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CASE REPORT

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Myolipoma of the round ligament: report of a case with a review of the English literature

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Abstract Tumours consisting of a mixture of mature adipose and smooth muscle tissues, including those designated lipoleiomyomas, fibrolipoleiomyomas and myolipomas, are exceedingly rare, but most often occur in the uterine corpus. We describe here a case of such a tumour arising in the right round ligament of a 44-year-old woman. The tumour, which measured approximately 20×15×10 cm, was well encapsulated and did not involve the intrapelvic organs. Intricate mixtures of adult adipose tissue and bland smooth muscle exhibited no cellular atypia or nuclear mitotic figures, and there was little vascular proliferation. We diagnosed the lesion as a myolipoma of soft tissue with dual differentiation, and have found only 13 cases of this tumour including our own in the English literature. The present tumour is the first reported in the round ligament. Although this tumour is rare, its recognition is important for the avoidance of erroneous diagnoses.

Key words Myolipoma · Soft tissue · Round ligament · Immunohistochemistry

Introduction

Mixed mesenchymal tumours consisting of mature adipose and smooth muscle tissues include tumours designated lipoleiomyomas [8, 13, 16, 18, 21], fibrolipoleiomyomas [10], and myolipomas [14, 15]. These tumours are rare, but occur most often in the uterine corpus [2, 8, 13, 18, 22], and develop in extrauterine sites only in exceptional cases [8, 14–16, 21]. We describe here a case of such a tumour arising in the round ligament, which we diagnosed as a myolipoma with dual differentiation [14, 15] rather than leiomyoma with fatty metaplasia. In the English literature, only 12 tumours of this kind have been reported previously. The present tumour is the first developing in the round ligament.

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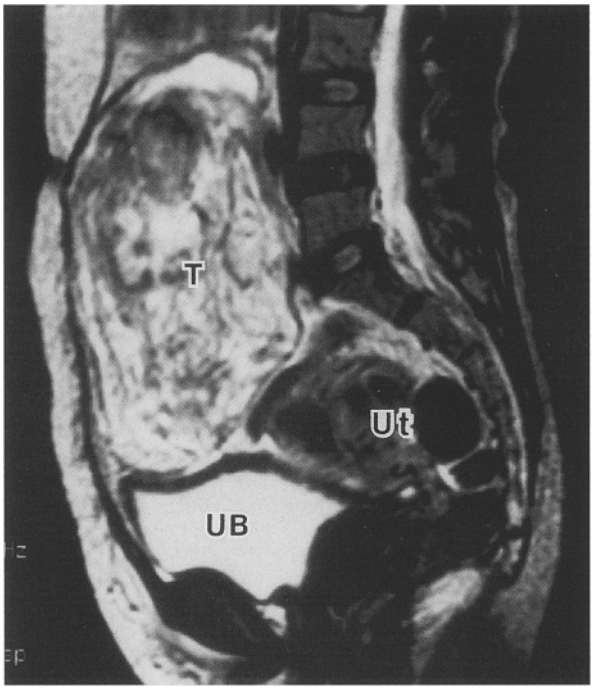

Fig. 1 Magnetic resonance computed tomography. The intraabdominal tumour (*T*) was demonstrated as a huge inhomogenous mass on T2-weighted imaging (*Ut* uterus, *UB* urinary bladder)

Clinical history

A 44-year-old Japanese woman, para 2, gravida 2, who was 156 cm in height and weighed 56 kg, presented for a medical checkup, although she was asymptomatic. A soft intraabdominal mass was palpable, and ultrasound sonography revealed a huge, non-homogeneous mass occupying the abdominal cavity. Magnetic resonance computed tomography with T1- and T2-weighted imaging demonstrated essentially the same features (Fig. 1), but there was no enhancement following gadolinium injection. On the

