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Pelvic Leiomyosarcoma obstructing vaginal opening – case report

Case report

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Abstract: We present a 42-year-old female admitted for a 4 month history of increasing pelvic discomfort and pain. Clinical examination revealed a large tumor obstructing the vagina. Tumor markers (CA 125, CEA, AFP and CA 19-9), white blood cells and biochemical tests were all within the normal limits. Pelvic ultrasound and magnetic resonance imaging scan confirmed the presence of a large retroperitoneal/pelvic mass. The tumor was surgically excised and pathohistologically diagnosed as a well differentiated leiomyosarcoma, staged IB. Six years after surgery the patient is well and disease free.

Keywords: *Leiomyosarcoma • MRI • Retroperitoneal • Surgery • Vagina*

1. Introduction

Soft tissue sarcomas are rare and histologically heterogeneous mesenchymal tumors, usually with poor outcome. They represent 1-1.5% of all malignancies in adults. Approximately 10-15% of soft tissue sarcomas arise from the retroperitoneum [1]. Histologically, the most common types are liposarcoma, fibrosarcoma, rhabdomyosarcoma, leiomyosarcoma and angiosarcoma [2]. Occasionally, retroperitoneal tumors may grow very large before the first clinical symptoms occur. The symptoms are often non-specific, or may be the result of infiltration or pressure on surrounding structures [1]. As ultrasound scanning is often inconclusive, in most cases magnetic resonance imaging (MRI) enables both determination of the exact location of the tumor mass, and its relation to surrounding structures, including nerves and large blood vessels. If possible, complete surgical excision is the recommended treatment. The completeness of excision depends on the involvement of adjacent structures. The outcome of treatment is usually poor due to the tumor's high recurrence rate and the metastatic potential [2,3].

2. Case report

We present a 42-year-old Caucasian female from a rural part of Serbia, admitted for a 4-month history of

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increasing pelvic discomfort, followed by lower abdominal, low-back and pelvic pain and dyspareunia. She had a 3 year history of hypermenorrhea accompanied with dysmenorrhea and general weakness. Complete vaginal and either tenaculum or bimanual examination was not possible, as the tumor mass completely obstructed the vaginal opening, being detected proximally and close to the introitus, but without infiltration of the vaginal walls. Thus, examination of the upper parts of the vagina as well as the cervix could not have been achieved. No local or generalized lymphadenopathy was noticed. The patient had three normal vaginal deliveries, the last one 15 years ago. Complete blood count (CBC) and erythrocyte sedimentation rate (ESR) were normal, and all tumor markers were within the normal range (CA 125: 10.1 U/ml, CA 19-9: 5.65 U/ml, CEA: 4.89 ng/ml, and AFP: 2.89 ng/ml). Pelvic ultrasound scan performed by convex probe was inconclusive, confirming the presence of an unusually large pelvic mass, localized behind and below the uterus level, more to the right pelvic side. Uterus was enlarged, measuring 86x59x67 mm, with several myomas up to 5 cm with secondary changes, and thin endometrium. Both ovaries were normal. There were no pathological findings on abdominal ultrasound and chest radiography as well. Abdominal and pelvic MRI confirmed the presence of a 15x12x9 cm, retroperitoneal tumor mass, in intimate contact with large blood vessels, without their infiltration and without enlarged lymph nodes. In order to prevent ureteral injury, a day before surgery cystoscopy was performed and bilateral ureteral catheters were placed, that were removed the second day after surgery. On surgery, the uterus was enlarged, with an uneven surface, with a few sub-serosal and intramural leiomyomas up to 5 cm in diameter. The ovaries and tubes on both sides were normal. In the retroperitoneal space, behind and below uterus, there was a large tumor measuring approximately 15 cm in maximum dimension, with a smooth and partly granular surface, without infiltration of surrounding blood vessels

Figure 1. Closer view on sacrouterine ligaments and upper part of tumor.

