Uterine atypical leiomyoma: a rare feature of a common disease. Case reports and review of literature

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Abstract

We present two cases of uterine atypical leiomyoma (AL) and a review of the literature which refers to the question raised by the use of conservative and microinvasive techniques in the context of the recurrence and dissemination potential of these tumors. The AL cases were identified in two patients who underwent trachelectomy and subtotal hysterectomy. Interestingly, one AL developed on a residual cervix and showed a cystic aspect in ultrasound. This AL was associated with an intense necrosis which may have been caused by insufficient vascularisation in the cervix. For the second case, which was diagnosed with a huge tumor, a laparascopic subtotal hysterectomy with morcellation was planned. Because of technical problems the surgery was converted into laparotomy. In both cases the histology shows cells with pleomorphic nuclei, low mitotic activity and only for the first case the presence of intense aseptic necrosis. The reported cases showed no recurrence after a one-year follow-up. Because of the rare occurrence of these tumors it is very difficult to develop guidelines; that is why information can only be obtained from the few studies which exist in the medical databases. It is recommended that these rare cases should be reported in medical journals in order to cumulate the experience in the diagnosis and treatment of AL patients.
Keywords: *uterine atypical leiomyoma, microinvasive surgery, recurrence risk*

Introduction

Atypical leiomyomas are rare forms of uterine tumors[1] accounting for about 0.5% of uterine mesenchymal neoplasms and belong to a grey zone from a morphological and clinical point of view[2,3]. Because their occurrence in the practical activity is rare, only a few studies are published in PubMed and in these studies the follow-up data is often incomplete[4]. This is one reason which motivates the reporting of these rare cases in journals in order to cumulate the experience in diagnosis and treatment of these patients[5].

From a hystopathological point of view an atypical leiomyoma is characterized by a number of up to 10 mitoses/10 high power fields while the normal leiomyoma has none and the uterine sarcoma is a malignant form of tumor. Clinically, atypical leiomyomas are often associated with a tumor that increases quickly in volume compared to classic uterine leiomyoma. They are usually asymptomatic until they reach a size large enough to cause pain or bleeding[6].

Since atypical leiomyoma is an atypical histological form, it raises the question of the locally postoperative recurrence and metastasization. These phenomena could be augmented by using the conservative, minimally invasive surgical treatment techniques and the intraperitoneal morcellation techniques. According to the rarity of these cases it is very difficult to give guidance for the treatment and follow-up of these patients, especially if a minimally invasive therapy like myomectomy is considered, and the patients want to preserve their fertility. The data within the specialized literature is controversial. Some sources mention the absence of recurrence after conservative surgery techniques like hysteroscopic myomectomy[7] or morcellation techniques[8]. Other articles conclude that the behavior of atypical leiomyoma treated with myomectomy is still not exactly known, because only a low amount of data about the follow-up of such patients is available[9]. Contrary to previous opinions other authors present cases with metastatic leiomyoma[10], malignant transformation after total hysterectomy[11] or local recurrence in the site of myomectomy[9].

We herein present two cases of atypical leiomyoma and discuss the literature.

Case 1. A 54-year-old woman with a history of supracervical hysterectomy performed 15 years ago because of uterus leyomatosus, came to our clinic with pain in the left iliac fossa that started 2-3 years ago. The ultrasound and MRI showed a tumor with a diameter of 30 mm on the upper left side of the cervix with uncertain provenience suggesting an ovarian cyst (Figure 1). The patient underwent an open surgical removal of the cervix and of annexes. The removed tumor showed a cystic aspect (Figure 2). The pathological examination showed an atypical leiomyoma with cells with pleomorphic nuclei, up to 2 mitoses/10 high power fields and intense aseptic necrosis and necrobiosis phenomena (Figure 3) considered to be caused by the poor vascularization from the cervix. The patient was scheduled for follow-up every 6 months and showed no recurrence 1 year after surgery.

Case 2. A 37-year-old secundipara with finished family planning came to our clinic because of anemia, heavy menstrual bleeding and the presence of a huge tumor in the lower abdomen. The ultrasound showed a uterine leiomyoma with a diameter of 150/100/80 mm (Figure 4). A laparoscopic supraistmic hysterectomy was planned for the patient, but it was converted into open surgery because of technical difficulties. The pathology examination showed an atypical leiomyoma with a number of 2 mitoses/10 high power fields and atypical cells. The patient

