PRESACRAL MYELOLIPOMA

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Myelolipomas are benign tumors that are mainly composed of fatty tissue interspersed with extramedullary hematopoietic elements [1]. Myelolipomas are most often found in the adrenal gland but may rarely occur in extra-adrenal locations. Previous reports of extra-adrenal myelolipomas revealed that more than half of these cases occurred in the presacral region [2]. Like most retroperitoneal tumors, symptoms do not develop until effects related to the mass occur. In this article, we discuss the pathology, morphologic differential diagnosis, and appropriate management of extra-adrenal myelolipoma to bring this condition to the attention of gynecologists.

A 65-year-old, gravida 2, para 2, postmenopausal woman complained of recent onset of lower abdominal fullness, swelling of her left leg, and constipation that had worsened over the past 2 months. Her medical history included coronary artery disease with an old myocardial infarction, arrhythmia, mild cirrhosis of the liver, transabdominal hysterectomy, appendectomy, and laminectomy of L5 because of spinal changes from tuberculosis. On physical examination, her blood pressure was 121/78 mmHg, heart rate was 80/minute, respiration rate was 18/minute, and temperature was 36.6°C. Her heart rate and lung sounds were normal, and abdominal examination showed moderate abdominal distension and a 12-cm longitudinal surgical scar over the abdomen. Her abdomen was otherwise soft, with mild tenderness in her lower quadrant without rigidity or palpable masses. Digital examination showed no tenderness or fullness. The left lower leg had mild edema.

Abnormal laboratory data included the following: hemoglobin, 11.4 g/dL; sodium, 133 mmol/L; potassium, 3.0 mmol/L; aspartate transaminase, 40 U/L; albumin, 2.1 g/dL; and CA125, 377 U/mL. A chest film revealed pleural effusion of the bilateral costophrenic angle and patchy opacity of the left lower lung zone.

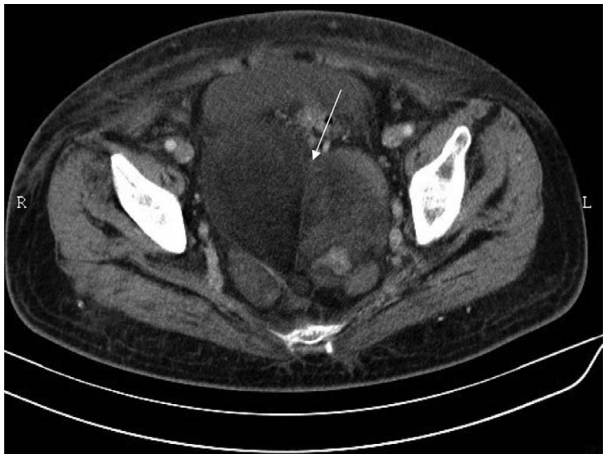

Figure 1. Pelvic contrast-enhanced computed tomography shows a well-circumscribed, presacral, soft-tissue mass. The arrow indicates the tumor.

An electrocardiogram showed an old infarction of the anterior wall. Ultrasonography of the pelvis revealed a large, heterogeneous, predominantly hyperechoic mass. Non-contrast and contrast-enhanced computed tomography (CT) of the abdomen and pelvis showed a presacral 11.5 × 8.5 × 5 cm mass with fatty and soft-tissue components (Figure 1). Other findings included mild hydronephrosis of both kidneys, irregular contour with hypertrophic change of the left lobe of the liver (favoring cirrhosis), small cysts in the left kidney, bilateral pleural effusion, and a small amount of ascites. The mass appeared to be adherent to the sacrum, encircling the rectum and causing mild hydronephrosis of both kidneys.

An exploratory laparotomy disclosed a large retroperitoneal tumor between the rectum and sacrum in the true pelvis (Figure 2). The tumor consisted of soft, greasy, yellow tissue. During removal of the tumor, significant bleeding was noted from the presacral region. Blood transfusions were required. Frozen section showed no evidence of malignancy.

The histopathologic study showed predominantly mature adipose tissue intermixed with extramedullary hematopoietic components. This was consistent with myelolipoma (Figure 3). No evidence of malignancy was seen. The tumor was 12 × 9 × 5 cm in size and weighed 405 g. The patient's leg edema subsided postoperatively.