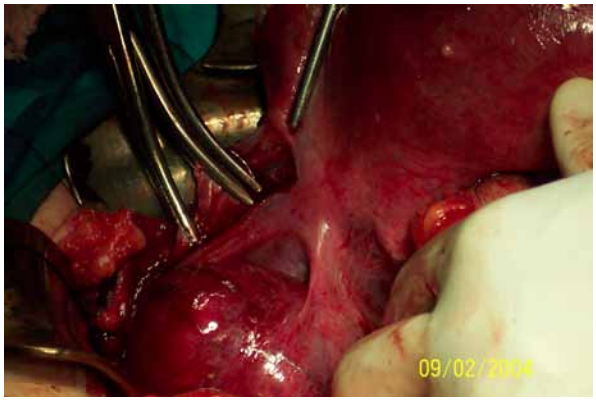

Figure 2. After hysterectomy, division of sacrouterine ligaments was done, retroperitoneal space is entered, and excision of the first part of tumor is accomplished.

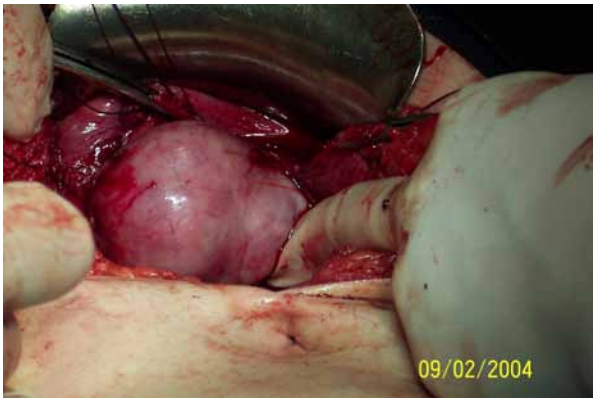

Figure 3. Retroperitoneal tumor was dissected from surrounding structures and removed completely.

and nerves (Figure 1). The parietal peritoneum of the anterior abdominal wall and the visceral peritoneum of the urinary bladder were unremarkable. Small and large bowel and other abdominal organs were without visible pathological change. No pelvic and/or paraaortic lymphadenopathy was found. The classical hysterectomy with bilateral salpingo-oophorectomy was performed. The retroperitoneal space was entered by cutting the peritoneum between sacrouterine ligaments (Figure 2). The retroperitoneal tumor was inclavated into the vaginal opening, but without infiltration of the vaginal walls. The tumor was removed completely and sent for histopathological analysis (Figure 3).

Macroscopically, the tumor measured 15x10x8cm and weighted 420 grams. On sectioning, it consisted of solid, grey/yellowish eddy tissue. Uterus showed a few intramural and sub-serosal leiomyomata up to 5cm. Ovaries/fallopian tubes were unremarkable.

On microscopy, the tumor was compactly hypercellular, composed of spindle cells showing mild atypia and on occasion pleomorphic nuclei. Mitotic count was 2/10 HPF (high-power fields) and atypical mitoses were observed. There was at least one focus of

coagulative tumoural necrosis and evidence of bleeding. The tumor showed strong expression of vimentin, smooth muscle actin and desmin. It was negative for CD117, CD34, S100 and AE1/AE3. Ki67 was 10%. The features were compatible with those seen in a well differentiated leiomyosarcoma.

As intraoperative findings indicated that the tumor was clearly marked, without infiltrating fascia or nearby structures, and histopathologically well differentiated, according to the AJCC protocol, it was staged as Ib, and no further treatment has been required. Follow up was performed using abdominal and pelvic computerized tomography (CT) scan in regular time intervals. During the first two years it was performed in 6 months intervals, and from the third year in the periods of 12 months. All those CT findings were normal over a period of 6 years.

