

A RARE FATTY SOFT TISSUE TUMOR: A CASE REPORT AND DISCUSSION OF THE LITERATURE

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HISTORY AND PHYSICAL EXAM

A 65-year-old woman presented with a history of a sarcoma in the left pretibial area removed in 1983 with post-operative radiation. She has no evidence of recurrent sarcoma since that time. She recently lost 25 pounds on purpose and felt a mass along the lateral aspect of her leg. She was evaluated with an MRI, bone scan and PET scan, which incidentally revealed a soft tissue mass in the thigh.

On examination, she walked with a normal gait and had full range of motion of her hips and knees. She has a well-healed transverse incision over the anterior aspect of her right knee at the level of the patellar tendon with atrophic skin consistent with prior radiation. There were several firm small nodularities associated with the underlying soft tissues, which were felt to be dystrophic calcifications. She had a large mass of her posterior thigh measuring about 19 cm in length that was well circumscribed, soft and mobile. She had no inguinal adenopathy. There were no motor or sensory deficits.

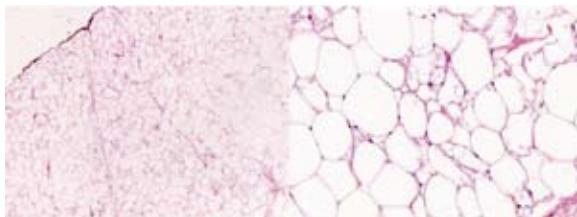


Figure 1: CT portion of PET-CT shows a large low density soft tissue mass in hamstrings in the left thigh with linear septa.

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IMAGING STUDIES

PET CT showed a mass of her right distal thigh (Figure 1). An MRI showed a mass measuring 8 x 14 x 16 cm within the posterior soft tissue of the mid to distal thigh between the biceps and semitendinosus. The lesion showed heterogeneously high signal intensity on T1 with low intensity septa. The mass was low intensity on STIR and demonstrated peripheral enhancement with contrast (Figure 2).

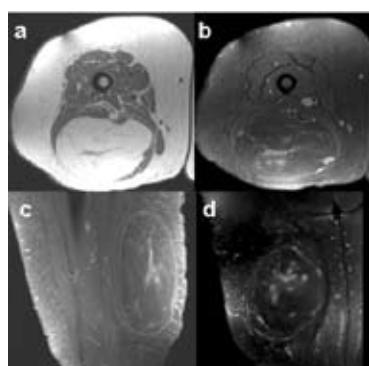


Figure 2: MRI shows a mass measuring 8 x 14 x 16 cm within the posterior soft tissue of the mid to distal thigh between the biceps and semitendinosus. (a) Axial T1 weighted image. The lesion is heterogeneously high signal intensity on T1 with low intensity septa. (b) Axial images with contrast which demonstrate peripheral enhancement. (c) Sagittal images with contrast. (d) Coronal T2 weighted image

DIFFERENTIAL DIAGNOSIS

Lipoma

Atypical Lipoma (low-grade liposarcoma)

INTRAOPERATIVE FINDINGS

There was a large intramuscular fatty tumor in the posterior thigh, infiltrating into the biceps and the medial hamstrings. Some muscle fibers were resected as a medial and lateral margin since it appeared to infiltrate. It was posterior to the sciatic nerve, which was identified and preserved. The mass was completely excised. Photomicrographs are shown.

HISTOLOGY

The mass measuring 16 x 17.5 x 7.3 cm and was yellow, soft and encapsulated. The specimen was serially sectioned to reveal tan solid yellow fatty cut surfaces. No areas of hemorrhage or necrosis were seen. Microscopically, the tumor was composed of a mixture of brown and white adipose cells. The brown adipose cells were characterized by polygonal multivacuolated cells, centrally located and had prominent nuclei (Figure 3).

DIAGNOSIS

Hibernoma

DISCUSSION

Hibernoma is an uncommon, slow-growing, benign soft tissue tumor resembling brown adipose tissue presenting in the fourth and fifth decades with a slight female predominance.



Figure 3: Hibernoma showing a mixture of brown and white adipose cells (H&E stain) right: x 40, left: x 200. The brown adipose cells are characterized by polygonal multivacuolated cells.

