Chapter 2
Vascular Tumors

- Vascular tumors (Table 2.1)
- Telangiectasias (Table 2.2)
- Port wine stain
  - Clinical
    - Presents at birth or shortly thereafter
    - Progressively darkens, thickens, and becomes more nodular with age
    - Associated with Sturge–Weber syndrome, Klippel–Trenaunay syndrome, and Cobb syndrome
    - Progressive ectasia believed to be caused by abnormal autonomic regulation – decreased nerves present in lesional skin
  - Histologic (Figs. 2.1 and 2.2)
    - Ectatic, thin-walled vessels in superficial to mid-dermis
    - Vascular ectasia becomes more prominent with increasing age
    - Difficult to diagnose in young children (without clinical history) – appears virtually normal

- Hereditary hemorrhagic telangiectasia
  - Clinical
    - Autosomal dominant inheritance
    - Nosebleeds in children
    - Telangiectasias appear on mucosal surfaces and diffusely on skin
Table 2.1  Vascular tumors

Telangiectias
Intravascular papillary endothelial hyperplasia
Hemangiomas
Lymphangiomas
Non-hemangiomatous tumors

Table 2.2  Telangiectasias

Nevus flammeus (port wine stains)
Hereditary hemorrhagic telangiectasia
Angiokeratoma
Venous lake

Fig. 2.1 Port wine stain demonstrates vascular ectasia but no increase in numbers of dermal blood vessels. Original magnification 40×

- Histologic
  - Ectatic post-capillary venules
  - Thin-walled vessels
  - No inflammation
Angiokeratoma

- Clinical
  - Acquired telangiectasia
  - Epidermal hyperplasia is secondary
  - Mibelli type – multiple telangiectasias on dorsa and sides of fingers
  - Fordyce type – multiple telangiectasias on scrotum
  - Associated with Fabry’s disease – multiple angiokeratomas, renal disease
  - Angiokeratoma circumscriptum is probably a true hemangioma (not a telangiectasia) – is probably misnamed

- Histologic (Figs. 2.3 and 2.4)
  - Dermal papillae expanded by dermal capillaries
  - Epidermal acanthosis with elongated rete ridges engulfing ectatic vessels
  - Deeper vessels may rarely be involved

Fig. 2.2 Widely ecstatic vessels are present in the papillary and superficial reticular dermis in port wine stains. Original magnification 100×
Fig. 2.3  Ectatic vessels are located high in the papillary dermis and surrounded by acanthotic epidermis in angiokeratoma. Original magnification 40×

Fig. 2.4  The superficial, ecstatic vessels in angiokeratoma are usually congested and often thrombosed. Original magnification 100×
• Venous lake
  – Clinical
    ○ Slowly enlarging ectatic vascular lesion usually in elderly men with marked sun exposure
  – Histologic (Fig. 2.5)
    ○ Dilated vessel, usually filled with erythrocytes and often thrombosed surrounded by marked solar elastosis
• Intravascular Papillary Endothelial Hyperplasia (IVPEH)
  – Clinical
    ○ Also known as Masson’s lesion
    ○ Tender-painful red-blue dermal nodule, often sudden appearance and growth
  – Histologic (Figs. 2.6 and 2.7)
    ○ Proliferation of endothelial cells confined to lumen of larger, thrombosed vessel
    ○ Papillary infoldings may resemble angiosarcoma, but no mitoses or cellular atypia
    ○ Represents a recanalized, thrombosed blood vessel

Fig. 2.5 A venous lake consists of a single ectatic vessels in the superficial dermis, often surrounded by marked solar elastosis. Original magnification 40×
Fig. 2.6 Intravascular papillary endothelial hyperplasia is a proliferation of endothelial cells confined to within the lumen of a vessel. Original magnification 40×

Fig. 2.7 Higher magnification demonstrates a florid proliferation of endothelial cells forming interanastamosing channels and extravasated erythrocytes in intravascular papillary endothelial hyperplasia. Original magnification 200×
• Hemangiomas (Table 2.3)
• Acquired capillary hemangioma
  
  − Clinical
  
  ◦ Small, red lesions acquired in adolescence and later in life
  ◦ Associated with increased estrogens, liver disease
  
  − Histologic (Fig. 2.8)
  
  ◦ Proliferation of dilated capillaries in the superficial dermis

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<tr>
<th>Table 2.3  Hemangiomas</th>
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<td>Acquired capillary hemangioma</td>
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<tr>
<td>Infantile capillary hemangioma</td>
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<td>Cavernous hemangioma</td>
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<td>Verrucous hemangioma</td>
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<td>Lobular capillary hemangioma (pyogenic granuloma)</td>
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<td>Arteriovenous hemangioma</td>
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<td>Acquired tufted hemangioma</td>
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<tr>
<td>Targetoid hemangioma (hobnail hemangioma)</td>
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**Fig. 2.8** Capillary hemangioma is characterized by a proliferation of bland appearing blood vessels in the superficial portion of the dermis. These vessels may be fully congested with erythrocytes. Original magnification 100×
• Infantile capillary hemangioma
  – Clinical
    ◦ Usually present within days of birth and grow rapidly for up to a year
    ◦ May involute in up to 75% of cases, usually starting at about age 5
    ◦ Usually in area surrounding parotid gland, but can grow to involve airways and to be life-threatening
  – Histologic (Fig. 2.9)
    ◦ Early lesions – marked cellularity, may have jig-saw puzzle-like appearance at low magnification
    ◦ Difficult to find vessels – very small
    ◦ Mitoses are abundant
    ◦ Older lesions with less cellularity and more prominent vessels

