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Case Report

A giant myxoid/round-cell liposarcoma of the hip in a pregnant woman: a case report and literature review

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Abstract: Giant myxoid/round-cell liposarcoma (MRCL), a liposarcoma morphological variant, of the hip during pregnancy is rare. Low- and high-grade myxoid and round cells can be observed simultaneously, showing a progressive trend. Current treatments are limited; surgery is mainly used to reduce the tumor burden and relieve local symptoms. Here, we report a 36-year-old woman with a mass on the right hip present since trauma to this area and the right-side limb one year prior; the mass had been gradually growing for four months and increased significantly during pregnancy, accompanied by defecation difficulties. She underwent a resection of the mass; a histopathological examination confirmed MRCL. She chose to terminate the pregnancy and received radiotherapy (60 Gy). She was followed up for 37 months, with no signs of recurrence or metastasis. Here we report a rare case of a pregnant patient with giant MRCL of the hip. Preoperative biopsy should be performed; preoperative angiography and embolization can be conducted if the mass is large with a clear blood flow source. Although not reported previously, we applied this treatment and achieved satisfactory results, confirming its feasibility and effectiveness. This report provides new ideas for future treatment, though more cases are needed for further research.

Keywords: Myxoid/round-cell liposarcoma, pregnancy, female

Introduction

Liposarcoma is a malignant tumor composed of abnormal adipocytes, and diversity is a major feature of this disease. The pathological changes exhibited by abnormal adipocytes include varying degrees of differentiation and atypia. Liposarcoma is one of the most common soft tissue sarcomas and consists of five histological types: well-differentiated, myxoid, round cell, pleomorphic, and dedifferentiated (WHO classification, 2002) [1]. Liposarcoma usually occurs in the fat-rich parts of the lower limbs, the abdomen and the retroperitoneum. Although myxoid/round-cell liposarcoma (MRCL) is common among these types, giant MRCL of the hip in pregnant women is rare. In addition, treatments for MRCL are currently limited, and surgery is the main method used to reduce the tumor burden and relieve local symptoms. However, the risks and difficulties of surgery are greater when the tumor is larger, and there are no standard surgical treatments.

In this paper, we report a rare case of a pregnant patient with giant MRCL of the hip. The chief clinical manifestation was that the tumor was found to increase significantly in size during pregnancy, and was accompanied by pain, lethargy, and defecation difficulties. Because the mass was large, we applied an operative treatment, including preoperative angiography and embolization, and achieved satisfactory results.

Case presentation

A 36-year-old woman presented with a one-year history of a mass on her right hip that had gradually been growing for four months.

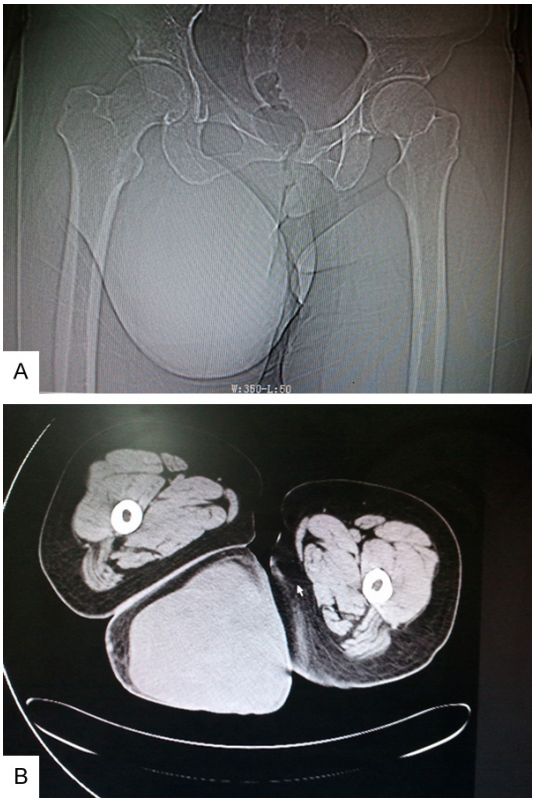

Figure 1. X-ray and CT findings. A. X-ray showing a large mass in the right hip. B. CT showing a large conglomeration shadow in the right hip, protruding into the pelvic cavity and causing local rectal and uterine compression. CT = computed tomography.

