Test yourself: answer – deep benign fibrous histiocytoma

Kerry L. Thomas · Evita Henderson-Jackson ·
Jamie T. Caracciolo

Published online: 14 February 2015
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Discussion

An AP radiograph shows a radiodense, superficial soft tissue mass in the proximal medial thigh without mineralization or femoral involvement (Fig. 1). Contrast-enhanced CT (Fig. 2) and MRI (Fig. 3) demonstrate a well-defined, smoothly marginated superficial soft tissue mass above the investing muscular fascia with an intervening fat plane and avid contrast enhancement with increased vascularity. While mostly homogenous, the mass has internal necrosis and hemorrhage with central vascularity. There is no definite hypointense pseudocapsule or T2-hyperintense peritumoral edema on MRI. Clinically, there was focal skin ulceration and eschar at the site of bleeding, possibly due to skin stretching in the setting of rapid enlargement. Given concern for hemorrhage, excisional rather than percutaneous needle biopsy was performed with electrocautery dissection around the mass to the underlying fascia, which was not involved.

Histopathology demonstrates a well-circumscribed spindle cell neoplasm without fibrous capsule/pseudocapsule exhibiting storiform architecture with focally striking branching hemangiopericytoma-like vessels arising in the subcutis and deep dermis (Fig. 4); focal ulceration of epidermis without epidermal tumor involvement. Spindle cells are

evenly distributed without pleomorphism, atypia, foamy histiocytes, or giant cells; the necrosis present is likely due to central infarction. Specimen margins were negative for tumor. Ischemic-like necrosis was noted at 20× and bland spindle cells without nuclear pleomorphism or hyperchromasia and rare mitoses at 40×.

Benign fibrous histiocytoma (BFH) is a superficial (relative to the investing fascia of the compartmental musculature) soft tissue neoplasm comprised of bland fibroblasts arranged in a storiform pattern with variable giant cells and foamy histiocytes [1, 2]. *Cutaneous* BFH (aka dermatofibroma or dermal histiocytoma) arises in the *dermis* with several histologic variants described [1, 2], while *deep* BFH arises in the *subcutis* and may histologically resemble cellular-type cutaneous BFH, but the presence of diffuse storiform architecture allows distinction. Forty percent demonstrate hemangiopericytoma-like changes with staghorn-like thin-walled branching vessels. Deep BFH often stains positive for CD34, unlike cutaneous BFH [3, 4].

Cutaneous BFH typically presents as a small (<2 cm), tan-brown, firm, painless skin nodule in the extremities of young adults [2]. Deep BFH also commonly occurs in the extremities, but typically achieves larger size (0.5–25 cm; median 3 cm). Although benign, deep BFH carries increased risk for local recurrence, up to 22 % in one series, and rare metastases have been reported [4]. Sampling with percutaneous or excisional biopsy is needed when lesions are large or necrotic to distinguish BFH from superficial malignant neoplasms such as undifferentiated pleomorphic sarcoma (UPS), cutaneous leiomyosarcoma (LMS), and dermatofibrosarcoma protuberans (DFSP). Pathologically, UPS may be differentiated from BFH by the presence of marked pleomorphism and numerous atypical mitoses. Cutaneous LMS are predominantly well-differentiated smooth muscle tumors exhibiting intersecting fascicular architecture. Like BFH, DFSP displays

The case presentation can be found at doi: [10.1007/s00256-015-2112-x](https://doi.org/10.1007/s00256-015-2112-x)

K. L. Thomas · J. T. Caracciolo (✉)
Department of Diagnostic Imaging, Moffitt Cancer Center,
Tampa, FL, USA
e-mail: jamie.caracciolo@moffitt.org

E. Henderson-Jackson
Department of Anatomic Pathology, Moffitt Cancer Center,
Tampa, FL, USA

a uniform cellular population with storiform architecture, but DFSP lacks overlying epidermal hyperplasia and typically demonstrates highly infiltrative margins [5].

A large retrospective analysis performed at the Armed Forces Institute of Pathology showed that soft tissue tumors of the lower extremity in young adults (26–35 years) are more often benign than malignant. Over a 10-year period, 60 % (534/891) of lower extremity soft tissue tumors were benign with BFH being the most common benign neoplasm [6]. Other benign entities in this demographic and location include non-neoplastic etiologies like hematoma and abscess and neoplasms including nodular fasciitis, lipoma, neurofibroma, and hemangioma [7].

Patient age, clinical history, and physical examination significantly contribute to the preoperative differential diagnosis, while imaging findings including size, margins, and location help narrow the differential. Sarcomas are typically large heterogeneous deep-seated tumors arising below the investing fascia with pseudocapsule and peritumoral edema [7]. Conversely, benign tumors tend to be small homogeneous superficial lesions without pseudocapsule, reactive zone, or involvement of the investing fascia [7]. However, there is notable overlap in the presentation of benign and malignant diseases. For example, superficial sarcomas certainly occur, often involving the investing fascia; myxomas may demonstrate peritumoral edema; large deep BFH may have a pseudocapsule and internal necrosis; and cutaneous LMS and DFSP may be small, superficial, and well defined [5, 7–9]. With that said, caution in preoperative diagnosis remains paramount [10]. In this case, despite the large size and internal heterogeneity, several imaging features suggested benignity including age, location, smooth margins, absence of fascial involvement, and lack of peritumoral edema [5, 7–10]. When considering these

findings in light of common benign neoplasms of the lower extremity, a confident preoperative diagnosis may be presented which, in turn, may influence surgical approach.

Conflict of interest The authors declare that they have no conflicts of interest.

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