The most common anatomic locations include the thigh, shoulder, back, neck, chest, arm, abdominal cavity and retroperitoneum¹. The name "hibernoma" was derived from the tumor's histological similarity to the brown fat in hibernating animals². Brown adipocytes contain a very large number of mitochondria and are innervated by the sympathetic nervous system³. Brown adipose tissue, the function of which is to promote nonshivering thermogenesis, is present in the fetus and gradually is replaced by white adipose tissue with advancing postnatal age. It persists, however, in varying amounts throughout adult life and may be found in the neck, axilla, mediastinum, and periaortica and perirenal zones⁴.

Clinically, hibernoma is a painless, slow-growing mass, freely movable and usually nontender⁵. The only symptoms they cause are from mass effect upon adjacent structures. Since the tumor is hypervascular, the overlying skin may be warm⁶.

Radiographically, hibernoma has characteristic, but not specific imaging features. The MR appearance of hibernoma may vary according to the histological composition, relative proportion of white and brown fat. These lesions are often intramuscular and tend to insinuate between the fascial planes and wrap around adjacent structures. On T1 weighted images, hibernoma demonstrates increased signal intensity and are slightly hypointense to subcutaneous fat. On T2 weighted sequences with fat suppression, the signal of the mass is equal to or higher than that of subcutaneous fat and demonstrates partial signal loss consistent with fatty elements. With intravenous contrast, hibernoma typically demonstrates diffuse enhancement. Hibernoma is well defined, heterogeneous in signal intensity and contains multiple low-signal bands, which are due to fibrovascular proliferations⁷⁻¹⁰. Doppler ultrasound has demonstrated a highly vascular tumor with large feeding surface vessels⁷. Some reports suggest that increased T1 signal intensity and large intratumor vessels may indicate a diagnosis of a hibernoma^{7, 11}. PET (Positron emission tomography) can demonstrate increased uptake due to high metabolic activity of brown fat^{12, 13}.

Grossly, hibernoma is a well circumscribed, lobulated and partly encapsulated yellow-tan mass. Histologically, the classic description is one of a mixture of brown and white adipose cells. Four histologic variants of hibernomas have been described: typical, myxoid, lipomal-like, and spindle cell¹. All tumors are composed partly or principally of coarsely multivacuolated fat cells with small, central nuclei and no atypia. Unlike classical lipomas that are composed of large fat cells with a single cytoplasmic fat vacuole and a peripheral nucleus, hibernomas have a variable histological appearance depending on the relative amounts of multivacuolated, mitochondria-rich brown fat cells associated with capillary proliferation and fibrovascular septa. The histological diagnosis is usually straightforward.

The differential diagnosis of a complex fatty mass on MRI is wide and includes benign conditions, such as lipoma, atypical lipoma, alveolar soft part sarcoma and clear cell sarcoma and hemangioma, as well as malignant lesion such as liposarcoma. The MRI features of hibernoma are not diagnostic and may mimic by lipoma variants and by atypical lipoma and are often indistinguishable. High-grade liposarcomas are rarely confused with lipomas or hibernomas as they contain little or no fatty elements. MR signal characteristics vary with the cellular composition and vascularity; however, the presence of thin tortuous vessels in some cases may provide a clue to the diagnosis of hibernoma^{11, 14}.

The treatment is marginal excision, which is curative. Hibernomas have generally been regarded as benign with no propensity to recur after resection and no potential distant metastasis⁵. Incomplete excision of hibernoma has been reported to lead to regrowth of the tumor¹⁵. Therefore, complete local excision is deemed the treatment of choice in most cases.

In our case, MRI had some features characteristic of hibernoma with low intensity structures in the tumor on T1; however, it was indistinguishable from atypical lipoma¹⁴. PET was grossly negative in this case (images not shown). In addition, hibernoma is sufficiently rare that our initial differential diagnoses included only lipoma and atypical lipoma. The diagnosis of hibernoma did not change our treatment plan although; one could consider the option of preoperative CT guided needle biopsy. Of note, high-grade liposarcomas are rarely confused with hibernomas as they contain little or no fatty elements.

In conclusion, hibernomas are rare fatty tumors that can arise at unusual sites. Hibernoma has characteristic, but non-specific imaging features of a well-defined, heterogeneous fatty mass with large vessels. On T1 weighted MR images, hibernoma demonstrates increased signal intensity that is similar, and may be lower than subcutaneous fat. The treatment is marginal excision, and hibernoma has a low potential of local recurrence. Hibernoma should be included in the differential diagnosis of a fatty mass on MR images.

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