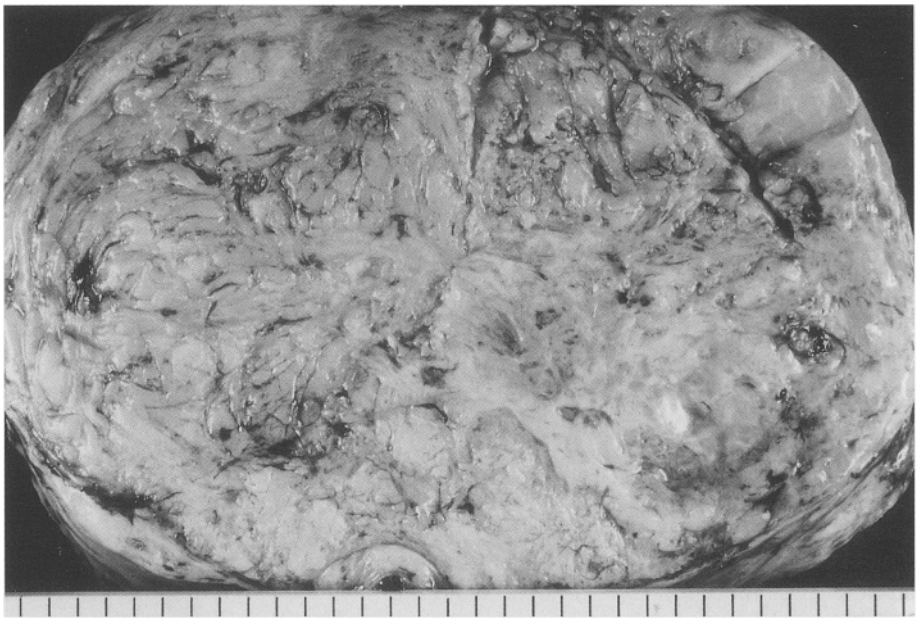

Fig. 2 Gross appearance of the extirpated tumour.The cut surface exhibits intricately mixed lipomatous and firm, whorled tissues throughout the lesion

basis of these findings a malignant tumour was suspected, although its site of origin was unclear.

On 7 May 1995, extirpation of the mass with total hysterectomy was performed. The well-circumscribed mass was found to arise from the right round ligament with no involvement of the surrounding organs, including the uterus. The patient is doing well 3 months after surgery, with no evidence of tumour recurrence or metastasis.

Materials and methods

For light-microscopy and immunohistochemistry, surgically resected tissues were fixed in 20% buffered formalin solution and embedded in paraffin using routine procedures. Dewaxed paraffin sections were stained with haematoxylin and eosin, Azan-Mallory, silver and Giemsa stains. Additional sections were stained with mouse monoclonal antibodies to alpha-smooth muscle actin (1A4, ×200; Dakopatts) and desmin (D33,×30; Dakopatts), and also rabbit polyclonal antibodies to S-100 protein (×1,000; Dakopatts) and factor VIII-related antigen (×400; Dakopatts), using the avidin-biotin immunoperoxidase complex (ABC) method (Histofine SAB-PO-Kit, Nichirei, Tokyo, Japan).

Pathologic findings

Grossly, the right round ligament mass, which measured approximately 20×15×10 cm, was elliptical in shape with a thick fibrous capsule. The maximum cut surface was solid, disclosing an intricate mixture of yellowish, lobulated adipose tissue and whitish, firm whorled tissue. The components were roughly equal in volume, and exhibited a rather regular, diffuse pattern of distribution throughout the lesion (Fig. 2). The uterus was unremarkable except for two firm nodules, measuring approximately 2 and 3 cm in diameter, within the myometrium.

Light microscopically, the mass consisted of two proliferating components: mature adipocytes forming a nest-like pattern, and bland smooth muscle cells with cigar-shaped nuclei and eosinophilic cytoplasm disclosing an interlacing fascicular pattern. The two components were intermingled with each other in varying proportions in a complicated fashion, and were accompanied by a stromal component of fibrovascular tissues (Fig. 3a). Neither nuclear atypia nor mitotic figures were observed. Many mast cells and eosinophils had infiltrated most of the smooth muscle tissues. On immunohistochemical examination, the adipocytes were found to be positive for S-100 protein (Fig. 3b), and the smooth muscle cells were positive for alpha-smooth muscle actin (Fig. 3c) and desmin. These findings led us to diagnose the lesion as a myolipoma of soft tissue of the round ligament. The two nodules in the uterus had features typical of leiomyoma but contained no adipose tissue.

