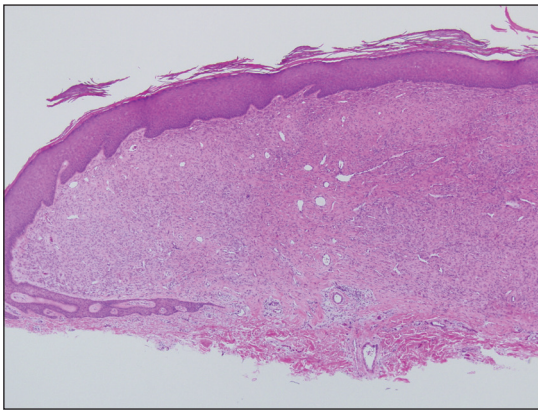


# Solitary Papule on the Leg

Qiong Wu, MD; Daniel C. Skipper, DO; Dirk M. Elston, MD

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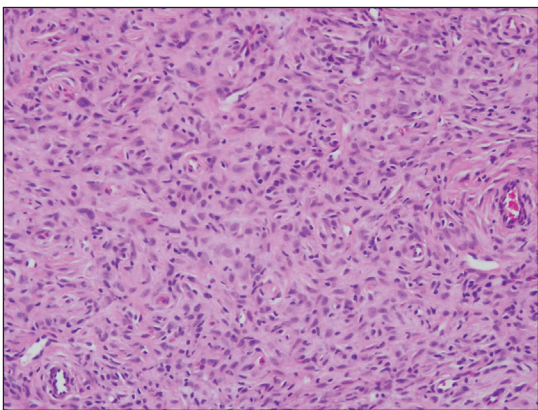
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A 28-year-old man presented with a growing asymptomatic papule on the right leg.

## THE BEST DIAGNOSIS IS:

- epithelioid histiocytoma
- myopericytoma
- polypoid Spitz nevus
- solitary fibrous tumor
- solitary reticulohistiocytoma



H&E, original magnifications  $\times 40$  (top) and  $\times 200$  (bottom).

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The authors report no conflict of interest.

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## THE DIAGNOSIS: Epithelioid Histiocytoma

**E**pithelioid histiocytoma (EH), also known as epithelioid cell histiocytoma or epithelioid fibrous histiocytoma, is a rare benign fibrohistiocytic tumor first described in 1989.<sup>1</sup> Epithelioid histiocytoma commonly presents in middle-aged adults with a slight predilection for males.<sup>2</sup> The most frequently affected site is the lower extremity. The arms, trunk, head and neck, groin, and tongue also can be involved.<sup>3,4</sup> It usually presents as a solitary asymptomatic papule or nodule, though cases with multiple lesions have been reported.<sup>5</sup> Anaplastic lymphoma kinase rearrangement and overexpression have been confirmed and suggest that EH is distinct from conventional cutaneous fibrous histiocytoma.<sup>6</sup>

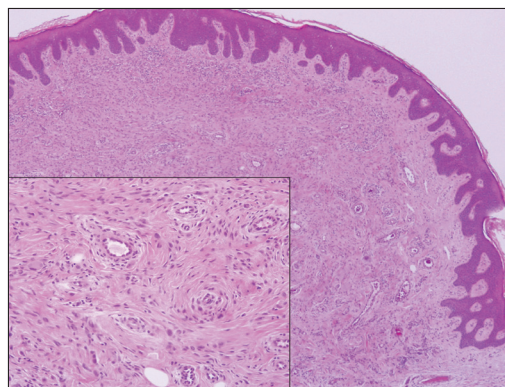
Histologically, EH appears as an exophytic, symmetric, and well-demarcated dermal nodule with a classic epidermal collarette. Prominent vascularity with perivascular accentuation of the epithelioid tumor cells is common. Older lesions may be hyalinized and sclerotic. Epithelioid cells commonly account for more than 50% of the tumor and are characterized by eosinophilic cytoplasm, vesicular nuclei, and small eosinophilic nucleoli. A small population of lymphocytes and mast cells are variably present (quiz image, bottom).<sup>1-3,7</sup> A predominantly spindle cell variant has been reported.<sup>8</sup> Other histopathologic variants include granular cell,<sup>9</sup> cellular,<sup>10</sup> and EH with perineuriomalike growth.<sup>11</sup> Immunohistochemical staining shows anaplastic lymphoma kinase positivity in most cases, and more than half of cases stain positive for factor XIIIa and epithelial membrane antigen. Tumor cells consistently are negative for desmin and cytokeratins.<sup>5,10,12</sup> Excision is curative.<sup>8</sup>

