Case Reports

Recurrent extraperitoneal low grade leiomyosarcoma with unusual localization

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Summary

Primary extragenital leiomyosarcoma is rarely found in pelvic localization. A 33-year-old multiparous woman who had recurrent low-grade leiomyosarcoma presented with the complaints of dyspareunia, pelvic pain and gait disturbance. Her past medical history revealed she had been subjected to maximal excision of a paravaginal mass by using vaginal and suprapubic transverse incision three years before. The pathology report showed that she had leiomyoma. Three years after the initial surgery, a paravaginal fixed mass was observed at the initial tumor bed and removed by the perineal approach. The histological examination of the specimen revealed a low grade leiomyosarcoma. She was discharged from hospital without any complications.

Key words: Low grade leiomyosarcoma; Extraperitoneal; Pelvis; Recurrence.

Introduction

Sarcomas account for 1% of all solid tumors that can arise anywhere in the body including the incidence of the lower extremities (46%), upper extremities (13%), and retroperitoneum (12%), truncal sarcomas (19%), head and neck sarcomas (9%) and frequently appear between the fourth and fifth decades of life [1, 2]. Intra-abdominal and retroperitoneal leiomyosarcomas are rare among the soft tissue sarcomas. Pelvic and especially paravaginal occurrence of such tumors are extremely rare and surgical excision with disease-free margins is the treatment of choice [3, 4]. The major determinants of malignancy of such tumors are mitotic activity, cellular atypia, and necrosis. The local recurrence rate of leiomyosarcoma is between 40 and 80% [5].

A patient presented with a recurrent soft tissue tumor located in the pelvic side wall and paravaginal area. The importance of magnetic resonance imaging (MRI) in the differential diagnosis is emphasized and the related literature is briefly reviewed.

Case Report

A 33-year-old woman, gravida 3, para 3, was admitted to our clinic with the complaints of dyspareunia, pelvic pain and gait disturbances. Her obstetric history was unremarkable. Three years before she had been admitted to another center with the complaints of pelvic pain and dyspareunia. She was subjected to excision of a paravaginal leiomyoma (Figure 2a) which was almost 10-15 cm in diameter via a vaginal and suprapubic transverse incision. Physical examination revealed no additional abnormalities. Pelvic examination revealed a large firm paravaginal mass at least 15 cm in diameter extending from the left posterolateral vagina to the sacrum. The uterine body, cervix, and ovaries were normal. Biochemical tests including renal and liver functions and tumor markers (CA-19.9, CA 15.3, CEA) were within the normal ranges except for a minimal elevation of serum CA 125 (59.31 U/ml (0-35)) levels.

Suprapubic and transvaginal ultrasound confirmed the finding of a large hypoechoic solid mass, which occupied approximately two-thirds of the pelvis and displaced the vagina to the right side. The mass seemed to originate from the left pelvis soft tissue, especially from the levator ani muscle. The mass was well shaped and without findings of local infiltration or ascites. After ultrasonography MRI was performed to better delineate the borders and extent of the tumor. Axial T1-weighted images before and after intravenous gadolinium administration, T2-weighted images in the axial and coronal plane with and without the fat suppressed technique all showed two well-defined solid individual masses (Figure 1). The larger one measured 9.8 x 7.1 cm in size and was located in the left paravaginal space, deviating to the right. The tumor extended superiorly to the obturator foramen, but it was confined within the pelvis. However another extrapelvic smaller mass measuring 2 x 1.5 cm in size was also noted within the gluteal muscles next to the sacral plexus. Since the MRI exam showed two separate but radiologically similar masses inside and outside the pelvis, the preoperative diagnosis was leiomyosarcoma given the prior history of a vaginal leiomyoma excision.

Because there were no findings of infiltration of the pelvic viscera and/or vessels, the patient underwent excision of the pelvic extraperioneal mass by the perineal approach. The resected mass measured 13 cm in length and 11 cm in maximum width. The genital system was normal. On microscopic examination, the tissue was composed of smooth muscle cells forming bundles and strands in a whorled pattern without capsule formation. The mitotic activity was 2-3 mitotic figures per 10 high power fields and particular cellular atypia and necrosis were detected in some areas (Figure 2b). There were

Revised manuscript accepted for publication October 25, 2003

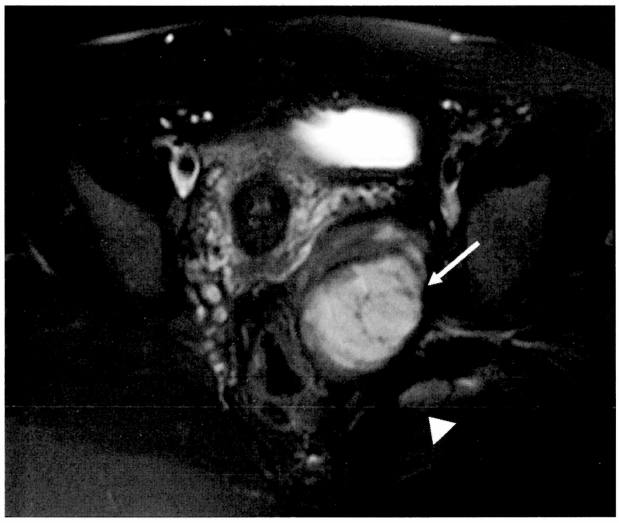

Figure 1. — Axial T2-weighted image shows a large extraperitoneal, predominantly high signal intensity mass at the left paravaginal space (arrow). The mass, measuring 9.8 x 7.1 cm in size, extends to the obturatory foramen. There is a second mass outside the pelvis in between the gluteal muscles. Two solid individual masses, the larger one at the left paravaginal space (arrow) and the smaller one (arrowhead) at the gluteus next to the sacral plexus, diffusely enhance after contrast administration.