Discussion

Certain rare tumours consisting of a mixture of mature adipose tissue and smooth muscle have been designated lipoleiomyomas [8, 13, 16, 18, 21], fibrolipoleiomyomas [10], mixed lipomas [2, 13], or myolipomas [14, 15]. They most frequently develop in the uterine corpus [2, 18, 22, 24], although they may in exceptional cases develop in the uterine cervix [8, 23], ovary [16], or soft tissue [14, 15, 21]. Uterine corpus tumours of this kind, usually called lipoleiomyomas, are in general considered to be leiomyomas with fatty metaplasia [18, 24], and are classified as a variant of leiomyoma in the WHO classification [19], despite the existence of other theories of their histogenesis, particularly of the adipose tissue component, which have included misplaced embryonic adipose cells [2], immature pluripotential cells [10], and lipoblasts migrating along uterine arteries or nerves [24]. In addition, unusual uterine lipoleiomyomatosis [2], leiomyosarcoma arising in uterine lipoleiomyoma [20] and

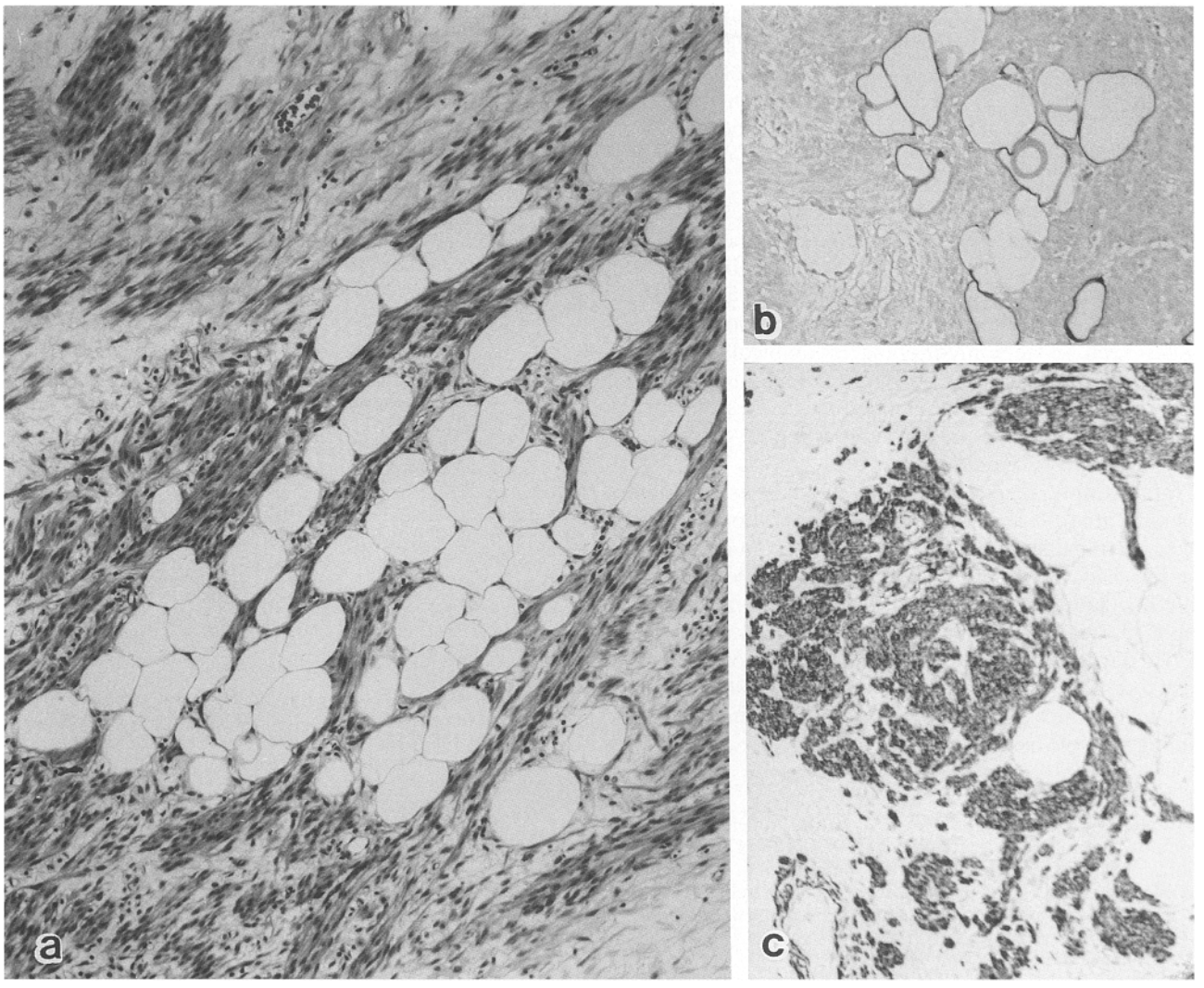

Fig. 3 a–c Light-microscopic features of the tumor. **a** Mature adipose cells and bland smooth muscle cells are intricately intermingled with each other (HE, ×200). **b** Adipose cells with S-100 protein-positive immunoreaction (ABC, ×200). **c** Smooth muscle cells with alpha-smooth muscle actin-positive immunoreaction (ABC, ×200)

lipoleiomyoma with bizarre epithelioid leiomyomatous components [4] have been described. These findings might support the histogenetical consideration that the adipose tissue is metaplastic in uterine lipoleiomyomas [18, 24].

Benign mesenchymal tumours are extremely rare in the round ligament, and include leiomyomas, adenomyomas, lipomas, and haemangiomas [5]. The histological features of the present tumour were similar to those of uterine lipoleiomyoma. However, except for two angiomyolipomas, no lipoleiomyomatous tumours arising in the round ligament have been described [17].

Meis and Enzinger [14] recently proposed that soft tissue tumours with mixed mature adipose and smooth muscle tissues should be called "myolipomas of soft tissue" and recognized as a new disease entity, and they considered, given the diffuse, regular pattern of distribution of the two tissues, that not only the smooth muscle but also the adipose tissue must be integral tumour components. Agreeing with this proposal, Michal [15] reported two similar retroperitoneal tumours as