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Case Report

Laparoscopic Radical Excision of Primary Round Ligament Perivascular Epithelioid Cell Tumor Mimicking Leiomyoma

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ABSTRACT Perivascular epithelioid cell tumors (PEComas) are a group of rare mesenchymal tumors including angiomyolipoma, clear cell sugar tumor, lymphangioleiomyomatosis, and other unusual clear cell tumors at various locations. We describe a 45-year-old female patient presenting with a painless mass at the left lower abdomen. Computed tomography showed a circumscribed mass 8 × 7 × 8 cm in the left round ligament of the uterus. The provisional diagnosis was leiomyoma. The patient underwent initial laparoscopic excision. The histological and immunohistochemical diagnosis was malignant PEComa. She subsequently underwent laparoscopic radical excision of the residual left round ligament and surrounding tissue. At 18 months after surgery, she remained well without clinical and radiographic evidence of recurrent disease. According to this report, primary PEComa of the round ligament can mimic leiomyoma. Laparoscopic radical excision might be a feasible and safe alternative treatment of this tumor with a favorable outcome. Journal of Minimally Invasive Gynecology (2009) 16, 626–9 © 2009 AAGL. All rights reserved.

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Perivascular epithelioid cell tumors (PEComas) are defined as ''mesenchymal tumor composed of histochemically distinctive perivascular epithelioid cells (PEC)''[1]. PEC was first proposed by Bonetti et al [2] as a description of a novel cell type that may originate from the walls of blood vessels, on the basis of the observation that these cells are frequently related to such structures. The histogenesis and the normal counterpart of PEC are unknown. For gynecologic tracts, the uterine corpus seems to be the most frequently reported anatomic site of origin. However, PEComas have also been found in the cervix, vagina, vulva, pelvis, broad ligament, and ovary [3-8]. We report a case of malignant PEComa of the round ligament and describe the clinical presentation, computed tomography (CT), pathologic and immunohistochemical diagnosis, minimally invasive treatment, and short-term outcome.

Case Report

A 45-year-old otherwise healthy woman presented with an asymptomatic, enlarging left lower abdominal mass that she had noticed for a few years. On examination, a firm, round mass was palpated in the left lower abdomen just above the left inguinal ligament. A contrast-enhanced abdominal pelvic CT scan showed a well-circumscribed mass 8 × 7 × 8 cm connecting to the thickened left round ligament of the uterus (Fig 1A). Multiple foci of mottle calcification were seen within this mass. No additional mass was noted, nor was adenopathy found in the remainder of the pelvis and abdomen. The initial diagnosis was leiomyoma. The patient underwent laparoscopically complete resection. The intraoperative findings revealed an extraperitoneal mass in the inguinal region just lateral to the medial umbilical fold (Fig 1B). This mass was found originating from the normal intraperitoneal portion of the left round ligament and continuing into the normal distal insertion of it. The right round ligament and other pelvic organs were normal. The laparoscopically excisional procedure