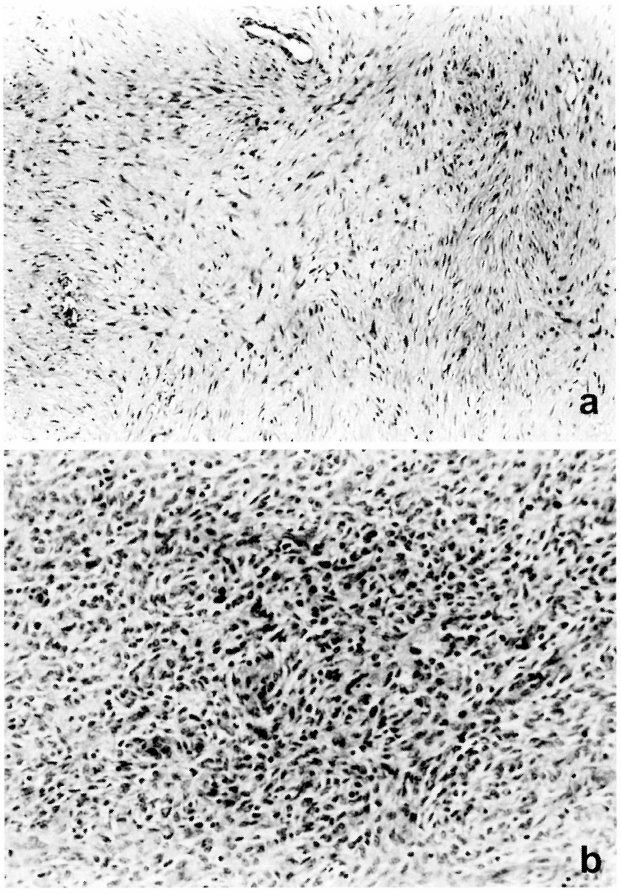

Figure 2. — (H&E x 100) Histological appearance of leiomyoma (a) and low-grade leiomyosarcoma (b). Figure 2b - moderate atypical mitosis; and increased cellularity compared to Figure 2a.

no tumors in the surgical margins. The diagnosis was low grade leiomyosarcoma of the pelvic connective tissue. On the tenth hospital day she was discharged from the hospital without any postoperative complications. Since the diagnosis was low-grade leiomyosarcoma, postoperative adjuvant chemotherapy and pelvic irradiation were not given.

Discussion

Leiomyosarcoma of the soft tissue is uncommon, but represents approximately 5-10% of sarcomas. The origin of pelvic soft tissue is very rare. The unique feature of pelvic side-wall sarcomas is their large size at the time of diagnosis, often reaching a tremendous size prior to diagnosis. The most common clinical presentation of retroperitoneal sarcoma is an abdominal mass. In the presented case the patient was admitted with the complaints of dyspareunia, pelvic pain and gait disturbances. The most important characteristic of these tumors is the high local recurrence rate. The most common site of recurrence is the bed of tumor removal [2]. The local recurrence rate ranges in between 40 and 80% [5] and local recurrence is mainly related to previous treatment failure. For pelvic side-wall sarcomas there is also increased risk of hematogenous seeding. Malignant transformations are more common in extrauterine leiomyomas, as in our case [2].

The diagnosis of a pelvic soft-tissue sarcoma is rarely made preoperatively. In cases of a pelvic cavity mass, abdominal and especially transvaginal ultrasound should be performed to exclude gynecologic pathologies [2, 4]. The anatomical site of origin of a non-gynecologic pelvic mass is a key factor in choosing the surgical approach. In the preoperative evaluation of such pelvic masses MRI provides the anatomical site of origin and helps in determining the feasibility of resection [6]. The best way to achieve an exact diagnosis of such tumors is histological examination [2, 4].

The only treatment for pelvic leiomyosarcoma associated with long-term survival is complete surgical removal of the tumor with adequate margins. After resection of the whole tumor, if the microscopic examination shows negative margins and low-grade tumor, only follow-up is recommended. In advanced staged tumors, tumoral debulking and adjuvant chemotherapy may be a choice [2]. In our case the tumor recurred three years later, and the final diagnosis of low-grade leiomyosarcoma implied that low-grade leiomyosarcoma might have been missed in the initial histological examination. However re-examination of the initial specimen did not confirm the diagnosis of leiomyosarcoma. The histological examination of both specimens suggested malignant degeneration of a pre-existing leiomyoma to leiomyosarcoma. In the pre-

sented case, there was no relationship with the pelvic vessels or viscera, no infiltration of adjacent structures, and the tumor was well circumscribed. These all are usually considered as positive prognostic factors [2, 3].

MRI with its multiplanner and multisequential imaging capability is the imaging of choice for pelvic tumors both before and after surgery. It perfectly delineates the borders of the tumor and its relation with normal soft tissues, vessels and nerves. With the given history of a vaginal leiomyoma excision, recurrent leiomyosarcoma with local metastasis was the initial diagnosis radiologically since two separate masses on both sides of the obturatory foramen were identified.

In this case study the recurrence of a pelvic soft-tissue low-grade leiomyosarcoma three years after the initial surgery is reported. To our knowledge, excisional therapy is the best choice for such tumors to relieve symptoms without infiltrating the pelvic viscera or vessels. This study also shows the importance of MRI in the preoperative evaluation in determining the feasibility of resection.

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