3. Discussion

Leiomyosarcoma is an aggressive soft tissue sarcoma, which mainly originates from the smooth muscle cells of the various body organs and structures including uterus, gastrointestinal tract, soft tissue and large blood vessels [1,2,4]. Leiomyosarcoma has specific clinical, pathohistological and radiographic features. Prognosis and survival rate of this tumor is among the lowest of all soft tissue sarcomas and directly depends on tumor location and size [4]. Leiomyosarcoma usually occurs in the fifth and sixth decade and the sex incidence depends on tumor location, with women making up the large majority of patients with retroperitoneal and inferior vena cava leiomyosarcomas. Retroperitoneal leiomyosarcoma is often presented by palpable abdominal mass and swelling, pain, loss of weight, nausea and vomiting [5].

Retroperitoneal leiomyosarcoma is usually large in diameter, an average of 11 cm [6]. The presence of a retroperitoneal tumor should be confirmed by using CT and MRI as the clinical and ultrasound findings may often be inconclusive. MRI is better for determination of tumor origin and involvement of the surrounding structures, while CT is superior for the evaluation of metastatic disease [6]. Prior to surgery, it is important to exclude the infiltration of large blood vessels. Several cases of large blood vessel involvement however, were described in the literature (iliac artery and vein, inferior vena cava, and even ovarian vein [7–9]. Preoperative evaluation is of particular importance for the planning of a surgical approach and procedures, in order to achieve an appropriate wide resection, which is crucial for the prevention of local recurrence and successful outcome [6]. In our case, MRI findings indicated that the tumor was in close contact with large blood vessels, but without their infiltration and without the existence of enlarged lymph nodes. Intraoperative findings corresponded to the MRI scan, and the tumor was completely removed. In cases when it is impossible to achieve adequate/complete surgical removal, radiotherapy should be considered, along with chemotherapy. For some cases the outcome depends on both radicality of surgery, and tumor characteristics (location, size, histopathological type and grade) [10]. American Joint Committee on Cancer (AJCC) staging system is the most commonly used, including histological stage, tumor diameter (larger or smaller than 5 cm), localization of the tumor and the presence of metastatic disease [2,11]. Macroscopic examination revealed that the tumor in our case measured 15x10x8 cm. Hystopathologicaly, it was a well-differentiated leiomyosarcoma, composed of spindle cells showing mild atypia, while the mitotic count was 2/10 HPF with atypical mitoses. Following the AJCC staging system this tumor was staged IB. According to the literature, when a tumor is larger than 5cm, with the present atypia, fields of necrosis and increased mitotic index, mortality rate is very high, ranging from 77-93% [12]. Despite the high literature mortality rate for this type of tumor and disease stage, the outcome of treatment in our case was successful. Local recurrences and metastases could be found in different locations, and in one case, nine recurrences with different locations were recorded during the period of 8 years after the first surgery [13]. Most tumor recurrences occur within 2 years of initial surgical resection [14]. As most recurrences are local, careful follow-up imaging should be done. Clinical follow-up in these cases is usually inconclusive, as up to 50% of patients are asymptomatic. One suggested imaging follow-up scheme is to obtain imaging at regular intervals using CT or MRI every 3–4 months for 2 years, then every 4–6 months for 3–5 years, and every 12 months thereafter. Follow up for greater than 5 years is recommended [14]. Our patient was followed up using abdominal and pelvic CT scan during the first two years in 6 months intervals, and from the third year once a year. All those CT findings were normal over a period of 6 years.

Leiomyosarcoma is an aggressive tumor with poor prognosis, particularly if it is retroperitoneal and in a close contact with surrounding tissues and blood vessels. Symptoms occurring in those patients are often nonspecific, while clinical and ultrasound findings are inconclusive. Therefore, retroperitoneal tumor needs additional imaging, either CT or MRI. These findings enable an appropriate surgical approach/treatment. Making a prompt diagnosis and referring a patient to a specialized institution where the most radical surgical excision could be performed is of great importance.

Sufficiently extensive surgery with disease free margins make the prognosis better, when the possibility of recurrence is reduced. Both a delay in diagnosis and incomplete excision could adversely affect the outcome of the disease.

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