Uterine bizarre epithelioid lipoleiomyoma with a myxoid stroma

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Summary

The first case of uterine bizarre epithelioid lipoleiomyoma with a myxoid component occurring in an 86-year-old woman is described. An intramural 22 cm mass in the anterior wall of the uterine body had a lipoma-like appearance with strands of fibrous tissue. Histologically, the tumor consisted of adipocytes which varied in size and shape, and epithelioid smooth muscle cells with nuclear atypia within a myxoid stroma. No mitotic features were noted despite an extensive search. The patient was well without disease 24 months after hysterectomy. Patients with this type of tumor need close and long-term follow-up because of the paucity of clinical information.

Key words:

Introduction

Uterine leimyomas contain various degrees of degeneration and/or exhibit a variety of unusual histologic subtypes including epithelioid leiomyoma, bizarre leiomyoma, myxoid leiomyoma and lipoleimyoma. We present the first reported case of the rare occurrence of bizarre epithelioid lipoleiomyoma with a myxoid component in the uterine body, focusing on the importance of its unusual morphological features, and discuss the differential diagnosis with the malignant counterparts.

Case Report

An 86-year-old woman presented with a two-month history of abdominal distention. Laboratory data and tumor markers including CA125, CA19-9, CEA and lactate dehydrogenase were within normal limits. On post-contrast computed tomography (CT), the tumor was heterogeneously enhanced with a well demarcated margin, and fat density was depicted in the mass lesion. The patient underwent laparotomy. At surgery, an enlarged uterus weighing 3300 g with an intramural mass measuring 20 x 14 x 11 cm in the anterior wall of the uterus was found. Total hysterectomy and bilateral salpingo-oophorectomy were performed. The postoperative course was uneventful. The patient is alive and well with no recurrences 24 months after surgery.

On pathologic examination, the cut surface of the mass had a mixture of whitish areas together with prominent yellow areas. There was no necrosis or hemorrhage (Figure 1). Microscopic examination showed a histologic variety characterized by a mixture of adipose tissue in places with strands of fibrous tissue and adjoining smooth muscle cells (Figure 2). The adipose tissue was predominantly composed of lipocytes with variations in size and shape. Neither obvious lipoblasts nor nuclear mitoses were found. The fibrous areas contained a proliferation of smooth muscle cells within a myxoid stroma. The smooth muscle cells showed bizarre or atypical nuclei and eosinophilic cytoplasm, but tumor cell necrosis or hemorrhagic areas were absent, and the mitotic rate was low (0-1 mitosis/10 high power fields) (Figure 3). The endometrium showed atrophic changes and the cervix was normal. Immunohistochemically, the lipocytes were strongly positive for S-100 protein, and did not stain for myogenic markers such as α-smooth muscle actin and desmin, and HHF35. The smooth muscle cells showed strong cytoplasmic staining of α-smooth muscle actin, HHF35 and desmin, and were negative for S-100 protein. The acellular matrix stained strongly with Alcian blue at pH 2.5 (digested by hyaluronidase) and negative for PAS, indicating that it consisted of myxoid material.

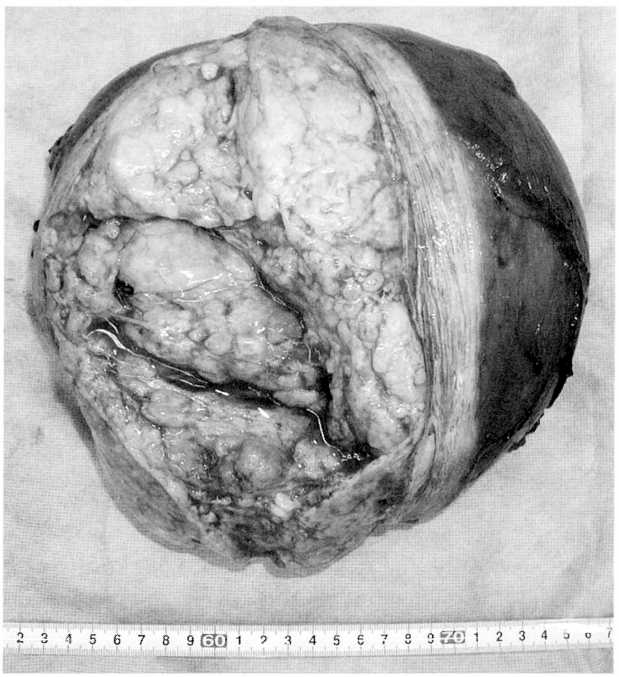

Figure 1. — The cut surface of the mass shows a mixture of whitish areas together with prominent yellow areas. No necrosis or hemorrhage is noted.