• Lobular capillary hemangioma (pyogenic granuloma)
  – Clinical
    ◦ Rapid onset of exophytic red nodule

Fig. 2.9 Juvenile (infantile) capillary hemangioma demonstrates a markedly cellular proliferation with scattered blood vessels that are often devoid of erythrocytes. Original magnification 200×
○ Usually ulcerated
○ Most common on fingers, mucous membranes, but can occur anywhere
○ Increased numbers associated with pregnancy
○ Can occur in any age patient

– Histologic (Figs. 2.10 and 2.11)

○ Collarette of rete ridges surrounds proliferation of well-formed small vessels
○ Feeder vessel at deep margin ramifies into many smaller vessels
○ Endothelial cell mitoses and mild cytologic atypia may be present
○ Infiltrate of inflammatory cells most prominent at surface where ulceration is common

Fig. 2.10 Lobular capillary hemangioma (pyogenic granuloma) is characterized by an exophytic proliferation of small blood vessels surrounded by an epidermal collarette. Original magnification 40×
Acquired tufted angioma (angioblastoma)
  - Clinical
    - Benign, progressive angiomatosis most common in children and younger adults
    - Slowly enlarging erythematous macules
    - Usually on trunk and upper extremities
  - Histologic (Figs. 2.12 and 2.13)
    - Cellular lobules dispersed throughout the reticular dermis
    - Ovoid or spindle-shaped cells without atypia
    - Cleft-like luminal spaces
    - Masses of cells protrude into and distort surrounding thin-walled vessels

Targetoid hemosiderotic hemangioma
  - Clinical
    - Most common in young to middle-aged men
    - Usually single, on trunk and proximal extremities
    - Central papular area with centrifugal areas of bronze discoloration
Vascular Tumors

Histologic (Figs. 2.14 and 2.15)
- Vascular spaces lined by plump endothelial cells with papillary projections into lumen
- Abundant hemosiderin deposits, especially laterally
- Poorly circumscribed and infiltrative growth pattern at periphery

Fig. 2.12 Acquired tufted angioma demonstrates a lobular pattern of dermal involvement. Original magnification 20×

Fig. 2.13 The lobules consist of angulated blood vessels surrounded by spindle-shaped cells in acquired tufted angioma. Original magnification 100×
**Fig. 2.14** Targetoid hemosiderotic hemangioma demonstrates thin vessels with prominent endothelial cells and surrounding hemosiderin. Original magnification 200×

**Fig. 2.15** Targetoid hemosiderotic hemangioma (hobnail hemangioma) demonstrates prominent endothelial cell protrusion into the lumen in a hobnail-like pattern. Original magnification 400×
• Lymphangiomas (Table 2.4)
• Lymphangioma circumscriptum
  – Clinical
    ◦ Grouped vesicular papules, often on flank and thighs
    ◦ May appear fluid filled or dark (if hemorrhagic and thrombosed)
  – Histologic (Fig. 2.16)
    ◦ Indistinguishable from angiokeratoma except if vessels are devoid of erythrocytes
    ◦ Epidermal collarette surrounds ectatic vessels in papillary dermis
• Non-hemangiomatous tumors (Table 2.5)

**Table 2.4  Lymphangiomas**

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<th>Lymphangioma circumscriptum</th>
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<tr>
<td>Cystic hygroma</td>
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<td>Acquired progressive lymphangioma</td>
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**Fig. 2.16** Thin-walled vessels, often containing serum but few cells, are present in the superficial dermis in lymphangioma circumscriptum. There is often an epidermal collarette. Original magnification 200×
**Table 2.5**  Non-hemangiomatous tumors

<table>
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<tr>
<th>Non-hemangiomatous Tumors</th>
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<tr>
<td>Angiolymphoid hyperplasia with eosinophilia (ALHE)</td>
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<td>Spindle cell hemangioendothelioma</td>
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<td>Glomus tumor (glomangioma)</td>
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<td>Hemangiopericytoma</td>
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- **ALHE**
  - **Clinical**
    - One or several erythematous papules, nodules
    - Most common around ears and on face
    - Young to middle-aged adults
    - Synonymous with epithelioid hemangioma
  - **Histologic (Figs. 2.17–2.19)**
    - Abundant small vessels with swollen endothelial cells, often occluding lumen – difficult to identify as vessels in some cases
    - Dense inflammatory infiltrate of lymphocytes and eosinophils surrounding vascular proliferation
    - Often completely filling dermis, may be germinal centers present

