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Extraperitoneal pelvic myolipoma

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ABSTRACT
Myolipoma is a very rare adipocytic tumor occurring most frequently in adults, and usually is located in the retroperitoneum or abdomen. It has been described in the retroperitoneum, spinal cord, orbita, breast, round ligament, subcutaneous tissue, pericardium, rectus sheath of the abdominal wall, and abdominal cavity with attachment to the abdominal wall. Most of these tumors are discovered incidentally and are large when discovered. Radiological findings are nonspecific due to the nonlipomatous component of the tumor. We present radiological findings of a large extraperitoneal pelvic myolipoma adjacent to the anterior abdominal wall, detected incidentally in an elderly woman with a presenting complaint of intractable hiccups.

Key words: • *myolipoma* • *abdomen* • *computed tomography*

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Myolipoma is a lipomatous soft tissue tumor, first described by Meis and Enzinger in 1991 (1). It is a rare tumor, and has been described in the retroperitoneum (1–3), spinal cord (4), orbita (5, 6), breast (7), round ligament (8), subcutaneous tissue (1), pericardium (9), rectus sheath of the abdominal wall, and abdominal cavity with attachment to the abdominal wall (1). Most cases are discovered incidentally and are large when discovered. Here, we present radiological findings of a large extraperitoneal pelvic myolipoma adjacent to the anterior abdominal wall, detected incidentally in an elderly woman with a presenting complaint of intractable hiccups.

Case report

An 83-year-old woman was admitted to our hospital with a complaint of hiccups for two months, unresponsive to medical therapy. She also had a feeling of fullness and pain in the midportion of her abdomen. Her past medical history revealed hypertension for 20 years, and bypass surgery 10 years ago. On physical examination, she was obese and had tenderness on the right and inferior part of the peri-umbilical region on deep palpation. Her laboratory studies were within normal limits.

Abdominal sonography showed a 10 x 5 x 5 cm mass composed of a central hyperechogenic area (which was the largest part of the tumor) and a peripheral hypoechogenic area (Fig. 1a). It was located in the pelvis anterior to the bowel, and anterosuperior to the urinary bladder. On color Doppler examination, the lesion did not show prominent vascularity (Fig. 1b).

In order to better characterize the lesion, we performed computed tomography (CT) and magnetic resonance imaging (MRI). CT images showed a smooth-contoured, encapsulated mass, containing fat densities in the central part of the lesion (-55 HU). A peripheral hypodense area was noted (Fig. 2a). On postcontrast images, the lesion showed heterogeneous enhancement centrally (Fig. 2b). On MRI examination, the lesion was hyperintense in T1-, and heterogeneous in T2-weighted images. On fat-suppressed T2-weighted images, the lesion showed significant signal loss. After administration of gadobenate dimeglumin, the lesion showed heterogeneous and peripheral enhancement (Fig. 3). We could not distinguish an intraperitoneal from an extraperitoneal location of the tumor by imaging. The differential diagnosis included benign and malignant fat-containing tumors such as lipoma, myolipoma, well differentiated liposarcoma, and teratoma, as well as inflammatory lesions such as an omental torsion.

The patient underwent surgery. During surgery, it was noted that the location of the tumor was superior to the urinary bladder in the extraperitoneal region. Macroscopically, the lesion was encapsulated. Some parts of the lesion were white, and some areas had a bright mucoid ap-