myolipomas and emphasized the presence of dilated non-neoplastic vascular structures in the tumours. Although the present lesion lacked this type of vascular pattern, it had essentially the same pattern of mixture of the two components as described in the series published by Meis and Enzinger [14]. We consider the features of this tumour to be consistent with myolipoma of soft tissue with dual differentiation.

Only 12 myolipomas of soft tissue are described in the English literature [14, 15, 21]. These lesions affect exclusively adults, and mostly women. They are completely or incompletely encapsulated, and are usually huge, with a greatest diameter larger than 10 cm. Apart from one in the subcutaneous tissue, the tumours occur in the deep soft tissues, and include six in the retroperitoneum, two in the abdominal wall, two in the inguinal canal, and one in the abdominal cavity. The present tumour is therefore the first to be found in the round ligament.

Myolipoma of soft tissue, which usually appears as a huge mass in the deep soft tissues including the retroperitoneum, may be confused with angiomyolipoma and well-differentiated liposarcoma [14, 15]. In order to avoid erroneous diagnoses, however, recognition of this rare soft tissue tumour is of great importance. In the present case, given the unusual extrauterine location of the tumour and its composition of adipocytes and spindle-cell components, it was also necessary to exclude angiomyolipoma [12] and well-differentiated liposarcoma [1, 6, 7] as well as spindle-cell lipoma [6]. The present tumour had none of the prominent proliferating vascular tissues seen in angiomyolipoma or atypical lipoblasts, which are observed in well-differentiated liposarcoma. In addition, spindle-cell lipoma exhibits features outwardly similar to those of the present tumour but no evidence of smooth muscle differentiation.

Interestingly, recent cytogenetical analyses have demonstrated that uterine lipoleiomyomas did not have only the same translocation t(12;14) as is detected in leiomyoma, but also abnormalities in chromosome 5q22 responsible for the lipomatous change, indicating a monoclonal origin [9, 11]. Further accumulation of cases and studies will be necessary for a detailed clarification of the nature of mixed lipomatous and leiomyomatous tumours arising both in the uterine corpus and in extrauterine sites, including the soft tissue.

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