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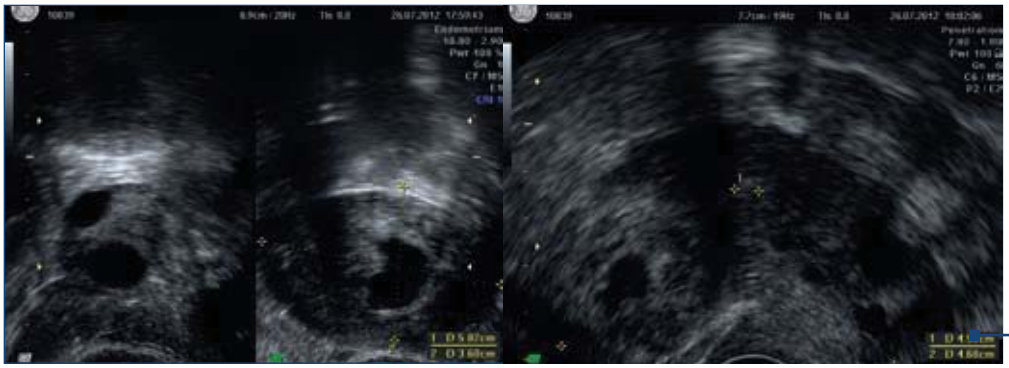

Figure 1. Ultrasound image of the atypical leiomyoma suggesting an ovarian cyst (58/36 mm) (case 1) (left - right ovary, middle - leiomyoma suggesting a cystic structure, right - cervix and cystic tumor)

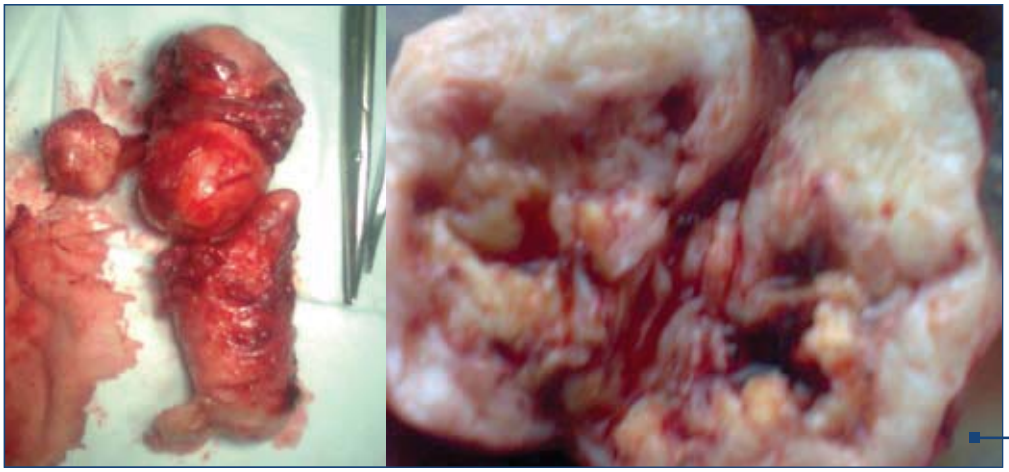

Figure 2. Macroscopic view of atypical leiomyoma removed from the cervix of the patient (case 1) (left - cervix and three leiomyomas, right - section through the cystic cavity of the atypical leiomyoma)

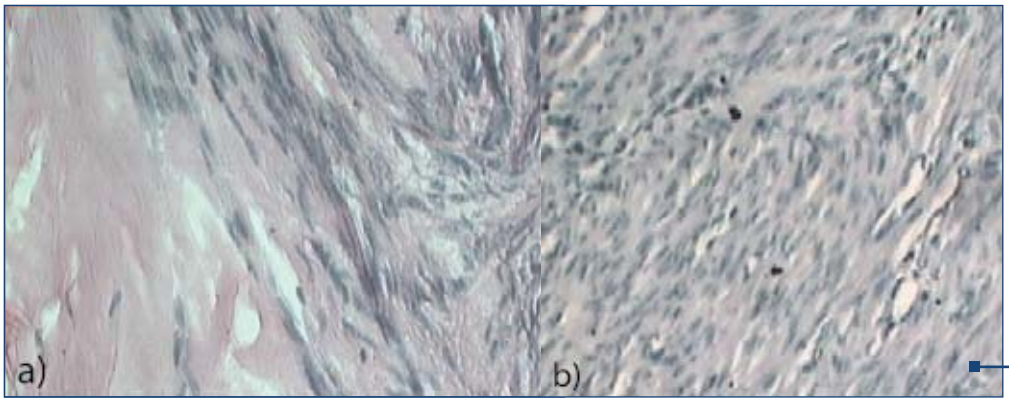

Figure 3. Hystopathological aspect (20x) of (a) atypical leiomyoma (case 1) with intensive necrosis and presence of atypia and (b) mitosis

was followed-up with ultrasound and MRI every 6 months and showed no complications 1 year after surgery.

Discussions

Atypical leiomyomata are a heterogeneous group of uterine mesenchymal tumors situated on a grey zone[2]. The hystopathological evaluation shows the presence of atypia, a low number of mitoses and no coagulated tumor necrosis[9]. Previous studies are controversial regarding the potential of local and distant dissemination of atypical leiomyoma. The majority of studies deny the long distance dissemination potential of atypical leiomyoma[7]. Other case reports present situations with distant dissemination in lungs or in iliac lymphatic nodes. In these cases a proper follow-up and prompt therapy give us the possibility to heal these complications[10]. Regarding the loco-regional relapse there is very little data in literature. It is generally accepted that if the uterus is totally removed, the risk of relapse is minimized. Careful follow-up or consideration of total hysterectomy for an atypical leiomyoma in which there was incomplete exclusion is also encouraged[12].