Polypoid Spitz nevus (PSN) is a benign nevus with a conspicuous polypoid or papillary exophytic architecture. The term was coined in 2000 by Fabrizi and Massi.<sup>13</sup> Spitz nevus is a benign acquired melanocytic tumor that typically presents in children and adolescents and has a wide histologic spectrum.<sup>14</sup> There is some debate on this entity, as some authors do not regard PSN as a distinct histologic variant; thus, it seems underreported in the literature.<sup>15</sup> In a review of 349 cases of Spitz nevi, the authors found 7 cases of PSN.<sup>16</sup> In another review of 74 cases of intradermal Spitz nevi, 14 cases of PSN were identified.<sup>14</sup> This polypoid variant is easily mistaken for a polypoid melanoma because it can show cytologic atypia with large nuclei. Polypoid Spitz nevus usually lacks mitoses, notable pleomorphism, and sheetlike growth, unlike melanoma (Figure 1).<sup>13,14</sup>

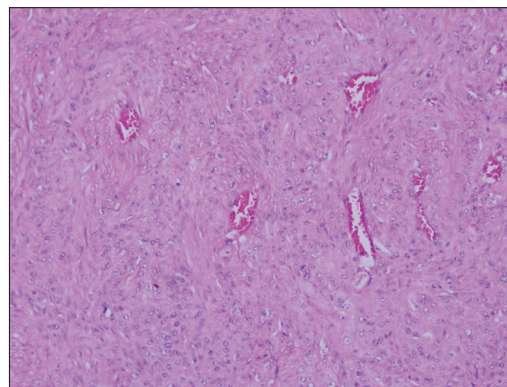
Myopericytoma is an uncommon benign mesenchymal neoplasm that typically presents as a solitary, slowly enlarging and painless nodule with a predilection for the lower extremities, usually in adult males.<sup>17-20</sup> Histologically, it consists of a well-circumscribed nodule with numerous thin-walled vessels and a proliferation of

ovoid to spindled myopericytes exhibiting a concentric perivascular growth pattern (Figure 2). Myopericytoma usually is positive for smooth muscle actin and h-caldesmon but is negative or only focally positive for desmin. The prognosis is good with rare recurrence, despite incomplete excision.<sup>17,18</sup>

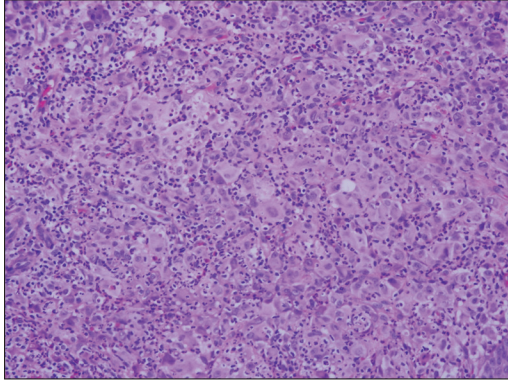
Solitary reticulohistiocytoma is a rare benign form of non-Langerhans cell histiocytosis.<sup>21,22</sup> Unlike its multicentric counterpart, solitary reticulohistiocytoma rarely is associated with systemic disease. It presents as a small, dome-shaped, painless papule or nodule that can affect any part of the body.<sup>22,23</sup> Solitary reticulohistiocytoma characteristically demonstrates cells with a ground glass-like appearance and 2-toned cytoplasm. A mixed