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Accepted: March 26, 2008

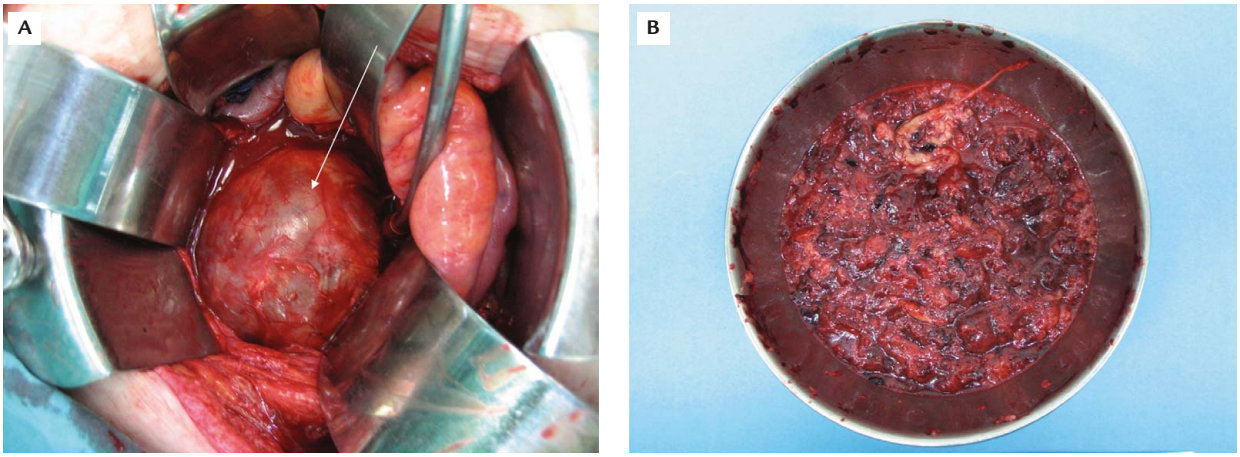

Figure 2. (A) A large retroperitoneal tumor (arrow) located between the rectum and sacrum is seen in the true pelvis. (B) The mass contains red friable tissue and yellow adipose tissue.

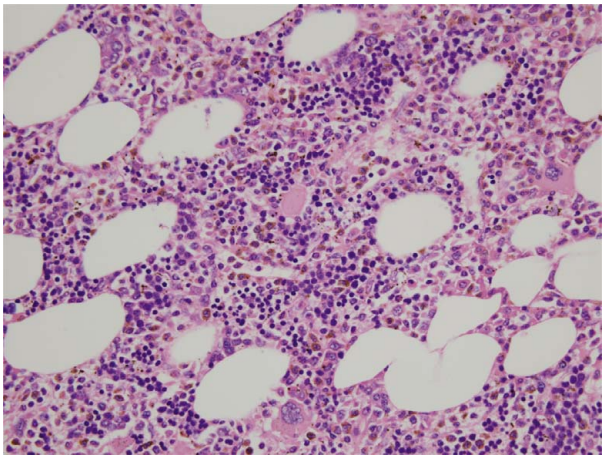

Figure 3. Histopathology of the tumor shows mature adipose tissue intermixed with normal hematopoietic cells (hematoxylin and eosin, 400×).

Myelolipomas are benign tumors composed of mature fatty tissue interspersed with hematopoietic tissue. The adrenal gland is the most common site, but myelolipomas also occur rarely in extra-adrenal locations. The etiology is still obscure. Some reports proposed that endocrine dysfunction be included in the etiology of adrenal and extra-adrenal myelolipoma [3]. Clinical examples like Cushing's syndrome, Addison's disease, adrenal hyperplasia, and chronic use of exogenous steroids have been associated with myelolipoma [3–5]. Experimental evidence like injecting pituitary extract subcutaneously into rats to induce myelopoiesis has been reported [3,6].

Singla et al [2] summarized 37 reported cases of extra-adrenal myelolipomas. Eight patients had a history of diabetes mellitus (21.6%), seven had a past history of cancer (18.9%), and six had a past history of steroid use (16.2%). The mean age of patients found to have this tumor was 65.2±11.2 years, and the male-to-female ratio was 13:24 (i.e. 0.54). These tumors were most commonly found in the presacral region (n=15; 40.5%), followed by the retroperitoneal (n=8; 21.6%), thoracic (n=5; 13.5%), pelvic (n=3; 8.1%), renal (n=2; 5.4%), gastric (n=1; 2.7%), hepatic (n=1; 2.7%), perivesicular (n=1; 2.7%) and multifocal (n=1; 2.7%) regions [2]. Our patient's tumor occurred in the most common site, the presacral region. Her presenting symptoms were related to the effects of the mass.

It is important to distinguish presacral myelolipomas from retroperitoneal tumors, which are more common and aggressive neoplasms. In a 1989 study, Pinson et al [7] divided retroperitoneal tumors into six categories: (1) benign, (2) germ cell, (3) lymphoma, (4) sarcoma, (5) carcinomas, and (6) undifferentiated malignancy. Of these tumors, 89% were malignant [3,7]. This figure is supported by several other studies [8].

For pelvic tumor, ultrasonography, CT scans (particularly spiral CT scans) and magnetic resonance imaging can provide much information about the location, consistency, extent, and possible origin of the mass. Needle biopsy can be useful in determining which therapeutic options could be considered. However, it is often difficult to label a retroperitoneal neoplasm as benign based on needle biopsy or radiographic findings, because both benign and malignant tumors can share similar characteristics [3].

Surgery is the preferred treatment for most primary retroperitoneal tumors. In this patient, even though there was no preoperative histologic diagnosis, frozen section played an important role in determining the extent of surgery needed. Because there was no evidence of malignancy, extensive resection was avoided (34% of retroperitoneal sarcomas require resection of adjuvant organs) [3,9]. This further decreased the possibility of surgical complications, such as easy bleeding, when the sacral attachments are resected [2].

In conclusion, extra-adrenal myelolipomas are rare benign tumors. Due to their fatty and soft-tissue components, it is not easy to distinguish them from other malignancies, like liposarcoma, from radiographic findings [3]. Diagnostic procedures, like CT-guided percutaneous fine-needle aspiration biopsy or frozen section, might be useful in the diagnosis [10–12]. We report this case to help gynecologists become aware of the possibility of extra-adrenal myelolipoma when they formulate a differential diagnosis for retroperitoneal tumors.

References

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