The diagnosis of these tumors is almost a postoperative surprise based on the hystopathological examination. Clini-

cally an atypical leiomyoma could be suspected if the tumor had a rapid growth and the routine examination could notice these changes. But this is not necessarily the development of all these tumors; in other situations, depending on vascularization, the growth could be only of small size (e.g. Case 2 with an atypical leiomyoma on residual cervix).

Considering the fact that the risk of relapse and metastasis is not clearly known, much attention must be given to the diagnosis, type of therapy and follow-up of these tumors. Since the diagnosis is mostly postoperative it raises the question if a conservative surgery technique (e.g. myomectomy or subtotal hysterectomy) is a sufficient therapy or a complete removal of the uterus is necessary.

The development of microinvasive laparoscopic surgery followed by morcellation techniques raise new hypotheses regarding the risk of loco-regional peritoneal and distance dissemination. A study on 1091 cases treated by laparoscopic hysterectomy shows that the occurrence of peritoneal recurrence after laparoscopic myomectomy of atypical leiomyoma is very rare[8].

The first case presented in our paper is a case of an atypical leiomyoma developed on a residual cervix after a subtotal supracervical hysterectomy. The imagistic diagnosis was very confusing both in ultrasound and MRI. Its imagistic and macroscopic aspect was like a cyst. Other authors reported similar cystic features in other patients, too[13]. The histopathology shows us up to 2 mitoses/10 high power fields, cellular atypia and intensive aseptic necrosis and necrobiosis probably consequent to an insufficient vascularization due to its origin in the residual cervix.

The second reported case came to our clinic with a huge uterine mass and with secondary anemia caused by menometrorrhagia. In this case we planned a laparoscopic subtotal hysterectomy but the operation was converted to open subtotal hysterectomy because of technical difficulties. During the procedure the myoma was partially fragmented and the macroscopic pieces were removed from the peritoneal cavity. Although postoperatively the histopathology showed an atypical leiomyoma with up to 2 mitoses/10 high power fields, the therapy consisted only in an open subtotal hysterectomy converted from a laparoscopic procedure.

Because of the hystopathological type of the tumor we raise the question whether subtotal hysterectomy is a sufficiently extensive treatment or a complete removal of the uterus would be necessary in order to minimize the risk of recurrence. Although the data in the literature regarding this subject is controversial, because of a very low reported rate of peritoneal or local relapse, our patient was not operated on again[14].

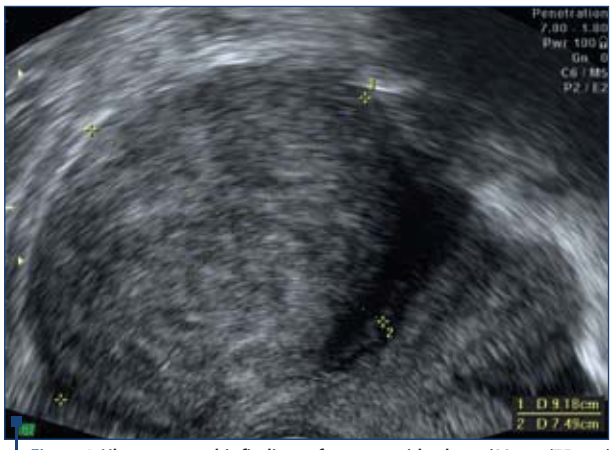

Figure 4. Ultrasonographic findings of a uterus with a huge (92 mm/75 mm) atypical leiomyoma (in case 2). (left side - myoma, right side - uterus)

Both patients were included in a 6-month follow-up program by ultrasound and MRI.

Conclusions

Our study presents two cases of patients with atypical uterine leiomyoma which raise difficulties in correct preoperative diagnosis, adequate therapy and follow-up. Not only the anatomopathological form but the imagistic aspect and the origin of the tumor could cover a variety of atypical aspects which can be pitfalls in preoperative diagnosis and intraoperative therapy. In the absence of a specific diagnostic marker, the diagnosis is a hystopathological surprise after the surgery was performed often in a conservative manner using conservative techniques such as subtotal hysterectomy, myomectomy or morcellation techniques which can spread cells in the peritoneum. These situations could bring to attention the possibility of local relapse or dissemination of atypical cells. In the absence of a guideline the therapy could be oriented according to a review of the significant studies published in the medical databases. The majority of authors recommend that the patients should be included in a carefully designed follow-up program or in case of a conservative surgical method the therapy should be completed to total removal of the uterus. ■

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