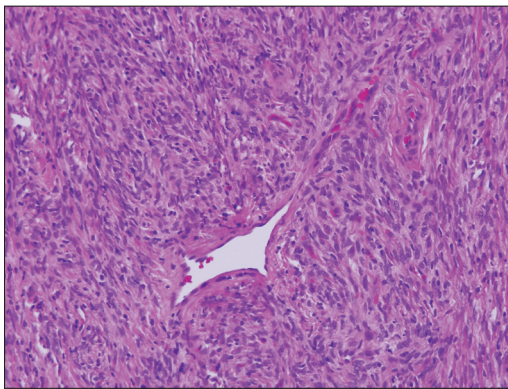
**FIGURE 1.** Polypoid Spitz nevus. A polypoid architecture with predominantly intradermal epithelioid and spindled melanocytes arranged as single units splaying between dermal collagen. The thick-walled vascular pattern is characteristic (H&E, original magnification  $\times 40$ ; inset, original magnification  $\times 200$ ).



**FIGURE 2.** Myopericytoma. A proliferation of concentric perivascular myopericytes around numerous thin-walled vessels. The cells are ovoid and plump spindled with eosinophilic cytoplasm (H&E, original magnification  $\times 200$ ).



**FIGURE 3.** Solitary reticulohistiocytoma. A dermal epithelioid histiocytic proliferation of cells with a ground glass-like appearance and 2-toned cytoplasm. A background mixed inflammatory infiltrate is present (H&E, original magnification  $\times 200$ ).



**FIGURE 4.** Solitary fibrous tumor. A dense, bland, spindled, fibroblastic proliferation with a "patternless pattern" and collagenized stroma with interspersed branching vessels (H&E, original magnification  $\times 200$ ).

inflammatory infiltrate including neutrophils, eosinophils, and lymphocytes commonly is present (Figure 3). The epithelioid histiocytes are positive for vimentin and histiocytic markers including CD68 and CD163.<sup>22</sup>

Solitary fibrous tumor (SFT) is an uncommon mesenchymal fibroblastic neoplasm that can arise at almost any anatomic site.<sup>24</sup> Cutaneous SFTs are more common in women, most often involve the head, and appear to behave in an indolent manner.<sup>25</sup> Solitary fibrous tumors are translocation-associated neoplasms with a *NAB2-STAT6* gene fusion.<sup>26</sup> The classic histology of SFT is a spindled fibroblastic proliferation arranged in a "patternless pattern" with interspersed stag horn-like, thin-walled blood vessels (Figure 4). Tumor cells usually are positive for CD34, CD99, and Bcl-2.<sup>27</sup> In addition, *STAT6* immunoreactivity is useful in diagnosis of SFT.<sup>25</sup>

