Pathologic Quiz Case

A 76-Year-Old Debilitated Woman With a Right Thigh Mass

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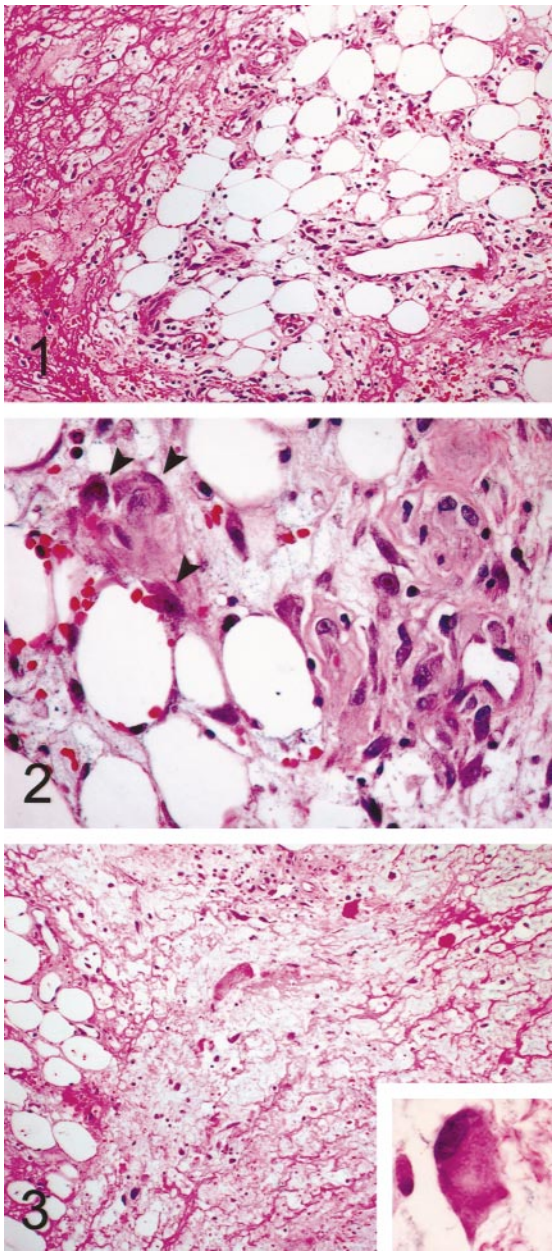
A 76-year-old white woman with multiple long-standing medical problems was noted to have an ill-defined mass in the deep soft tissues of her right upper lateral thigh. The patient had been a nursing home resident for many years and was essentially bedridden during that period of time because of severe arthritic disease. The mass was painless and had been present for an undetermined amount of time; however, during a previous physical examination 3 months earlier, the lesion had not been detected. On physical examination, the mass was located over the greater trochanter of the right femur and was mobile and soft to rubbery in consistency. The overlying skin appeared slightly erythematous, yet skin erosion or ulceration was absent. An excisional biopsy was performed.

Gross examination showed a 4.0 × 2.5 × 2.4-cm, yellow-tan mass involving the subcutaneous adipose tissue and fascia. The mass was poorly circumscribed and vaguely multinodular with mottled tan-brown intermixed with focal areas of gray-white fibrous tissue. Microscopically, broad areas of deeply eosinophilic paucicellular fibrinoid necrosis were seen (Figure 1, top right), interfacing with more vascularized cellular areas and intact adipose tissue (Figure 1, right). These zones of interface were sharply defined, with the more cellular areas containing variably sized spindled cells intermixed with occasional polymorphonuclear leukocytes, lymphocytes, and rare plasma cells. The spindle cells often appeared enlarged and atypical, with abundant basophilic cytoplasm, hyperchromatic smudged nuclei, and prominent viral inclusion–like nucleoli (Figure 2, arrowheads). The vascular component often resembled granulation tissue; however, the lining endothelial cells occasionally appeared enlarged, hyperchromatic, and epithelioid. Focal zones of abundant myxoid change were seen primarily involving the areas of fibrinoid necrosis (Figure 3). Within these areas, single cells were found floating within the myxoid substance and, not uncommonly, these cells resembled ganglion cells (Figure 3, inset). The margins of excision were involved by the process.

Immunohistochemical studies showed that the atypical spindle cell component strongly and diffusely expressed vimentin and CD68, with focal expression of muscle-specific actin and CD34. Stains for cytokeratin, S100 protein, HMB-45, and leukocyte common antigen (CD45) were negative.

What is your diagnosis?



Accepted for publication May 10, 2004.

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The authors have no relevant financial interest in the products or companies described in this article.