The patient had a history of right hip and right-side limb trauma one year prior. After the trauma, she found an egg-size mass that was accompanied by pain, and the surface of the mass was purple. Four months prior, the tumor was discovered to be significantly increasing in size during her pregnancy. At the time of admission, the mass was approximately 25×10 cm in size and was accompanied by pain, lethargy, and defecation difficulties. The physical examination of the right hip showed a significantly convex mass that was approximately 25×12×10 cm in size and soft in texture; the surface was smooth, the boundary was clear, and the mobility was good, with palpable lymph nodes in the inguinal region. Ultrasonography revealed a hypoechoic subcutaneous mass in the right hip (with a high likelihood of having originated from the muscularis), with a clear boundary, a visible envelope, a lobulated morphology, a width of approximately 16.0 cm, uneven echo signals, and dark liquid areas, indicating internal venous and low-resistance-type arterial blood flow. A clinical diagnosis of rhabdomyoma or sarcoma of the right hip was made. The x-ray and computed tomography (CT) findings were as follows: a large conglomeration shadow in the right hip protruding into the pelvic cavity with local rectal and uterine compression and a small section with a slightly increased density; the shadow was 22×15×12 cm in size, and the average CT value was 25 HU. The maximum diameter was approximately 23 cm (**Figure 1A**, **1B**). A clinical diagnosis of a benign tumor in the right hip was made.

However, on the fifth day after admission, the skin of the patient's right hip ruptured and bled after activity. As the CT showed a clear source of blood flow, the patient underwent inferior mesenteric artery and double internal iliac artery angiography and embolization. To clarify the nature of the tumor, she underwent a biopsy of the right hip mass, and a rapid frozen histopathological examination confirmed that the tissue was composed of small round cells of the same size, accompanied by necrosis, which tended to indicate malignant soft tissue tumors. Subsequently, a resection of the large right hip mass was performed, and the histopathological examination confirmed MRCL (**Figure 2A**, **2B**). The immunohistochemical staining results of the tumor were as follows: S-100 (-) (**Figure 3A**), desmin (Des) (-) (**Figure 3B**), smooth muscle actin (SMA) (-) (**Figure 3C**), and an antigen Ki-67 (Ki-67) index of 3% (**Figure 3D**). Our patient chose to terminate the pregnancy and received radiotherapy (60 Gy) after complete tumor resection and incision healing. The patient was followed for 37 months, and no signs of recurrence or metastasis were observed.

Discussion

Liposarcoma is a malignant tumor composed of abnormal adipocytes. There are five histological types of liposarcoma: well-differentiated, myxoid, round cell, pleomorphic, and dedifferentiated (WHO classification, 2002) [1]. Different histological subtypes of liposarcoma have different prognoses. The important related factors include the resection margin, continuous organ resection and the age of the patient [2]. MRCL is a common subtype of liposarcoma, accounting for more than 33% of lipo-

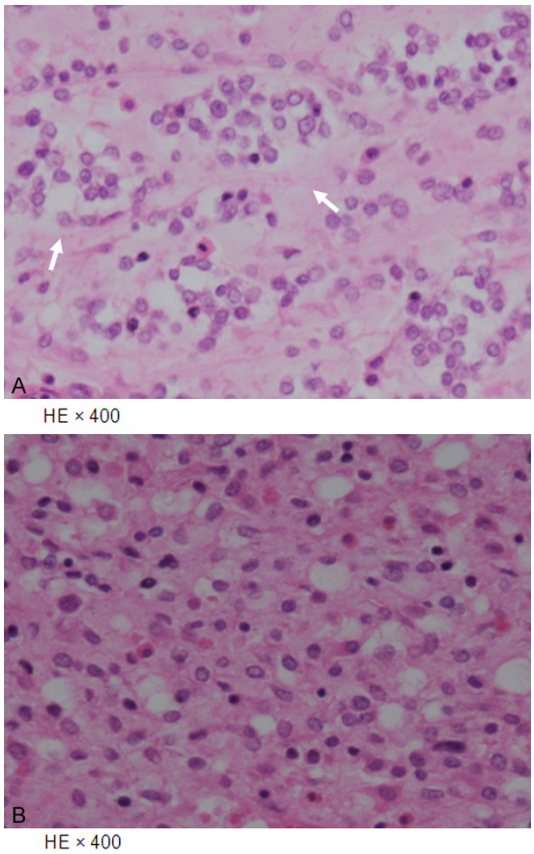

Figure 2. Histopathology using hematoxylin and eosin staining (400×). A. Stained section showing tissue composed of small round cells of the same size, accompanied by necrosis, which tends to suggest malignant soft tissue tumors. B. Myxoid/round-cell liposarcoma was confirmed.