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Fig. 2

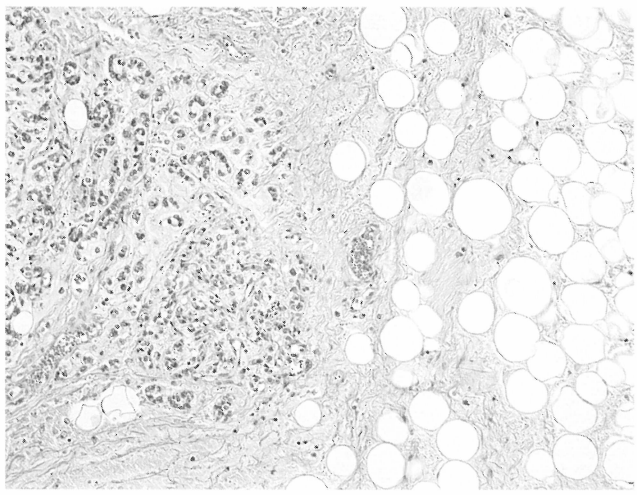

Fig.

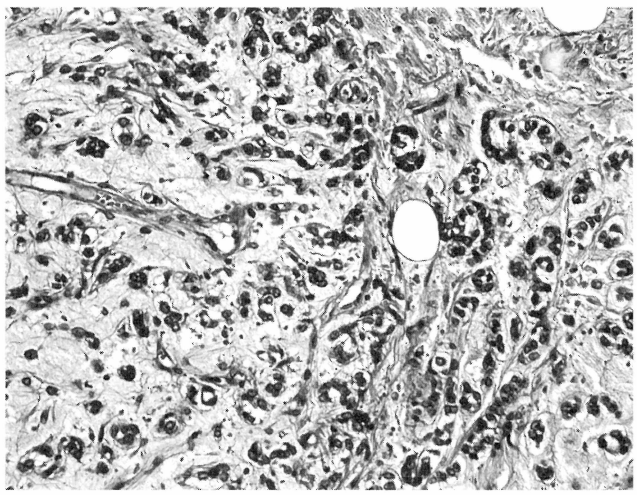

Figure 2. — Tumor shows a histologic variety characterized by a mixture of lipocytes with variations in size and shape in places with strands of fibrous tissue, and adjoining epithelioid smooth muscle cells with rounded cells and nuclear variation (H&E).
Figure 3. — The smooth muscle cells show bizarre or atypical nuclei with low mitotic rate in a prominent myxoid background (H&E).

Discussion

Benign lipomatous lesions of the uterus are uncommon. Such tumors can occur as pure lipomas or as lipoleiomyomas. The fat cells are usually mature as in our case, but occasional lipoblasts can be found. On rare occasions, smooth muscle cells are polygonal and they may grow in sheets or cords instead of fascicles and bundles. The term "epithelioid smooth muscle tumors" is used to designate smooth muscle tumors which show substantial epithelial differentiation. Myxoid leiomyomas of the uterus are uncommon. They are characterized by a myxoid stroma and myogenic phenotype without mitotic activity and infiltrative growth patterns. Regarding the combination of these entities, there have been only two reports of bizarre lipoleimyoma [1, 2] and one report of bizarre epithelioid lipoleiomyoma [3]. Lin and Hanai [1] reported a case of atypical lipoleiomyoma of the uterus without lipoblasts in a 55-year-old postmenopausal woman. Recently a case report described a 58-year-old postmenopausal woman who developed bizarre lipoleiomyoma with an atypical adipocytic component of the uterus [2]. Brooks *et al.* [3] reported a bizarre epithelioid lipoleiomyoma of the uterus in a 41-year-old premenopausal woman in which atypical smooth muscle cells and lipoblasts were present. Unlike previous reports of bizarre lipoleiomyomas, the present case is characterized by the coexistence of adipocytes which varied in size and shape, and bizarre epithelioid smooth muscle cells coupled with a myxoid background.

Pathologically, the differential diagnosis of atypical smooth muscle tumors from their malignant counterpart is often difficult. The presence of either an epithelioid morphology or nuclear atypia may pose some difficulties in defining the malignant potential and in the differential diagnosis of a uterine smooth muscle tumor. Clinically malignant tumors (i.e., epithelioid leiomyosarcomas) typically have a combination of significant nuclear atypia (either grade 2 or grade 3 nuclei) and some mitotic activity (usually at least 3 to 4 MF/10 HPF); most also have tumor cell necrosis [4]. Presumably, the same would hold true for bizarre epithelioid lipoleiomyoma. So far, no recurrence has been documented in the reported cases of bizarre lipoleiomyoma with a limited follow-up. In the present case, despite nuclear pleomorphism the tumor was confined in the myometrial wall, and lacked mitotic activity and coagulative tumor necrosis, which is completely consistent with a benign course for our patient without disease 24 months after surgery. Finally, bizarre epithelioid lipoleiomyoma with myxoid change is a diagnostic challenge because of its heterogeneous appearance. Careful evaluation of the above described morphological features should enable the differential diagnosis to be made. However, patients with this type of tumor need a close and long-term follow-up because of the paucity of clinical information.

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