![Fig. 2.17](image)  Angiolymphoid hyperplasia with eosinophilia demonstrates a marked inflammatory infiltrate at lowest magnification. Original magnification 40×
Fig. 2.18 At high magnification, swollen endothelial cells may make vessels appear to be small granulomas in angiolymphoid hyperplasia with eosinophilia. The percentage of eosinophils present in the infiltrate is highly variable. Original magnification 400×

Fig. 2.19 In some cases of angiolymphoid hyperplasia with eosinophilia, there are many eosinophils surrounding the thickened blood vessels. Original magnification 400×
• Spindle cell hemangioendothelioma
  – Clinical
    ◦ May be reactive, and not neoplastic
    ◦ 50% of cases in people <25
    ◦ Single or multiple, but multiple confined to single region
    ◦ Hands and feet most frequent
    ◦ Local recurrence after removal
  – Histologic
    ◦ Three major components
      Thin-walled cavernous vessels that may contain thrombi
      Solid area of spindle cells with slit-like vascular spaces
      (like Kaposi’s sarcoma)
      Plump endothelial cells in groups or lining vascular
      spaces – intracytoplasmic vacuoles
    ◦ Minimal cytologic atypia or mitotic activity

• Glomus tumor
  – Clinical
    ◦ Erythematous to blue nodules, most common on
      extremities
    ◦ Tender/painful
  – Histologic (Fig. 2.20)
    ◦ Normal blood vessels surrounded by proliferation of
      bland, uniform appearing cells with central dark nuclei
      and relatively scant cytoplasm
    ◦ Cells express smooth muscle actin
    ◦ “Glomangioma” is variant with concomitant proliferation
      of vessels and glomus cells
    ◦ Glomangiosarcoma – very rare

• “Malignant” vascular tumors (Table 2.6)
• Kaposi’s sarcoma
  – Clinical
    ◦ Four subtypes
      Classic – men – fifth to seventh decades, E. European
      Jews, lower extremities, long chronic course
African (endemic) – marked male predominance, can resemble classic type or involve nodes, also frequent EBV infection in these patients

Immunosuppressive – younger patients, no male predominance, tumors appear with treatment and abate with cessation, more aggressive than classic – may result in death from GI hemorrhage

HIV associated – intravenous drug abusers and homosexual men, 10% of AIDS patients, widespread lesions, mucosal involvement, not usually cause of death

○ All associated with HHV-8 infection

- Histologic (Figs. 2.21–2.24)

○ Patch – irregular vascular spaces surrounding pre-existing vessels, around appendages, between collagen bundles; lymphocytes and plasma cells

**Table 2.6**  “Malignant” vascular tumors

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<tr>
<th>Angiosarcoma</th>
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<td>Kaposi’s sarcoma</td>
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**Fig. 2.20**  Glomus tumors are characterized by a proliferation of monomorphic, cuboidal cells with small, dark, central nuclei surrounding small blood vessels. Original magnification 400×
Fig. 2.21 Patch-stage Kaposi’s sarcoma is characterized by a sometimes subtle proliferation of endothelial cells forming slit-like vascular spaces in the superficial to mid-dermis. Original magnification 100×

Fig. 2.22 Normal vessels are often surrounded by the slit-like vessels and increased numbers of endothelial cells in Kaposi’s sarcoma. Extravasation of erythrocytes is commonly seen. Original magnification 400×
Fig. 2.23 In nodular lesions of Kaposi’s sarcoma, the proliferation of endothelial cells may be quite dense but there is little cytologic pleomorphism, often despite brisk mitotic activity. Original magnification 400×

Fig. 2.24 Degenerating erythrocytes may appear as small pink globules within vessels or in the surrounding dermis in Kaposi’s sarcoma. Original magnification 600×
• Plaques and tumors – dermal proliferation of interlacing bundles of spindle cells and poorly defined vascular spaces; variable nuclear pleomorphism, rare mitoses, extravasated erythrocytes, hyaline globules

- Angiosarcoma
  - Clinical
    - Head and neck type
      - Elderly patients, M>F
      - Single, blue – violaceous nodules and plaques
        - 15% 5-year survival
    - Arising in lymphedematous limbs (Stewart–Treves)
      - 12.5 years post-surgery
    - Post-irradiation
      - 10–20 years post-irradiation

Fig. 2.25 Angiosarcoma is characterized by a proliferation of angulated, interanastamosing channels throughout the dermis. The early lesions may be subtle. Original magnification 100×
Fig. 2.26 The interanastamosing channels are lined by prominent protruding endothelial cells in angiosarcoma. Original magnification 400×

Fig. 2.27 In some cases, the proliferating endothelial cells may be markedly spindly in shape. Original magnification 100×
Histologic (Figs. 2.25–2.28)

- Poorly circumscribed proliferation of interanastamosing vascular spaces
- Vessels lined with atypical endothelial cells
- May be solid areas
- Dense lymphocytic infiltrate in some cases may obscure vascular proliferation
- Well-differentiated can resemble benign angiomatous process, poorly differentiated can be difficult to identify as vascular

Fig. 2.28 In other cases, the endothelial cells may demonstrate pronounced atypia and have an epithelioid morphology, making it difficult to determine the nature of the malignant process. Original magnification 200×
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