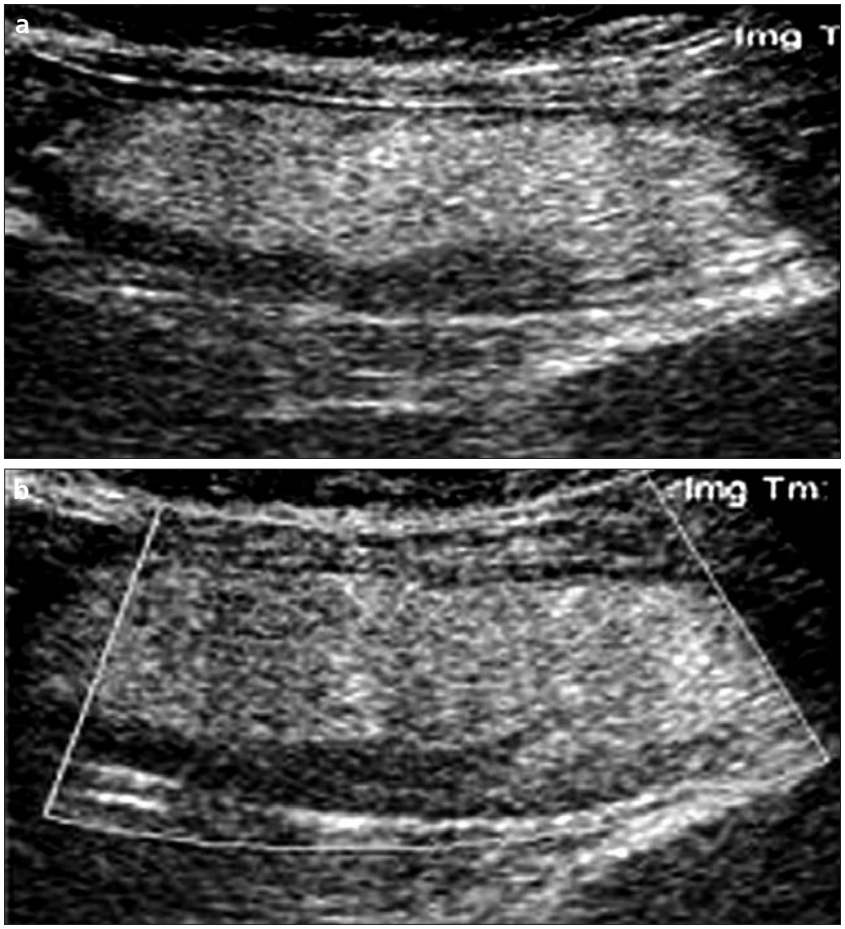

Figure 1. a, b. Abdominal sonography **(a)** shows a hyperechoic solid mass with a peripheral hypoechogenic area in the pelvis. On color Doppler examination **(b)** there was no prominent vascularity in the interior of the lesion.

pearance. In serial sections, fatty tissue was observed. Microscopically, the tumor was a mesenchymal lesion composed of smooth muscle cells, mature fatty tissue interspersed in loose connective tissue and, to a lesser extent, vascular structures. Mitoses, necrosis, and atypia were absent. In cut sections, a myxoid type of loose connective tissue was noted peripherally, close to the surgical margins (Fig. 4). Immunohistochemical examination was negative for HMB-45, which aided in excluding angiomyolipoma from the differential diagnosis. Accordingly, the overall diagnosis was myolipoma. After the operation, persistent hiccups did not resolve.

Discussion

Myolipoma, also called as lipoleiomyoma, is a very rare adipocytic tumor occurring most frequently in adults, and usually located in the retroperitoneum or abdomen. It is a large tumor composed of benign smooth muscle and mature adipose tissue. Histopathologically, the smooth muscle component of the tumor usually is regularly interspersed with the adipose tissue, creating a sievelike appearance (10). Retroperitoneal myolipoma is very rare and, to our knowledge, there have been only eight reported cases (1–3, 11–13). In retroperitoneal myolipoma, the main clinical presentation is abdominal distension or abdominal mass (Table). Our patient had intractable hiccups that did not respond to medical therapy.

In the literature, persistent hiccups are defined as those lasting longer than 48 hours, whereas intractable hiccups are those lasting longer than one month (14). Persistent or intractable hiccups may lead to fatigue, sleep disturbances, and dehydration. The exact mechanism of hiccups is not

Table. Clinicopathological data of retroperitoneal myolipoma reported in the literature

Case[a]	Age	Sex	Presentation	Radiological features	Size (cm)	Reference
1	53	F	Incidental	ND	17 x 7 x 5	1
2	63	F	Incidental	ND	20 x 17 x 11	1
7	55	F	Abdominal mass	MRI: Heterogenous mass, predominantly with fat intensity	30 x 15 x 8	2
8	34	F	Abdominal pain and distension	CT: Low density	ND	3
5	79	F	Fever, abdominal pain	CT: Heterogenous low density, mild enhancement, contains areas of calcifications	30 x 30 x 17	11
3	52	F	ND	ND	22 x 20 x 9	12
4	62	F	ND	ND	20 x 15 x 10	12
6	56	F	Abdominal distension	US: Heterogenous echoic mass, with multiple hyperechoic foci of variable size CT: Heterogenous mass, containing low-density areas	23 x 21 x 12	13

[a] Numbered in chronological order of their reporting in the literature
F, female; ND, no data; MRI, magnetic resonance imaging; CT, computed tomography; US, ultrasonograpy

