Myolipoma: A clinical-Radiologic-Pathologic Mimic of Malignant Retroperitoneal Tumors

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Abstract

Myolipoma is a rare benign soft tissue tumor that can mimic a malignant retroperitoneal tumor. It consists of mature adipose tissue mixed with bundles of well-differentiated smooth muscle cells, and is most often found in the retroperitoneum and abdomen. It can grow to a large size, and is often found late as an incidental finding due to non-specific abdominal and pelvic discomfort or pain. Imaging will reflect the dual composition of the mass, with the possibility of a soft tissue sarcoma high on the differential. Histologically, the differential diagnosis is well-differentiated/dedifferentiated liposarcoma, lipoleiomyosarcoma, spindle-cell lipoma, angiolipoma, angiomyolipoma, and leiomyoma with fatty degeneration. Surgical removal is usually necessary, since myolipomas in the retroperitoneum can be very large. A myolipoma is completely benign, has no metastatic potential, and has not been reported to recur in all of the cases to date, therefore an accurate diagnosis is essential.

Introduction

Here we present a case of a 59 year old female with a history of leiomyomas and a new complaint of pelvic pain. After imaging studies were completed, a soft tissue sarcoma in the right lower quadrant was high on the differential diagnosis. The patient underwent surgery for the leiomyomatous uterus and the right lower quadrant mass, which was ultimately diagnosed as a myolipoma. On histologic examination, it was found to contain mature adipose tissue mixed with bundles of well-differentiated smooth muscle cells, indicating it was most likely benign, with immunohistochemical stains supporting this diagnosis. It could easily be misdiagnosed as a malignant neoplasm, such as a liposarcoma, due to the location and characteristics seen on imaging and pathologic examination.

Case Report

This 59 year old G0P0 Caucasian female was seen for her annual gynecologic exam, complaining of suprapubic tenderness, pain and dyspareunia for 10 days. She also complained of urinary stress incontinence with laughing and jogging. She had been seen by her gynecologist approximately one and a half years earlier and was diagnosed with a posterior uterine fibroid on office ultrasound and had also been previously treated for lower back pain. On exam, her cervix was tender, and the uterus was not palpable due to size of the abdomen. She was then referred for abdominal and pelvic ultrasound imaging a couple of days later. The abdominal ultrasound was grossly unremarkable. The pelvic ultrasound revealed multiple intramural fibroids, one of which was located in the fundus measuring 2.4 cm. The endometrium was abnormally thickened, measuring 1.9 cm. The uterus measured 8.1 x 5.0 x 5.8 cm. A large hyperechoic area in the right lower quadrant was also seen (Illustration 1 – white arrow), without peristalsis, and was measuring approximately 6 cm. This hyperechoic area was the site of the patient's pain. According to the radiologist's report, it could represent bowel versus a mass, and CT of the pelvis with contrast was recommended.

Two days later the patient went for a CT scan of the pelvis with IV and oral contrast, which showed a well-defined minimally enhancing ovoid shaped mass (Illustration 2 - white arrow), separate from bowel and uterus, in the right lower quadrant measuring 10 x 4 cm in size with some intrinsic low attenuation. The bladder was normal and the endometrium was thickened (Illustration 2 - red arrow). Heterogeneity in the uterus was consistent with a leiomyomatous uterus (Illustration 2 – yellow arrow). Radiologic impression for the right lower quadrant mass was a soft tissue sarcoma. However, an inflammatory or infectious process, hematoma or spontaneous hemorrhage could not be excluded.

The patient was then scheduled for a laparotomy with total abdominal hysterectomy and bilateral salpingo-oophorectomy about 2 weeks later. After the hysterectomy was completed, dissection of the pelvic mass seen on CT was commenced. During surgery, this mass was actually found to be outside of the pelvis and in the right lateral abdominal wall behind the peritoneum, just below the appendix and above the iliac vessels and ureter. On inspection, the mass was soft and about 10 cm in size.
The mass was submitted to pathology for frozen section evaluation. On gross inspection the mass was noted to be slightly ovoid and slightly nodular, measuring 10.5 x 8.2 x 3.6 cm weighing 154.9 grams. The surface was mostly smooth and pink and pink-tan in color. The mass was serially sectioned and revealed lobulated soft and rubbery pale to glistening tan cut surfaces, without any necrosis. On frozen section, however, the mass was diagnosed as a fatty mesenchymal tumor that had to be referred to permanent sections for a definitive diagnosis. No ovarian tissue was identified on frozen section evaluation. The surgeon proceeded to remove more fatty tissue residing along the lateral abdominal wall overlying the iliac vessels, in light of the fact that liposarcoma was a concern. Histologically, on permanent sections, the mass was composed of adipose tissue mixed with scattered bundles of smooth muscle (Illustration 3, 4). The smooth muscle cells did not show cytologic atypia, as there were uniform looking oval nuclei, without cytological pleomorphism or mitotic activity. Some scattered small vessels were noted, but not significantly prominent. Also, necrosis and hylalnized vessels were not present. Immunohistochemical stains were then performed. Staining for smooth muscle actin (Illustration 5) was positive as expected. Ki-67 labeling was very low in both the smooth muscle and adipocytes, indicating the mass was most likely benign. HMB-45 and HHV-8 stains were negative. Estrogen receptor expression was not seen, however, staining for progesterone receptor expression was focally positive (Illustration 6). Due to the above findings, the mass was diagnosed as a myolipoma.