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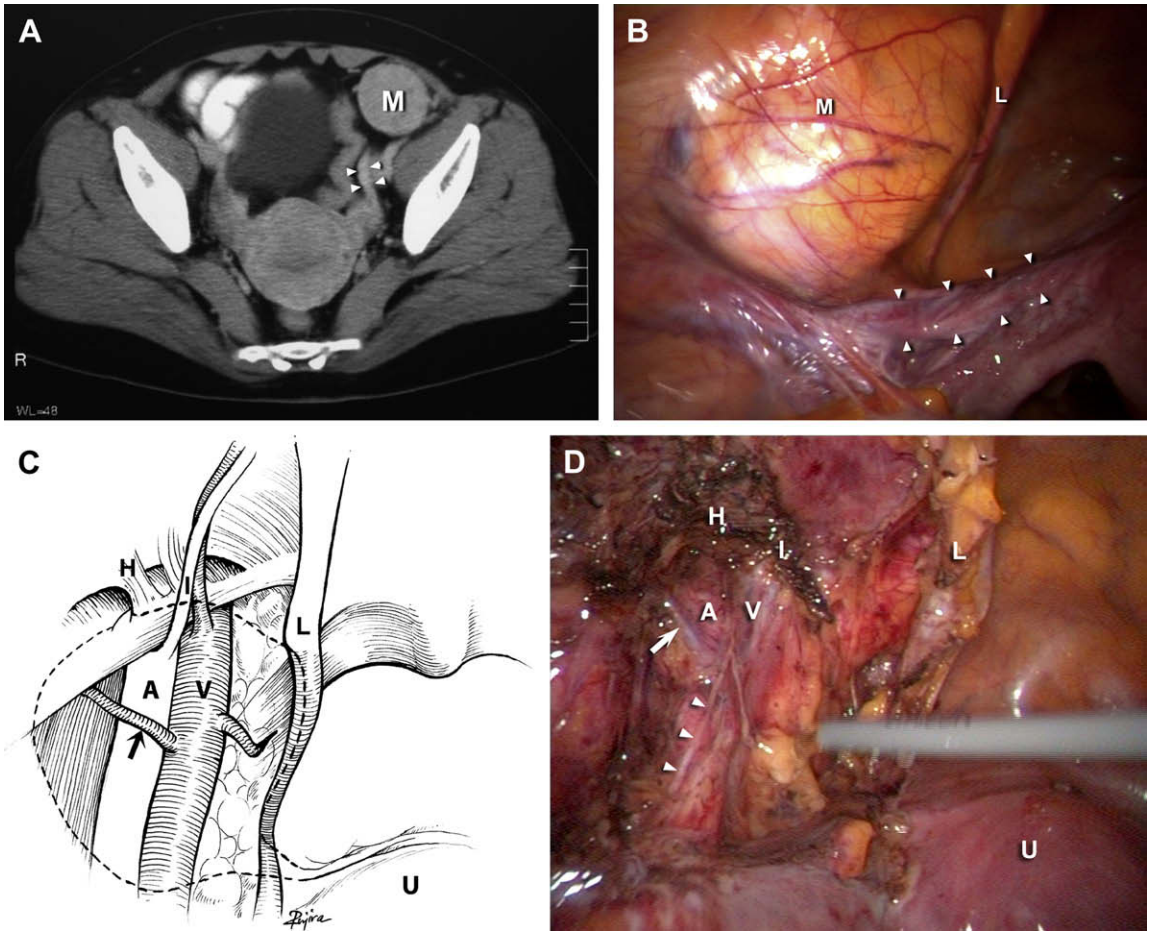

Fig. 1. Left round ligament PEComa. Preoperative axial enhanced CT scan of the pelvis **(A)** and intraoperative laparoscopic view **(B)** shows an extraperitoneal mass *(M)* at the left inguinal region originating from the left round ligament *(arrowheads)* of the uterus *(U)*. A diagram **(C)** depicting the outline of the mass *(dotted line)* and the final laparoscopic view **(D)** shows the relationship between the tumor and essential anatomic landmarks, including the uterus *(U)*, left medial umbilical fold *(L)*, external iliac artery *(A)*, external iliac vein *(V)*, deep circumflex iliac vein *(arrow)*, inferior epigastric vessels *(I)*, genitofemoral nerve *(arrowheads)*, and Hesselbach ligament *(H)*.

comprisal opening the peritoneum, approaching the retroperitoneal space, and excising of the whole mass. The specimen was placed and morcellated in an endobag before being removed from the pelvic cavity.

Pathologic Study

Microscopically, this mass was composed of polygonal cells containing clear or eosinophilic cytoplasm (Fig. 2, *A*). These cells were arranged in small nests, trabeculae, and sheet-like patterns with intervening capillary network. The tumor illustrated prominent vasculature composed of delicate capillaries and vessels with a thick hyalinized wall in some portions (Fig. 2B). The nuclear size was variable, with marked nuclear atypia in several areas (Fig. 2, C1). Neither a mitotic figure nor necrosis was seen. The tumor border was infiltrative (Fig 2, C2). There were a few lymphovascular space invasions (Fig. 2, C3). Immunohistochemical staining was positive for HMB-45 (Fig. 2D), but negative for other antibodies, including cytokeratin, EMA, CAM 5.2, desmin, smooth-muscle actin, chromogranin, synaptophysin, vimentin, S-100 protein, CD117, and calretinin. The diagnosis was a malignant PEComa on the basis of the criteria recently proposed by Folpe et al [6].

Laparoscopy

Because of malignant potential tumor and infiltrative margins, the patient subsequently underwent a second laparoscopic radical excision of the residual left round ligament. The proximal and distal parts of the left round ligament were transected at it abutting on the uterus and at the internal inguinal ring, respectively. The left inferior epigastric vessels were used as the landmark of the inferomedial aspect of the internal inguinal ring, and transected at their origin. The left deep circumflex iliac vein was used as the landmark of the inferolateral aspect of the internal inguinal ring, and preserved. All soft tissue among those described structures was removed en bloc with avoiding injuries of the genitofemoral nerve, iliac vessels, as well as the interfoveolar (Hesselbach) ligament, a condensation of the transversalis fascia at the medial border of the internal inguinal ring (Fig. 1C, 1D). The entire