# Multiple nodules on the unilateral upper limb

Luisa Lobato Macias, MD,[a] Luciana Mendes dos Santos, MD,[a] Patricia Motta de Morais, MD,[a,b] Patricia Amaral Couto, MD,[a] Patricia Chicre Bandeira de Melo, MD,[a] Mariana Santiago Bernardes, MD,[a] Laísa Ezaguy de Hollanda, MD,[a] Joelly Taynara Lapinski Levermann, MD,[a] Aline Sales Mendes Záu, MD,[a] Ana Paula Coelho Rocha, MD,[a] and Monique Freire Santana, MD[c]

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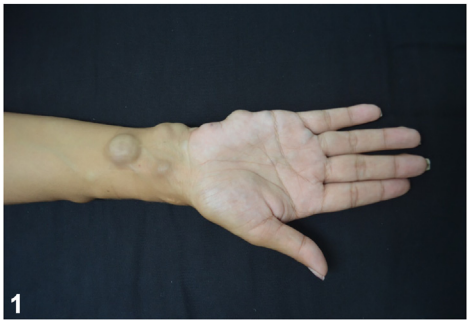

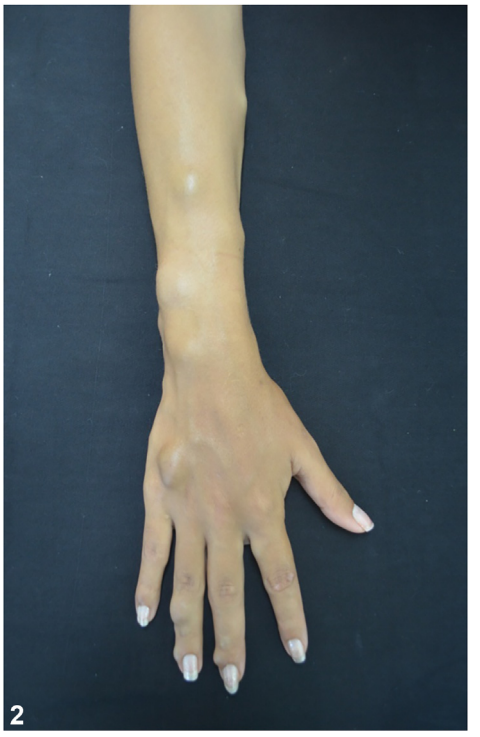

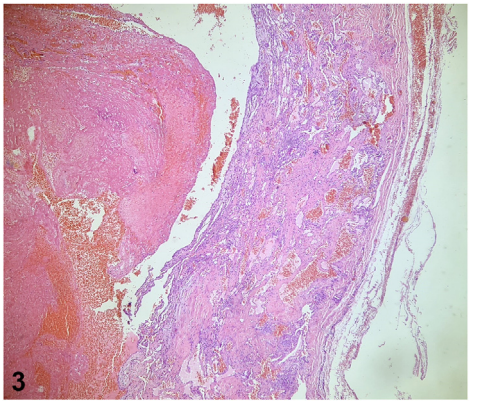

A 28-year-old woman presented with multiple bluish nodules of various sizes, which are painless, of soft consistency, not adhered to deep planes, and segmentally distributed along the right upper limb (Figs 1 and 2) with a 5-years evolution. Magnetic resonance imaging demonstrated heterogeneous nodular images in the subcutaneous adipose planes, and soft tissue ultrasonography pointed to the presence of echogenic material and thrombosis intravascular histopathological examination of the incisional biopsy (Fig 3).

**Question 1: What is the most likely diagnosis?**

**A.** Segmental neurofibromatosis

**B.** Angiolipomas

**C.** Angiosarcoma

**D.** Masson's tumor

**E.** Epithelioid hemangioendothelioma

**Answer:**

**A.** Segmental neurofibromatosis — Incorrect. Neurofibromatosis type 5 is characterized by segmental and unilateral neurofibromas associated, or not, with Café-au-Lait patches. Neurofibromas are neural tumors that are brownish, soft, smooth-surfaced, painful nodules with a herniated ring to palpation.[1]

**B.** Angiolipomas — Incorrect. These are benign tumors with excessive vascular proliferation in the middle of the fatty tissue, and fibrin thrombi are common. Clinically, they are painful, encapsulated nodules in the subcutaneous compartment. They are usually numerous and are located on the arms, legs, and abdomen.[1]

**C.** Angiosarcoma — Incorrect. Angiosarcoma is a rare vascular neoplasm, with aggressive behavior, high recurrence rates, and early metastases and generally evolving to death. It manifests as an erythematous plaque or nodule, with an infiltrative aspect, soft consistency, and rapid growth. The cutaneous form is the most common, affecting the scalp, face, and neck of elderly Caucasian men.[1]

**D.** Masson's tumor — Correct. Intravascular papillary endothelial hyperplasia (IPEH), or Masson's tumor, is a rare, benign intravascular lesion caused by proliferation of papillary endothelial structures associated with degenerative alterations. It usually affects the subcutaneous and dermis, as a single lesion, more common in women. Clinically, Masson's tumor presents as a firm or soft nodule, not adherent to deep planes, bluish-red in color, without pulsatility.[2]