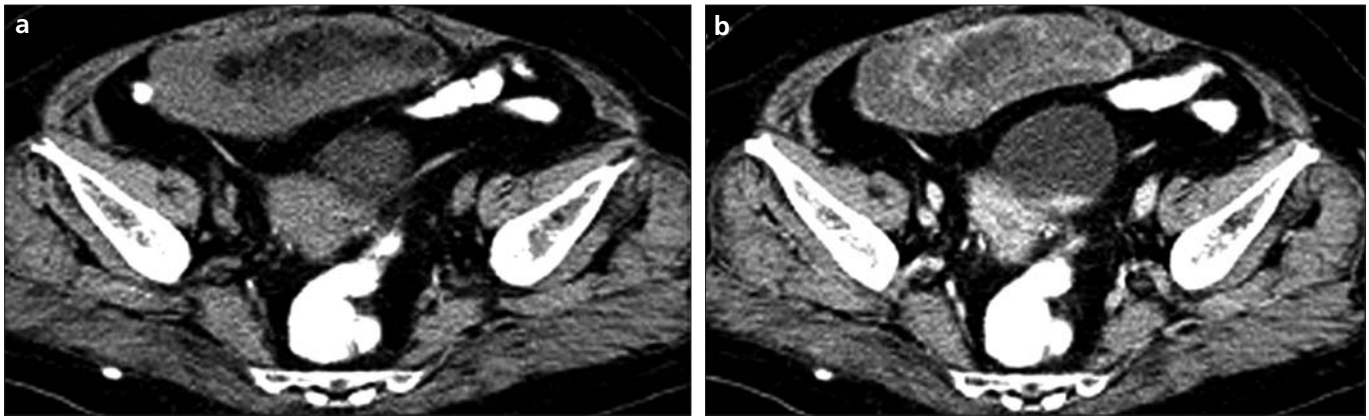

Figure 2. a, b. Precontrast CT image **(a)** demonstrates the mass, which contains fat densities, located anterosuperior to the urinary bladder. There was a hypodense area in the peripheral part of the lesion. On postcontrast CT image **(b),** the lesion showed heterogeneous central enhancement and capsule formation at the periphery.

