Fırat Tıp Derg/Firat Med J 2015; 20(1): 57-59

Case Report

Degenerated Primary Retroperitoneal Leiomyoma: A Rare Case

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ABSTRACT

Primary retroperitoneal leiomyomas are a very rare clinical condition. We present the case of degenerated primary leiomyoma in retroperitoneal area. A 33-year-old primiparous woman presented with chronic constipation. On examination, there was a large firm unpainful palpable lumbar mass in the left side. The diagnostic workup included contrast computer tomography, transabdominal ultrasonography, and colonoscopy that revealed a 7,5-cm heterogeneous retroperitoneal tumor in the left infrarenal area. At laparotomy, the mass arising from the retroperitoneum was excised. Histopathology showed a degenerated leiomyoma. Preoperative imaging can only give provisional diagnosis and can be misguiding. Considering current limitations in radiological diagnosis, surgical excision of these tumors is necessary to rule out malignancy.

Key words: *Retroperitoneal space, Degenerated leiomyoma, Retroperitoneal leiomyoma*.

ÖZET

Dejeneratif Primer Retroperitoneal Leiyomyom: Nadir Bir Vaka

Primer retroperitoneal leiyomyomlar oldukça nadir görülen bir durumdur. Biz retroperitoneal alandaki dejenere olmuş bir primer leiyomyom vakasını sunuyoruz.33 yaşında tek doğum yapmış bir kadın hasta kronik konstipasyon şikayetiyle geldi. Fizik muayenesinde sol lomber bölgede ağrısız palpe edilebilen büyük kitlesi ele geliyordu. Tanı için kullanılan kontrastlı bilgisayarlı tomografi, ultrason ve kolonoskopi sonucunda sol böbrek altında retroperitonda 7,5 cm çapında heterojen kitle olduğu saptandı. Laparotomi ile kitle retroperitonden çıkarıldı. Histopatolojisi dejenere olmuş leiyomyom geldi.Operasyon öncesi incelemeler bizim kesin tanı koymamızı zorlaştırabilir ve yanlış yönlendirebilir. Radyolojik tanıdaki güncel sınırlamalar göz önünde bulundurulduğunda bu vakalar için malignensiyi dışlama için cerrahi eksizyon yapılmalıdır.

Anahtar Sözcükler: *Retroperitoneal alan, Dejeneratif leiyomyom, Retroperitoneal leiyomyom.*

Leiomyomas are benign smooth muscle tumors that usually arise from the uterus being the most common benign tumors in women. However, primary leiomyoma of the retroperitoneum without the co-existence of uterine leiomyoma or disseminated disease is very rare (1). When a retroperitoneal mass is detected in the scan, these tumors are usually believed to be malignant. Also, preoperative diagnosis of a retroperitoneal leiomyoma is rarely possible (2). We report a rare case of a degenerated retroperitoneal leiomyoma.

CASE REPORT

A 33-year-old primiparous woman was presented to the department of gastroenterology at our hospital with the 3-month history of constipation. Colonoscopy revealed a partial obstruction at the distal end of the descending colon without mucosa involvement. A heterogeneous retroperitoneal mass in the left lumbar area was found on the abdominal ultrasound. She was referred to our clinic with the suspicion of retroperitoneal malignancy. On examination, there was a large firm unpainful palpable lumbar mass in the left side. Computer tomography demonstrated a retroperitoneal mass of 8x6x5 cm located inferior to the left renal with compression of the descending colon (Figure 1). Magnetic resonance imaging confirmed the finding of the computer tomography (Figure 2). Routine laboratory investigations and tumor markers including alpha-fetoprotein, CEA, CA 19-9, CA-125 were revealed to be within normal range. Chest X-ray did reveal no pathologic finding. Ultrasound guided tru-cut biopsy was performed from the retroperitoneal mass, and pathologic examination of the specimen revealed hypocellular connective tissue and a very little striated muscle, however the specimen was inadequate for tissue diagnosis.

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Received/Geliş Tarihi: 02.04.2013 Accepted/Kabul Tarihi: 02.07.2014