sarcomas and for approximately 10% of all soft tissue sarcomas [3]. MRCL is a morphological variant of liposarcoma, with a proportion of round-cell components greater than 5% [4]. Importantly, in MRCL, low- and high-grade myxoid cells and round cells can be observed simultaneously, showing a trend of progression. The incidence of MRCL peaks between 40 and 50 years old and is slightly more common in males than females [5]. Indeed, MRCL is rare in female patients younger than 30 years, and it is very rare for a pregnancy to coexist with MRCL. In the current English-language literature, fewer than 15 cases have been published. Additionally, no solid evidence of an increased incidence or poorer outcome has been revealed. The clinical manifestation of MRCL is a large painless mass usually located in deep soft tissue, and more than 67% of cases occur in the lower limbs [6]. MRCL rarely appears in the hips. Interestingly, our case differs from previous reports in that a large mass was present in the right hip in a relatively young woman; the tumor was also found to increase significantly in size during pregnancy, and was accompanied by pain, lethargy, and defecation difficulties. The clinical manifestation in our case differed from that in previous cases, and the clinical characteristics were similar to those of some other diseases, such as angiolipoma, leiomyoma, or schwannoma. Therefore, additional methods were needed to determine the nature of the tumor.

The diagnosis of MRCL depends on its clinical manifestations as well as the imaging and histopathology findings [7]. The clinical manifestations are described above. The relevant imaging methods include X-ray, CT, and magnetic resonance imaging (MRI). These examinations should be performed before the biopsy and as early as possible. For surgeons, the above techniques are necessary to measure the size of a tumor, determine its location, and understand the preoperative conditions. In particular, CT and MRI are important and reliable means of examination for the diagnosis of MRCL [8], providing surgeons with useful information to accurately distinguish between the different types of tumors. Moreover, these assessment methods are extremely important for obtaining prebiopsy information [9, 10]. However, needle biopsy or surgery remains the most reliable diagnostic method for MRCL, as the gold standard is still histopathological examination of a surgical specimen [11]. The evidence needed for differential diagnosis stems from immunohistochemical analysis of S-100, Des and SMA. Immunohistochemical positivity for S-100, Des, or SMA suggests that the sarcoma is derived from nerve tissue, striated muscle tissue, or smooth muscle tissue, respectively. The final diagnosis of MRCL is established according to the morphology of the tumor tissue, an examination of the tumor cells through microscopy, and the immunohistochemistry results. Our patient was diagnosed with MRCL according to the above criteria. Preoperative biopsy is a great necessity for surgeons to determine the nature of a tumor, choose the surgical treatment, and assess the risks and difficulties of surgery. Therefore, a preoperative biopsy should be performed.

The principle of treatment is comprehensive treatment based on surgery. Currently, complete surgical resection for MRCL is the main

