Pathologic Quiz Case

A Large Recurrent Thigh Mass in a 79-Year-Old Woman

Michael O. Lovell, MD; Ronald P. Williams, MD, PhD; Josefine Heim-Hall, MD

A 79-year-old obese woman presented with a large soft tissue mass that markedly distended her right thigh. Her past medical history was significant for the removal of a 1.1-kg, 21-cm (in greatest diameter), deep soft tissue mass from the same area approximately 7 years earlier. The original mass had been present for about 18 months and was diagnosed as an atypical lipoma at another facility.

Magnetic resonance imaging of the recurrent tumor revealed a large, lobulated heterogeneous soft tissue mass of the right thigh. The majority of the mass displayed uniform fat density, but one eccentric region displayed a dense, heterogeneous appearance. After incisional biopsy established the diagnosis, the patient underwent preoperative radiation therapy followed by a limb-sparing procedure.

A 4.8-kg multilobulated yellow soft tissue mass that was 34 cm in greatest dimension was received fresh. The majority of the tumor lobules were soft, yellow, and homogeneous, but one lobule was 8.5 cm in diameter and firm, tan-white, and fibrous in consistency (Figure 1).

Histologically, the firm, tan-white area corresponded to nodules of inflammatory cells separated by broad bands of fibrous tissue (Figure 2). The inflammatory cells consisted predominantly of lymphocytes and plasma cells. Scattered large, atypical cells with hyperchromatic, lobated pleomorphic nuclei were seen in both the fibrous and inflammatory areas (Figure 3). Rare lipoblasts were also identified (Figure 4). The remainder of the tumor consisted of well-differentiated adipocytes separated by fibrous septa. Rare large, atypical cells were also identified in these areas.

Postoperatively, the patient had no significant complications. She is without evidence of recurrence or metastases 12 months postoperatively.

What is your diagnosis?

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From the Departments of Pathology (Drs Lovell and Heim-Hall) and Orthopedic Surgery (Dr Williams), University of Texas Health Science Center at San Antonio, San Antonio, Tex.

Corresponding author: Josefine Heim-Hall, MD, Department of Pathology, University of Texas Health Science Center at San Antonio, 7703 Floyd Curl Dr, MC 7750, San Antonio, TX 78229-3900 (e-mail: heim@uthscsa.edu).

Pathologic Diagnosis: Inflammatory Liposarcoma Occurring as a Component of a Recurrent, Well-Differentiated, Lipoma-like Liposarcoma

Liposarcoma is one of the most common soft tissue sarcomas of adults. Liposarcomas consist of a heterogeneous group of tumors sharing lipogenic differentiation as a common factor. This group of tumors is remarkable not only for their potential to attain great size but also for their marked histologic and biologic variability. Histologically, liposarcomas display a spectrum of appearances, ranging from very well-differentiated tumors that closely resemble normal adipose tissue to high-grade pleomorphic tumors in which adipose differentiation is not apparent.

Inflammatory liposarcoma is a recently recognized rare variant of liposarcoma in which the lipomatous nature of the lesion may not be apparent.[1–3] The differential diagnosis of this lesion includes a number of neoplastic and nonneoplastic lesions such as Hodgkin and non-Hodgkin lymphoma, Castleman disease, and inflammatory myofibroblastic tumor (inflammatory pseudotumor).[1–3] The correct diagnosis can be made by recognizing a component of more typical well-differentiated liposarcoma within the same lesion, most often of the lipoma-like or sclerosing subtypes.[2] Scattered large, atypical cells with lobated or multiple nuclei are also typically present. The lymphoplasmacytic infiltrate in this tumor has been shown to be benign and reactive in nature.[2]

The majority of the cases in the 2 series reported to date have occurred in the retroperitoneum,[1,2] with only 1 reported case occurring in the thigh.[2] Of 3 additional more recently reported cases, 1 was retroperitoneal,[4] 1 was subdiaphragmatic,[5] and 1 occurred in supraclavicular subcutaneous soft tissue.[6]

The combination of different subtypes of well-differentiated liposarcoma within the same tumor is commonly observed, and recurrences may contain different combinations of these subtypes. The recurrence of a lipoma-like liposarcoma as an inflammatory liposarcoma, as in this case, is therefore not surprising, although this subtype is still considered relatively rare.

The literature on this entity is limited, and the first detailed description appeared under the name of ''lymphocyte-rich well-differentiated liposarcoma'' in 1997.[2] The term ''well-differentiated inflammatory liposarcoma'' was used in a subsequent article by Kraus et al.[1] Reference is made to the potential for confusion when using the term ''inflammatory liposarcoma,'' because it has been used in the past to refer to cases of pleomorphic liposarcoma with large numbers of neutrophils.[7] Because pleomorphic liposarcoma is considered a high-grade sarcoma, distinction of these entities is important. To avoid confusion, it has been suggested that the term inflammatory liposarcoma should be reserved only for those cases that display characteristic features of nodular aggregates of chronic inflammatory cells within a paucicellular stroma with scattered large atypical cells.[1]

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