**E.** Epithelioid hemangioendothelioma — Incorrect. It is a rare vascular tumor with intermediate aggressiveness that presents as a solitary, painful, angiomatous soft tissue tumor usually occurring in adults. The diagnosis is histological, where there is proliferation of endothelial cells of epithelioid aspect forming vascular channels.[1]

**Question 2: Choose the correct alternative about Masson's tumor below:**

**A.** Fast-growing, aggressive behavior

**B.** No involvement of endothelial growth factors

**C.** Presents as a primary vascular tumor

**D.** Most cases arise from trauma

**E.** Histologically, angiosarcoma is its main differential diagnosis

**Answer:**

**A.** Fast-growing, aggressive behavior — Incorrect. Masson's tumor is a benign, well-circumscribed, slow-growing lesion,[3] of a reactive character.[4]

**B.** No involvement of endothelial growth factors — Incorrect. Excessive endothelial proliferation stimulated by local production of growth factors is postulated in the etiology of IPEH. Studies have

From the Dermatology Department, Universitary Hospital Getúlio Vargas, Manaus, Amazonas, Brazil[a]; Afredo da Matta Foundation[b]; and Pathology Department, Federal University of Amazonas, Manaus, Amazonas, Brazil.[c]

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Correspondence to: Luisa Lobato Macias, MD, Dermatology Department, University Hospital Getúlio Vargas, Professor Marciano Armond St, n° 963, São Francisco, CEP: 69079-015, Manaus, Amazonas, Brazil. E-mail: luisalmacias@gmail.com.

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revealed increased expression of fibroblast growth factor beta, suggesting its secretion by endothelial cells.[4]

**C.** Presents as a primary vascular tumor − Incorrect. Although IPEH arises within a vessel, it is not a primary vascular tumor. After vascular injury, there is thrombus formation within a vessel, followed by inflammation and vascular stasis, with proliferation of endothelial cells. Chemotaxically attracted macrophages release basic endothelial fibroblast growth factor, which stimulates the formation of IPEH.[2]

**D.** Most cases arise from trauma − Incorrect. There are 3 origin-based categories: type I representing a 'de-novo' incident arising from normal blood vessels, while type II can develop from a preexisting vascular process: hemangioma, pyogenic granuloma, or hematoma. Type III, the least common variant, has an extravascular location and typically arises from post-traumatic hematomas; only 4% of patients report such a history.[5]

**E.** Histologically, angiosarcoma is its main differential diagnosis − Correct. Histologically, its main differential diagnosis involves angiosarcoma, which is distinguished by the absence of necrosis, cell atypia, and mitotic figures.[4]

### Question 3: Choose the alternative that represents the histopathology of Masson's tumor:

**A.** The presence of spiculated cells with little atypia and few mitoses

**B.** Compacted vascular or capillary clumps are observed throughout the dermis, with a typical "cannonball" pattern

**C.** The presence of cystic vascular structures with hemorrhagic content, with intraluminal papillary formations covered by an endothelium with soft cytology

**D.** The presence of infiltrate at the subcutaneous tissue level, papillary endothelial hyperplasia, prominent nucleoli, mitotic figures, significant cytological atypia

**E.** Endothelial cell lobules arranged in solid strands and masses, with few capillary lumens in superficial dermis

### Answer:

**A.** The presence of spiculated cells with little atypia and few mitoses − Incorrect. the presence of spiculated cells with little atypia and few mitoses suggests the diagnosis of hemangioendothelioma.[1]

**B.** Compacted vascular or capillary clumps are observed throughout the dermis, with a typical "cannonball" pattern − Incorrect. the presence of vascular clumps with a typical "cannonball" pattern, with crescent-shaped spaces around the clumps, and with lymphatic-like spaces in the tumor stroma is suggestive of angioma.[1]

**C.** The presence of cystic vascular structures with hemorrhagic content, with intraluminal papillary formations covered by an endothelium with soft cytology − Correct. the histopathology of Masson's tumor is characterized by the presence of cystic vascular structures with hemorrhagic content, with intraluminal papillary formations covered by an endothelium with soft cytology. No necrosis, cell pleomorphism, or mitotic figures are observed.[4]

**D.** The presence of infiltrate at the subcutaneous tissue level, papillary endothelial hyperplasia, prominent nucleoli, mitotic figures, significant cytological atypia − Incorrect. subcutaneous infiltration, papillary endothelial hyperplasia, prominent nucleoli, mitotic figures, significant cytological atypia, and dermal collagen dissection suggest angiosarcoma.[5]

**E.** Endothelial cell lobules arranged in solid strands and masses, with few capillary lumens in superficial dermis − Incorrect. the description is compatible with infantile hemangioma in the proliferative phase, with the presence of lobules of endothelial cells arranged in solid strands and masses, with few superficial dermal capillary lumens.[1]

### Abbreviation used:

IPEH: intravascular papillary endothelial hyperplasia

**Conflicts of interest**

None disclosed.

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