis and upper subcutaneous tissue. There is increased echogenicity of the lower subcutaneous tissue. (**c**) Power angio (longitudinal view) demonstrates increased blood flow within the lesion (*). (**d**) 3D reconstruction of the lesion (*, transverse axis). (**e**) 3D power angio reconstruction of the lesional area (5–8 s sweep). (**f**) Histology (HE 40 × zoom) shows a dermal vascular proliferation. *Abbreviations*: *e* epidermis, *d* dermis, *st* subcutaneous tissue, *m* muscle



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