## REFERENCES

- Jones EW, Cerio R, Smith NP. Epithelioid cell histiocytoma: a new entity. *Br J Dermatol.* 1989;120:185-195.
- Singh Gomez C, Calonje E, Fletcher CD. Epithelioid benign fibrous histiocytoma of skin: clinico-pathological analysis of 20 cases of a poorly known variant. *Histopathology.* 1994;24:123-129.
- Felty CC, Linos K. Epithelioid fibrous histiocytoma: a concise review [published online October 4, 2018]. *Am J Dermatopathol.* doi:10.1097/DAD.0000000000001272.
- Rawal YB, Kalmar JR, Shumway B, et al. Presentation of an epithelioid cell histiocytoma on the ventral tongue. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2005;100:75-83.
- Cangelosi JJ, Prieto VG, Baker GF, et al. Unusual presentation of multiple epithelioid cell histiocytomas. *Am J Dermatopathol.* 2008;30:373-376.
- Doyle LA, Marino-Enriquez A, Fletcher CD, et al. ALK rearrangement and overexpression in epithelioid fibrous histiocytoma. *Mod Pathol.* 2015;28:904-912.
- Silverman JS, Glusac EJ. Epithelioid cell histiocytoma—histogenetic and kinetics analysis of dermal microvascular unit dendritic cell subpopulations. *J Cutan Pathol.* 2003;30:415-422.
- Murigu T, Bhatt N, Miller K, et al. Spindle cell-predominant epithelioid fibrous histiocytoma. *Histopathology.* 2018;72:1233-1236.
- Rabkin MS, Vukmer T. Granular cell variant of epithelioid cell histiocytoma. *Am J Dermatopathol.* 2012;34:766-769.
- Glusac EJ, Barr RJ, Everett MA, et al. Epithelioid cell histiocytoma. a report of 10 cases including a new cellular variant. *Am J Surg Pathol.* 1994;18:583-590.
- Creytens D, Ferdinande L, Van Dorpe J. ALK Rearrangement and overexpression in an unusual cutaneous epithelioid tumor with a peculiar whorled "perineurioma-like" growth pattern: epithelioid fibrous histiocytoma. *Appl Immunohistochem Mol Morphol.* 2017;25:E46-E48.
- Doyle LA, Fletcher CD. EMA positivity in epithelioid fibrous histiocytoma: a potential diagnostic pitfall. *J Cutan Pathol.* 2011;38:697-703.
- Fabrizi G, Massi G. Polypoid Spitz naevus: the benign counterpart of polypoid malignant melanoma. *Br J Dermatol.* 2000;142:128-132.
- Plaza JA, De Stefano D, Suster S, et al. Intradermal Spitz nevi: a rare subtype of Spitz nevi analyzed in a clinicopathologic study of 74 cases. *Am J Dermatopathol.* 2014;36:283-294; quiz 295-287.
- Menezes FD, Mooi WJ. Spitz tumors of the skin. *Surg Pathol Clin.* 2017;10:281-298.
- Requena C, Requena L, Kutzner H, et al. Spitz nevus: a clinicopathological study of 349 cases. *Am J Dermatopathol.* 2009;31:107-116.
- Mentzel T, Dei Tos AP, Sapi Z, et al. Myopericytoma of skin and soft tissues: clinicopathologic and immunohistochemical study of 54 cases. *Am J Surg Pathol.* 2006;30:104-113.
- Aung PP, Goldberg LJ, Mahalingam M, et al. Cutaneous myopericytoma: a report of 3 cases and review of the literature. *Dermatopathology (Basel).* 2015;2:9-14.
- Morzycki A, Joukhar N, Murphy A, et al. Digital myopericytoma: a case report and systematic literature review. *J Hand Microsurg.* 2017;9:32-36.
- LeBlanc RE, Taube J. Myofibroma, myopericytoma, myoepithelioma, and myofibroblastoma of skin and soft tissue. *Surg Pathol Clin.* 2011;4:745-759.
- Chisolm SS, Schulman JM, Fox LP. Adult xanthogranuloma, reticulohistiocytosis, and Rosai-Dorfman disease. *Dermatol Clin.* 2015;33:465-472; discussion 473.
- Miettinen M, Fetsch JF. Reticulohistiocytoma (solitary epithelioid histiocytoma): a clinicopathologic and immunohistochemical study of 44 cases. *Am J Surg Pathol.* 2006;30:521-528.
- Cohen PR, Lee RA. Adult-onset reticulohistiocytoma presenting as a solitary asymptomatic red knee nodule: report and review of clinical presentations and immunohistochemistry staining features of reticulohistiocytosis. *Dermatol Online J.* 2014. pii:doi\_21725.
- Soldano AC, Meehan SA. Cutaneous solitary fibrous tumor: a report of 2 cases and review of the literature. *Am J Dermatopathol.* 2008;30:54-58.
- Feasel P, Al-Ibraheemi A, Fritchie K, et al. Superficial solitary fibrous tumor: a series of 26 cases. *Am J Surg Pathol.* 2018;42:778-785.
- Thway K, Ng W, Noujaim J, et al. The current status of solitary fibrous tumor: diagnostic features, variants, and genetics. *Int J Surg Pathol.* 2016;24:281-292.
- Erdag G, Qureshi HS, Patterson JW, et al. Solitary fibrous tumors of the skin: a clinicopathologic study of 10 cases and review of the literature. *J Cutan Pathol.* 2007;34:844-850.