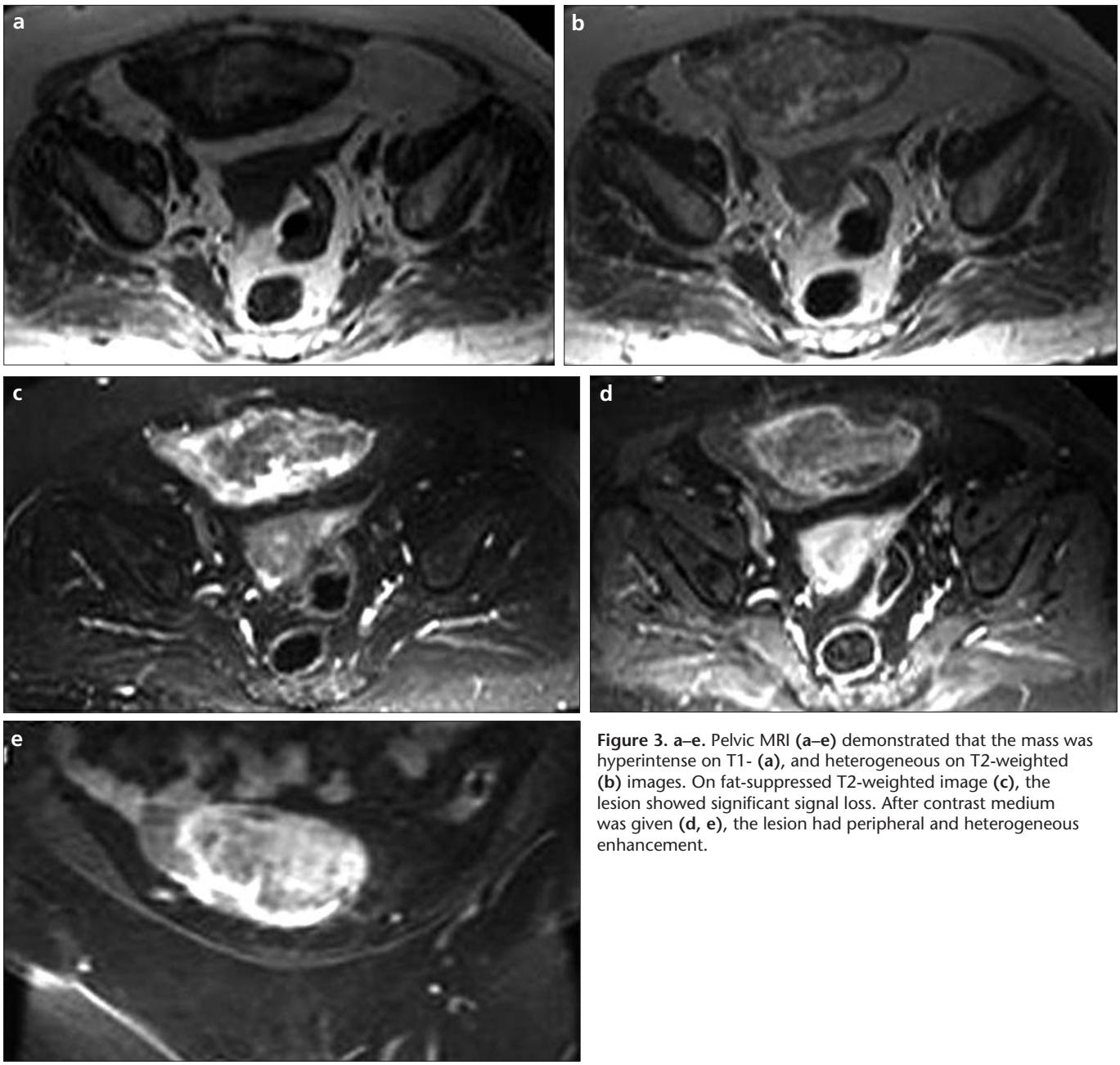

Figure 3. a–e. Pelvic MRI **(a–e)** demonstrated that the mass was hyperintense on T1- **(a)**, and heterogeneous on T2-weighted **(b)** images. On fat-suppressed T2-weighted image **(c)**, the lesion showed significant signal loss. After contrast medium was given **(d, e)**, the lesion had peripheral and heterogeneous enhancement.