As of this writing, approximately four and a half years after the patient’s surgery, the patient is alive and well. Discussion

Myolipoma of soft tissue is a rare tumor, initially described by Meis and Enzinger in 1991 [1]. It mostly occurs in the abdomen and retroperitoneum, but has also been reported in the round ligament, broad ligament, fallopian tube, inguinal region, orbit, pericardium, subcutaneous tissue, breast [1-5]. The term “extrauterine lipoleiomyoma” when referring to a myolipoma has been discouraged, in order to avoid confusion with masses arising from the uterus [6]. Lipoleiomyoma, once termed “leiomyoma with fatty degeneration”, was traditionally thought to be a variant of leiomyoma [7]. This is controversial however. It is now believed that it is an entirely separate tumor, since the fat component is an integral part of the lesion, and not just a focally degenerated area of smooth muscle tissue [7,8]. All of the retroperitoneal myolipoma cases that have been reported in the literature have been found in women. They can grow to be very large, in some cases up to 30 cm in greatest dimension [3]. As most cases are in the retroperitoneum and abdomen, patients will present with vague abdominal pain or distension. In some cases the mass was found incidentally on imaging or during surgery. The most immediate concern for a fatty retroperitoneal mass is a sarcoma, such as liposarcoma or leiomyosarcoma, since these are the majority of fat containing retroperitoneal masses and are malignant tumors. Often, myolipoma cases are misdiagnosed initially as liposarcoma, since it is difficult on imaging to distinguish between the two and there is a very small amount of information in the literature describing the radiologic features of a myolipoma [9]. The features seen on ultrasound, CT and MR imaging reflect its intermixed dual tissue composition, as noted in pathologic examination. The US, CT, and MR imaging appearances vary along a spectrum, representing varying proportions of mature adipose tissue and poorly defined non-adipose tissue representing smooth muscle. On CT, both fat and soft-tissue attenuation areas are noted (as identified in our case). Presence of calcifications varies and more commonly occur in large-sized lesions [9]. On MRI, smooth muscle elements appear as areas of intermediate signal intensity on T1 and intermediate to high signal intensity on T2 weighted sequences. The fat component of these lesions is bright on T1 and T2 weighted sequences [10]. These features might also suggest a well-differentiated liposarcoma, and the imaging appearance does not allow for distinction between these two lesions [11]. Grossly, myolipomas are soft to firm in consistency, with a smooth or lobulated surface. On cut surface they can appear homogenous or septated by bands of fibrous tissue [2,5]. Histologically, the tumors normally present as an admixture of mature smooth muscle cells and mature adipose tissue without any lipoblasts, flioret-like giant cells, or cytologic atypia. Our case was consistent with this pattern. Also, Ki-67 labeling was low, further demonstrating the benign nature of this tumor. Immunohistochemical stains for smooth muscle actin, desmin and vimentin are usually positive. Staining for HMB-45 has been negative. S-100 has also shown to be positive in the adipocytes in some cases [5], however it was negative in several other cases within the smooth muscle component [8]. Our case was not evaluated for S-100 activity. Estrogen and progesterone receptor status has rarely
been documented in myolipomas. Our case showed focal progesterone positivity in the smooth muscle component, and staining for estrogen receptors was negative. Expression for estrogen receptors and progesterone receptors in both the smooth muscle cells and adipocytes has been shown to be positive in some cases, leading to the theory that there could be a common Mullerian origin between the two components of the mass [5]. Therefore, the significance of the hormone receptor status in the pathogenesis of this type of tumor is uncertain. It is interesting to note, however, that in one review of clinical data from 249 patients with non-gastrointestinal stromal tumor soft tissue sarcomas, the coexpression profile of estrogen receptor negativity and progesterone receptor positivity was found to be a negative prognostic factor [12].

