DERMATOLOGY WORLD
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# Vascular neoplasms

by Jesse Hirner, MD

| Tumor | Comments | Histopathology |
|---|---|---|
| **Benign tumors** | | |
| Infantile hemangioma | Risk factors: prematurity, female sex, placental abnormalities; Beard area hemangioma – risk for airway hemangioma Periorbital hemangioma – risk for amblyopia & astigmatism Treatment: systemic or topical β-blockers are 1st line treatment; can use PDL for ulcerated hemangiomas | Small dermal +/- subcutaneous capillaries with variably dense endothelial cell proliferation, GLUT-1 positive |
| Rapidly involuting congenital hemangioma (RICH) | Fully developed at birth, rapidly involutes by 1 year | GLUT-1 negative |
| Noninvoluting congenital hemangioma (NICH) | Fully developed at birth, grows proportionally with patient, does not involute | GLUT-1 negative |
| Pyogenic granuloma (lobular capillary hemangioma) | Rapidly growing, frequently ulcerated; associations: trauma, pregnancy, OCPs, retinoids, indinavir, EGFR-inhibitors May occur intraorally or on lips, Bacillary angiomatosis & verruga perruana (2nd stage of Oroya fever) may resemble PGs Treatment: procedural in most cases, shave removal +/- curettage and electrodessication, PDL, C02 laser | Exophytic, lobular, capillary proliferation, epidermal collarette present, may be ulcerated |
| Bacillary angiomatosis | Red papulonodules; Bartonella henselae and B. quintana. Immunocompromised patients, especially HIV patients, often cat exposure. Treatment: Erythromycin 1st line, can use doxycycline, azithromycin, clarithromycin; Jarisch-Herxheimer reaction may occur. | Similar to pyogenic granuloma with neutrophils; can use GMS or Warthin-Starry stains to see organisms |
| Glomus tumor | Tumor of perivascular modified smooth muscle cells of the Sucquet-Hoyer canal Most common site is subungual, painful, adult onset | Abundant glomus cells around vascular lumens; SMA, desmin, vimentin positive |
| Glomangioma (glomuvenous malformation) | Arises in infancy or childhood, often multiple, nonpainful May be sporadic or inherited in AD fashion, glomulin mutation | Large vascular lumens, surrounded by one to a few layers of glomus cells |

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| Tufted angioma | Most commonly neck or trunk, congenital or acquired, grows slowly, may be a/w Kasabach-Merritt phenomenon | "Cannonball" highly cellular lobules in dermis +/- subcutis |
| Glomeruloid hemangioma | Red-purple papules, trunk & extremities, a/w POEMS syndrome & less commonly multicentric Castleman's disease, ↑ VEGF, POEMS – polyneuropathy, organomegaly, endocrinopathy, M-protein, skin changes | Dilated dermal vascular lumen containing multiple capillary loops resembling renal glomerulus |
| Epithelioid hemangioma (angiolymphoid hyperplasia with eosinophils) | Grouped papules or nodules, head & neck, often around ear, young to middle aged adults, treatment - excision | Large & small lumens, hobnail endothelial cells with intracytoplasmic lumen formation, lymphocytic inflammation with eosinophils |
| Targetoid hemosiderotic hemangioma (hobnail hemangioma) | Acquired, targetoid clinical appearance, legs most common location, children and middle-aged adults | Large, ectatic, thin walled vessels superficial to smaller vessels, hobnail endothelial cells (protrude into lumen), hemosiderin deposition |
| **Tumors of borderline or low-grade malignant potential** | | |
| Kaposi sarcoma | Purple to red-brown macules to plaques, HHV-8 present in 100%, 4 variants, ranges from indolent to aggressive, may affect mucosal surfaces and viscera<br>HHV-8 also a/w Castleman's disease & primary effusion lymphoma<br>Treatment: retinoids, surgery, cryotherapy, laser, radiation, chemotherapy for extensive disease, HAART if AIDS-related | Patch, plaque, and nodular stage, spindle cells, slit-like & staghorn vessels, promontory sign, extravasated RBCs, hemosiderin, plasma cells, intra- & extracellular eosinophilic globules ("Dorf balls"), less cytologic atypia than angiosarcoma, nuclear positivity for latency-associated nuclear antigen 1 (LANA-1) IHC stain |
| Kaposiform hemangioendothelioma | Vascular macule, patch, plaque, or tumor, onset usually before age 2, can occur in retroperitoneum and present as ecchymoses<br>A/w Kasabach-Merritt phenomenon: consumptive coagulopathy → apparent growth of tumor, purpura, ↓ plts, DIC, hemolytic anemia, bleeding, risk of high-output cardiac failure<br>Treatment: steroids & vincristine | Densely cellular lobules in dermis, spindle cells, narrow lumens, CD31 & CD34 positive, GLUT-1 and factor VIII negative |

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| Atypical Vascular Lesion (AVL) | Erythematous to violaceous lesion on breast in patient s/p radiation for breast cancer; a minority progress to angiosarcoma<br>Treatment: typically excised | Vascular type AVL: proliferation of capillary sized vessels in dermis (CD31, CD34 positive, D2-40 negative)<br>Lymphatic type: thin-walled, anastomosing vessels (D2-40 positive<br>May have some atypia, but less than angiosarcoma |
| Hemangiopericytoma | Expanding dermal or deep soft tissue mass<br>Infantile hemangiopericytoma (<10% of cases): More commonly involves skin than adult type, birth to 1 year old, better prognosis than adult hemangiopericytoma, possibly related to infantile myofibromatosis<br>Adult hemangiopericytoma (>90%): Typically deep soft tissue, more aggressive than infantile form<br>Treatment: Resection, sometimes radiation used, some cases have spontaneously regressed. | Adult type: "Staghorn" vessel lumens and densely packed spindle cells<br>Infantile type: Same as adult with surrounding lobules that are less densely cellular<br>CD34 and CD99 positive |
| **Malignant tumors** | | |
| Angiosarcoma | Seen in:<br>- elderly on sun-damaged sites, often men on scalp and face<br>- lymphedematous sites (Stewart-Treves syndrome), often from lymphaedenectomy after breast cancer<br>- postradiation, often after breast cancer<br>Treatment: Excision +/- radiation, high rate of recurrence | Cytologic atypia with hyperchromatic nuclei and pleomorphism, hemorrhage, narrow vascular spaces, multilayered endothelial cells, mitoses; CD31, CD34, ERG, FLI-1, ulex europaeus, & factor VIII positive, MYC positive in postradiation angiosarcoma |

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