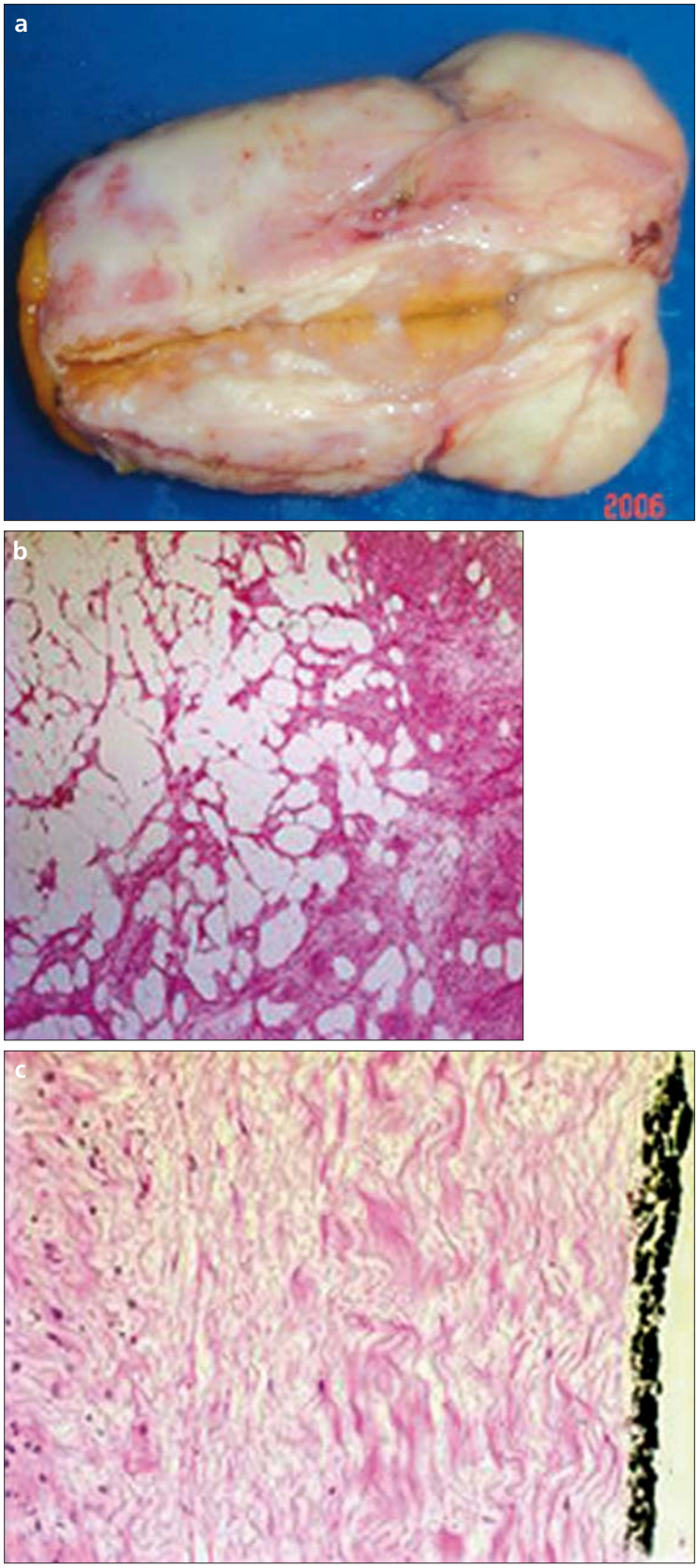

Figure 4. a–c. Gross pathological examination **(a)** showed that the lesion was encapsulated, and some parts of the lesion were white. Microscopically **(b, c)**, the tumor was a mesenchymal lesion composed of smooth muscle cells, mature fatty tissue interspersed in a marked loose connective tissue and, to a lesser extent, vascular structures **(b)**. In cut sections, a myxoid type of loose connective tissue was noted peripherally, close to the surgical margins **(c)**.

completely understood. Hiccups result from stimulation of the central or peripheral components of a hiccup reflex arc. Over 100 known organic causes of hiccups have been identified, and multiple etiologies have been proposed. The most common are of gastrointestinal origin, such as gastric distension. Metabolic derangements and drugs are also frequently implicated as causes for hiccups (14). In our patient, the hiccups were unrelated to the retroperitoneal mass and, therefore, did not resolve after the operation.

There are only a few reports concerning the radiological appearance of myolipoma (2, 3, 11, 13), listed in Table. Ultrasound findings are suggestive but not diagnostic, due to hyperechogenicity of the lipomatous component of the tumor. Lipomatous components of the tumor have specific CT and MRI features of fat tissue; however, the nonlipomatous component has nonspecific MR intrinsic features of the tumor, with intermediate signal intensity on T1-, and intermediate to high signal intensity on T2-weighted images. Coarse calcifications have been reported (11). In our case, there was also peripheral hypoechogenicity on ultrasonography, and hypodensity on CT, which represented the myxoid type of loose connective tissue on pathological examination.

The differential diagnosis of myolipoma includes lipoma, liposarcoma, teratoma, spindle cell lipoma, angiolipoma, leimyoma with fatty degeneration, omental infarction, and inflammatory lesions engulfing fat tissue (15). The most important entity in the differential diagnosis of pelvic myolipoma is retroperitoneal liposarcoma. Well differentiated and myxoid subtypes of liposarcoma can mimic myolipoma because of fat content. Internal thick fibrous septations of well differentiated liposarcomas have the same CT and MRI features as those of muscle, and may enhance after gadolinium administration (15). Histopathologically, myolipoma differs from the liposarcoma by the presence of encapsulation, and also by its non-invasiveness. Myolipoma also does not contain lipoblasts, atypical cells, or mitotic figures microscopically (2). Some lipomas with prominent fibrous septa and nodularity can mimic myolipoma and well differentiated liposarcoma (15). Retroperitoneal teratoma must be kept

in mind in the differential diagnosis of myolipoma because of fat content, calcifications, and soft tissue. Omental infarction usually occurs in the right lower or upper quadrants, and clinically resembles appendicitis or cholecystitis. CT of this lesion shows a large cake-like mass centered in the omentum. The inflammatory mass may or may not lie immediately adjacent to the colon, and is irregular in contour (15). In addition, omental infarction is painful, which is an important clue in the clinical differential diagnosis of fatty tumors. Angiolipoma is frequently associated with tuberous sclerosis. Histologically, it contains medium-sized arteries with a thick muscular wall. Immunohistochemical staining demonstrates positivity for melanoma markers (HMB-45) (2, 3). Spindle-cell lipoma is composed entirely of adipose tissue (2). In leiomyoma with fatty degeneration, adipose tissue is not an integral component of the lesion, and shows focal distribution, in contrast to the scattered distribution in myolipoma (3).

Myolipomas are cured by surgical resection (10). There are no reports of local recurrence, metastatic disease, or malignant transformation.

In conclusion, myolipoma is a rare tumor for which the radiological findings are nonspecific. It should be considered in the differential diagnosis of fat-containing lesions of the abdomen.

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