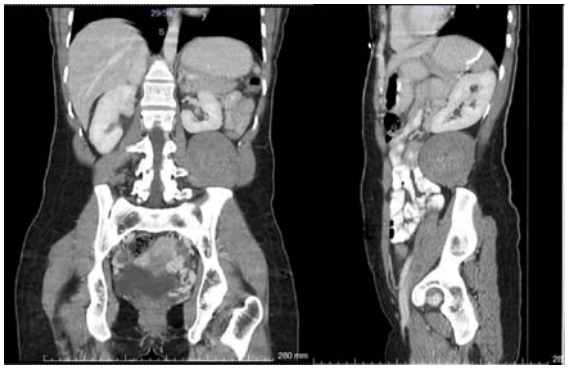

Figure 1. *Computer tomography shows a retroperitoneal mass.*

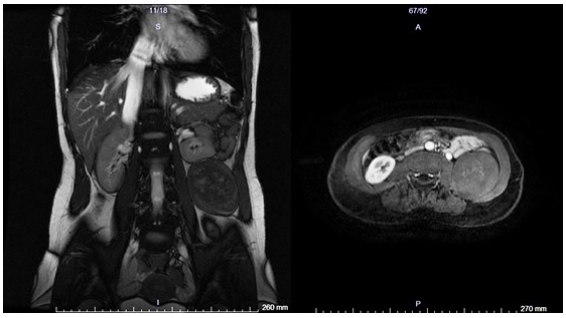

Figure 2. *Magnetic resonance imaging shows a retroperitoneal mass.*

At laparotomy, an encapsulated firm tumor without renal and descending colon connection but adhesive to the psoas muscle was found, arising from the left retroperitoneal region at the L3-L5 level of the spine. The descending colon was compressed partially by the mass and the proximal section of the descending colon did not dilate. There was no connection of the tumor to the genital tract. The mass of size 8x5x5 cm was excised by both blunt and sharp dissection (Figure 3). The postoperative period of the patient was uneventful. On gross examination it was a solid, well-circumscribed and firm mass. Microscopically, the lesion were consisted of vast areas of hyalinization with very few foci having barely recognizable smooth muscle bundles (Figure 4). The patient was examined 2 weeks after the procedure. She had no sign of urinary and bowel dysfunction. There has been no sign of recurrence and the patient remains asymptomatic for 3 months. The patient provided written consent to use the information for the case report.

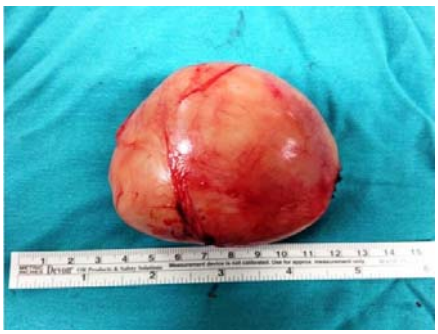

Figure 3. *Macroscopic appearance of the mass after surgical removal.*

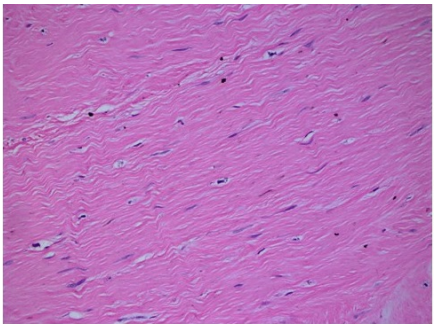

Figure 4. *Within intensely hyalinized stroma attenuated smooth muscle cells are seen. H&E stain x 20.*

DISCUSSION

Leiomyoma is more frequently found in the fourth and fifth decades of life, and the uterine corpus is the most common site of location. However, leiomyomas occasionally occur with unusual growth patterns or in unusual locations (3). Although there have been reports on various atypical localizations for leiomyomas, their growth in the retroperitoneum is extremely rare. It could be related to uterine leiomyomas since more than 40% of patients with retroperitoneal leiomyoma have a concurrent uterine leiomyoma or a history of hysterectomy for leiomyoma (4). The pelvis is the most common location for retroperitoneal leiomyoma (73%). The leiomyoma in the present case is extremely rare, because of the localization and history. The leiomyoma was not in pelvis and was not related to a concurrent uterine leiomyoma or a history of hysterectomy for leiomyoma.

Etiology and pathogenesis of leiomyomas are still poorly understood. Since these tumors probably arise from smooth muscle cells, including those in blood vessel walls, they can originate wherever smooth muscle cells exist (1). Zaitoon explained that uterine leiomyomas that adhere to surrounding structures acquire an auxiliary blood supply and become detached from the uterus (5). Such leiomyomas have been termed “parasitic” leiomyomas. Stutterecker et al. speculated that embryonal remnants (e.g., the tubes of Müller or Wolff) or local vessel musculature might be responsible for the development of retroperitoneal leiomyomas (6).

Retroperitoneal leiomyoma may enlarge considerably, yet remain asymptomatic and be detected incidentally at a routine check-up or autopsy. The most common clinical feature of a retroperitoneal leiomyoma is palpation of a pelvic mass, which is present in nearly 90% of patients (3). Although this patient had chronic constipation, the findings of the colonoscopy and intraoperative visual inspection showed that the compression of the mass in the descending colon could not cause chronic constipation.

Although preoperative imaging and gross appearance are frequently typical for uterine leiomyomas, retroperitoneal leiomyomas are frequently misdiagnosed as malignant tumors. Ultrasonographyc examination provides good initial orientation for retroperitoneal masses. Computer tomography and especially magnetic resonance imaging are most useful screening tools in evaluating and distinguishing the exact nature of the tumor and its relationship with adjacent organs and vascular structures. However, no radiological diagnostic modality appears highly sensitive or specific in ruling out malignancy and differential diagnosis on the basis of radiological finding alone is difficult (1). For the expectant management of asymptomatic patients refusing operation, ultrasonography or computer tomography-guided fine-needle aspiration or biopsy may be histologically beneficial. Complete tumor excision with pathologic investigations is still the mainstay for diagnosis and treatment (7).

If a retroperitoneal mass is diagnosed as leiomyoma either preoperatively or intraoperatively, the surgeon should excise the tumor as completely as possible, especially in symptomatic patients. However, the tumor can displace important retroperitoneal and vascular structures; therefore, sometimes resection of the tumor might be incomplete (2). Abdominal hysterectomy along with the resection depends on the age of the patient, her symptomatology, and associated uterine myoma (8). We excised only the mass because it did not connection any organs or structures.

Retroperitoneal leiomyomas of located outside the pelvis are extremely rare and generally detected incidentally at a routine check-up. No radiological diagnostic modality appears highly sensitive or specific in ruling out malignancy and differential diagnosis on the basis of radiological finding alone is difficult. Complete tumor excision with pathologic investigations is still the mainstay for diagnosis and treatment.

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