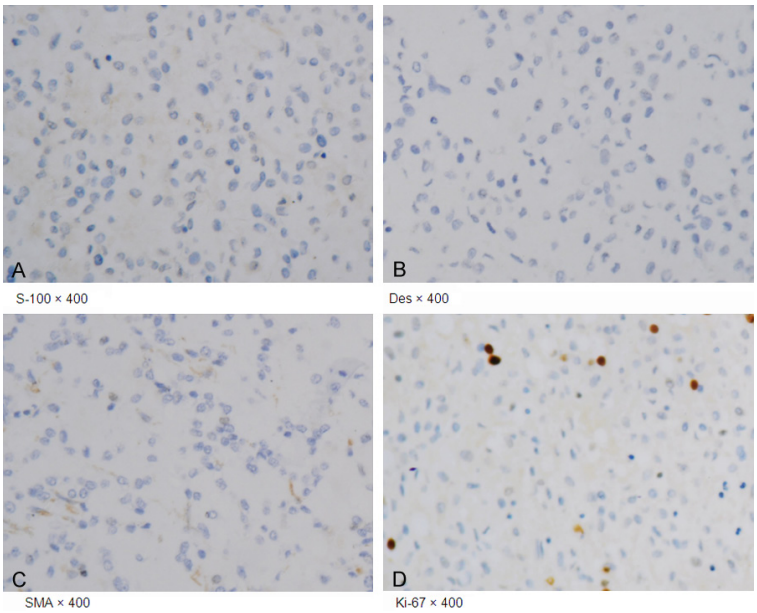

Figure 3. Immunohistochemical analysis (400×). The tumor cells were negative for S-100 (A), Des (B) and SMA (C). The Ki-67 index was 3% (D). S-100 = soluble protein 100, Des = desmin, SMA = smooth muscle actin, Ki-67 = antigen Ki-67.

treatment for reducing the tumor burden and alleviating the local symptoms [12]. Other treatments include radiotherapy and chemotherapy [13]. Surgery must include a wide excision with negative margins (R0). If the mass is large and CT or MRI shows a clear source of blood flow, preoperative arteriography and embolization can be performed. This method can reduce the mass size, thereby reducing the risks and difficulties of surgery. Our patient underwent inferior mesenteric artery and double internal iliac artery angiography and embolization, and satisfactory results were achieved. This approach has not been reported in previous studies. Moreover, radiotherapy is a standard and effective treatment for patients with locally advanced disease or unclear surgical margins. Although it can decrease the local and distant recurrence rates, it cannot alter the overall survival rate of MRCL [9, 10]. Some current studies have suggested that adjuvant chemotherapy may reduce distant and local recurrence rates in high-risk patients. Regardless, there is a lack of robust evidence based on large-sample clinical studies [14]. Current clinical practice guidelines recommend adjuvant chemotherapy as an option for high-risk patients with MRCL rather than as a standard therapy [4]. In clinical work, it is clear that existing treatments for MRCL are limited, and new research is needed to develop new treatments. Pollack reported that NY-ESO-1 is an important target antigen for the treatment of MRCL [3]. Other studies have found that if the expression of carboxypeptidase M (CPM) is decreased, EGFR signaling will be inhibited, and liposarcoma cells will be induced to undergo apoptosis [15, 16]. These findings may be helpful for obtaining new molecular therapies for MRCL. Based on our case, hormone level changes during pregnancy might stimulate rapid tumor growth. Nonetheless, this hypothesis needs to be verified in future research. More significantly, whether pregnancy can be continued during treatment of the disease needs to be further explored. Ultimately, our patient received radiotherapy after surgery according to these criteria and principles.

In conclusion, we report a rare case of a pregnant patient with giant MRCL of the hip. Clinically, MRCL manifests as a large, painless mass, usually located in deep soft tissue. The diagnosis of MRCL depends on its clinical manifestations and imaging and histopathological findings. Among these examination methods, preoperative biopsy should be performed. The principle of treatment is comprehensive treatment based on surgery. Other treatments include radiotherapy and chemotherapy. In particular, if the mass is large and there is a clear source of blood flow, preoperative arteriography and embolization can be carried out. This method can reduce the mass size, thus decreasing the risks and difficulties of surgery. Our patient underwent inferior mesenteric artery and double internal iliac artery angiography and embolization, which yielded satisfactory results, and this has not been reported in previous studies. Our case confirms that this is a feasible and effective method.

However, there are some limitations to this work. The results of this study do not demonstrate that elevated hormone levels stimulate

rapid tumor growth. In the future, we will explore the implications of hormone level changes in MRCL progression using basic research. Furthermore, the small number of clinical cases and retrospective analysis should also be considered limiting factors. We will continue to study similar cases after obtaining written informed consent.

Acknowledgements

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Disclosure of conflict of interest

None.

Abbreviations

S-100, soluble protein 100; Des, desmin; SMA, smooth muscle actin; Ki-67, antigen Ki-67; MRCL, myxoid/round-cell liposarcoma; NY-ESO-1, cancer-testis antigen (also known as cancer germ cell antigen); LPS, liposarcoma; CPM, carboxypeptidase M; EGFR, epidermal growth factor receptor.

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