The differential diagnosis of myolipomas on histologic examination includes well-differentiated/dedifferentiated liposarcoma, spindle-cell lipoma, angiolipoma, angiomyolipoma, and leiomyoma with fatty degeneration. In well-differentiated/dedifferentiated liposarcomas, there is mature fat present, but, lipoblasts and atypical hyperchromatic cells with mitotic activity are also present [6]. These features distinguish liposarcoma from myolipomas, as lipoblasts and cytologic atypia are absent in the latter. The typical spindle-cell lipoma would contain equal amounts of mature fat and spindle cells that are negative for smooth muscle actin and desmin, since no smooth muscle component is present. Also, spindle cell lipomas are generally found as a subcutaneous mass in the neck, shoulders, and back. Angiolipomas are usually found subcutaneously rather than in the retroperitoneum. Microscopically, they consist of mature fat cells and small vessels that are interspersed throughout [6]. There is no smooth muscle component to this lesion, as there is in a myolipoma. Angiomyolipoma is another tumor on the differential of a myolipoma. Similarly, it can grow to a very large size, in some cases up to 36 cm [14]. However, angiomyolipomas contain medium-sized arteries with thick muscular walls, and are HMB-45 positive [6,14]. As we mentioned before, some small scattered vessels without hyalinization were present in our case, distinguishing our mass from an angiomyolipoma. Angiomyolipomas are associated with tuberous sclerosis [6], which our patient did not have. Lastly on the differential would be leiomyoma with fatty degeneration. However, this mass tends to show only focal areas of mature fat cells and not the regular distribution found in a myolipoma [6].

Our case is consistent with the other reported cases of retroperitoneal myolipomas, clinically and histologically. The patient presented with non-specific abdominal and pelvic pain. Also, the mass grossly appeared similar to other cases, and demonstrated similar benign histological features, such as mature adipose tissue regularly interspersed with mature smooth muscle cells, and a lack of cytological pleomorphism or mitotic activity on H&E. The immunohistochemical workup in our case was consistent with previous reported cases, as smooth muscle actin staining was positive and HMB-45 staining was negative. Hormone receptor positivity has been found in other cases as well. The mass was thought to be completely benign and would not recur after surgical removal, which indeed has been the case.

Surgical resection is the treatment for both myolipomas and liposarcomas. However, resection for a retroperitoneal sarcoma needs to be aggressive because of the invasive nature of the tumor [15]. There have been no reported cases of local recurrence or metastasis after complete surgical removal of a myolipoma. Therefore, initial treatment and prognosis will differ greatly.

**Conclusion**

A malignant mass, such as a liposarcoma, is always a concern on the differential diagnosis for a retroperitoneal mass. Clearly, the clinical presentation will not allow for the distinction between a malignant and benign mass, and even radiologic imaging for these types of masses makes it very difficult, if not impossible, to distinguish. Perhaps if one could make the distinction, a patient with a small benign mass that presents without any symptoms clinically, could be spared surgery. Ultimately, the pathologic examination is crucial, whether intraoperatively or postoperatively. Histologically, much of the mass can look very similar to a liposarcoma, aside from the few key features mentioned above. The diagnosis will affect the surgeon’s decision on how aggressive the resection should be, and also will change the patient’s prognosis drastically, as a liposarcoma has the chance to recur or metastasize in the future. Therefore, it is always important to consider the possibility of a benign neoplasm, such as a myolipoma, and to make an accurate diagnosis in these types of cases.

**References**

Illustrations

Illustration 1

Illustration 1: Transabdominal ultrasound shows hyperechoic mass like area (white arrow) adjacent to urinary bladder that measures 6 cm.

Illustration 2

Illustration 2: CT with IV and oral contrast of pelvis showing large, well-defined, heterogeneous mass in right lower quadrant (white arrow) with internal low fat densities (white arrowhead). Note leiomyoma in uterus (yellow arrow) and thickened endometrium (red arrow).
Illustration 3

Illustration 3: H&E stain showing mature smooth muscle cells and adipose tissue regularly distributed

Illustration 4

Illustration 4: H&E stain showing mature smooth muscle cells and adipose tissue regularly distributed
Illustration 5

Illustration 5: smooth muscle actin positivity

Illustration 6

Illustration 6: focal progesterone receptor positivity
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