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Pathologic Diagnosis: Ischemic Fasciitis (Atypical Decubital Fibroplasia)

The term *ischemic fasciitis* (IF) is used to describe a juxtaarticular pseudosarcomatous fibroblastic proliferation that occurs predominately in immobilized or debilitated elderly patients. Montgomery et al[1] first described IF in 1992 under the synonymous term *atypical decubital fibroplasia* (ADF), and to date their report has been the largest published series, consisting of 28 patients. In these patients (12 men, 16 women), the lesion was characterized as a painless, ill-defined, focally myxoid mass that ranged from 1 to 8 cm. Patient ages ranged from 15 to 95 years, with a peak age of incidence in the eighth and ninth decades.

Histologically, the lesion tends to involve the underlying subcutaneous adipose tissue and fascia with occasional extension into the dermis. The lesion primarily demonstrates a zonal pattern, with central hypocellular fibrinoid necrosis surrounded by more cellular areas containing prominent proliferating neovessels and fibroblasts resembling exuberant granulation tissue. Focal areas of myxoid change are often found; multivacuolated muciphages mimicking lipoblasts also rarely can be seen within these myxoid zones. One of the hallmarks of the lesion is the presence of proliferating atypical fibroblasts within the more cellular peripheral regions. Cytologically, these fibroblasts may be markedly atypical with significant nuclear enlargement, hyperchromatic smudged (''degenerated'') chromatin, prominent macronucleoli, and abundant basophilic cytoplasm. These cells are often seen in close proximity to the proliferating vasculature and may be seen to merge imperceptibly with the adventitial walls of the blood vessels. Fibrin thrombi can frequently be seen occluding peripheral vessels. Mitotic figures often are identified, ranging from numerous to extremely rare. Unusual findings include ganglion-like fibroblastic cells resembling those seen in proliferative fasciitis/myositis, multinucleated osteoclast-like cells, numerous elastic fibers, and chondroid metaplasia.[2]

The immunohistochemical characterization of these lesions is limited, but the atypical fibroblastic cells have been noted to consistently express vimentin with frequent focal expression of muscle-specific actin and CD68.[1,3] Not uncommonly, staining for CD34 is found, which has been postulated to indicate early endothelial differentiation.[4]

In approximately one third to one half of reported cases, a malignant histologic diagnosis was considered.[1–3,5,6] The major differential diagnoses include other benign reactive conditions, such as nodular fasciitis, proliferative fasciitis, and proliferative myositis, as well as malignant lesions, such as epithelioid sarcoma, myxoid liposarcoma, and myxoid malignant fibrous histiocytoma. Although epithelioid sarcoma often manifests a histologic picture of cellular regions punctuated by zones of necrosis reminiscent of IF, it primarily arises in adolescents and young adults, and it has a propensity for the distal extremities. In addition, the cellular constituent of epithelioid sarcoma consists of nodules of deeply eosinophilic epithelioid cells that stain strongly for cytokeratin. The muciphages occasionally identified in IF may lead to confusion of these cells as lipoblasts. When these cells are seen in the background of a myxoid matrix, a misdiagnosis of myxoid liposarcoma may occur; however, unlike myxoid liposarcoma, IF lacks the delicate plexiform vasculature characteristic of this entity. Myxoid malignant fibrous histiocytoma lacks the zonal architecture seen in IF. The neoplastic cells of myxoid malignant fibrous histiocytoma also fail to show the degenerative smudgy chromatin of the atypical reactive fibroblasts, the fibrin thrombi, and the fibrinoid necrosis of IF.

With the ever-increasing use of fine-needle aspiration biopsy as a diagnostic tool, keeping IF/ADF in mind is paramount in order to avoid mislabeling this benign reactive proliferation as a high-grade malignancy. Unfortunately, fine-needle aspiration findings often are interpreted as suspicious or strongly suggestive of malignancy; therefore, conservative caution in evaluating fine-needle aspiration specimens from patients with a characteristic clinical history as described in this article is essential to avoid misdiagnosis.[6] Only rarely will the clinical history deviate from the prototypical one, but the pathologist should be aware that rare cases of IF/ADF have been described in young patients.[1,7]

The pathogenesis of this entity has been speculated upon, but most authors agree that the lesion represents a variant of decubitus ulcer that primarily involves the deep soft tissues.[1,3,5] Perosio and Weiss[5] postulated that the lesion arises from local ischemia to an area due to prolonged vascular compression from underlying bony prominences. Unlike cutaneous decubital ulcerations, the overlying skin in IF/ADF is usually intact, and this may be due to the fact that the ischemia is likely of a less severe or intermittent nature, so that the development of overlying cutaneous ulceration does not occur.

The prognosis of IF/ADF is universally excellent, and no instances of malignant transformation, metastasis, or death have been reported. In general, conservative local excision has been accepted as the treatment of choice. Of the approximately 40 published cases, 4 patients developed local recurrence, with 1 patient developing 2 recurrences.[1–3,5–7]

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