Contents lists available at ScienceDirect



Case report

International Journal of Surgery Case Reports



journal homepage: www.elsevier.com/locate/ijscr

Occult leiomyosarcoma simulating malignant ovarian tumor: A case report

Herlangga Pramaditya, Brahmana Askandar Tjokroprawiro

Department of Obstetrics and Gynecology, Faculty of Medicine, Universitas Airlangga/Dr. Soetomo General Academic Hospital, Surabaya, Indonesia

ARTICLE INFO

Giant leiomyosarcoma

Keywords:

Ovarian tumor

Malignancy

Re-surgery

Case report

ABSTRACT

Introduction and importance: Leiomyosarcoma (LMS) is a malignant tumor of the uterine smooth muscle. It is highly aggressive and accounts for 1 % of all uterine malignancies. Symptoms often include a rapidly growing mass, with an average course of illness ranging from six to eight months. Leiomyosarcomas are rare in younger individuals. *Case presentation:* A 31-year-old nulliparous woman was diagnosed with a suspected malignant ovarian tumor. A

Case presentation: A 31-year-old numperous woman was dragnosed with a suspected marginant ovarian tumor. A unilateral salpingo-oophorectomy and surgical staging were planned to preserve the patient's fertility. However, the large solid mass was found to be arising from the uterus during surgery. In light of the preference for fertility preservation, a myomectomy was performed, with subsequent pathology results showing a malignant leio-myosarcoma. Following counseling and discussion with the patient and her spouse, follow-up surgery was performed to ensure a free surgical margin.

Clinical discussion: Leiomyosarcomas at a young age is very rare, and early diagnosis is quite a challenge, especially when the tumor characteristics are not atypical. Thus, surgical evaluation and staging are paramount. *Conclusion:* Surgical diagnosis of any mass with a history of rapid enlargement and characteristics of malignancy is very important, especially when anatomical landmarks are distorted due to the enlarged mass.

1. Introduction

Leiomyosarcoma is a malignant tumor of the uterine smooth muscle. It is highly aggressive and accounts for 1 % of all uterine malignancies. The incidence ranges from 0.35 to 0.64 in 100,000 people in the United States, with the majority of cases occurring in the perimenopausal to postmenopausal ages [1]. Leiomyosarcoma typically does not appear in individuals younger than 43–53 years [2,3]. Symptoms often include a rapidly growing mass, with an average course of illness ranging from six to eight months [4]. During surgery, a flaky and easily bleeding mass is usually found, but it is not uncommon for leiomyosarcoma to be diagnosed only after a pathological examination. This usually indicates the need for follow-up surgery. We present the case of a woman diagnosed with a suspected malignant ovarian tumor. During surgery, it was found that the mass originated from the uterus and had the characteristics of uterine leiomyoma; however, pathology results following surgery reported a leiomyosarcoma. This case report was prepared per the SCARE guidelines [5].

2. Case presentation

We present a 31-year-old nulliparous woman of Asian ethnicity who was married for eight years and received preconception counseling in 2015. An ovarian cyst measuring 3 cm in the right ovary was found during the examination. The patient was advised to undergo surgery but refused. During the following years, the patient had never been checked by an obstetrician-gynecologist (OB/GYN), there were no notable changes or complaints in the patient's menstrual cycle, and she did not report taking any medications or undergoing any surgical procedure. The patient also denied any history of chronic or familial disease and never consumed any alcohol or smoking.

Later the patient presented with fullness and enlargement of the abdomen beginning in November 2021. At that time, she was examined by an OB/GYN at the referring hospital, and a cystic solid mass measuring $17.63 \times 20 \times 17.3$ cm was found in the pelvic cavity. The mass was suspected to originate from the adnexa. Hydronephrosis and bilateral hydroureter were also noted, likely caused by obstruction due to the enlarging mass. The patient was referred to our gynecological oncology polyclinic for follow-up. Upon examination, her BMI was 24,

https://doi.org/10.1016/j.ijscr.2022.107758

Received 18 August 2022; Received in revised form 14 October 2022; Accepted 21 October 2022 Available online 24 October 2022

^{*} Corresponding author at: Department of Obstetrics and Gynecology, Faculty of Medicine, Universitas Airlangga/Dr. Soetomo General Academic Hospital, Jl. Prof. Dr. Moestopo 6-8, Surabaya 60286, Indonesia.

E-mail address: brahmanaaskandar@fk.unair.ac.id (B.A. Tjokroprawiro).

^{2210-2612/© 2022} Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

her BP was 120/70 mm Hg, her pulse rate was 82 beats/min, and her respiratory rate was 20 breaths per minute. No anemia, icterus, cyanosis, edema, or significant lymphadenopathy was present. The cardiovascular and neurological examination was normal. Upon physical examination at our hospital, we found a mobile abdominal mass measuring 25-30 cm in diameter (Fig. 1). Gynecological examination revealed a normal external genitalia with a soft cervix; however, a 30 cm-mass was palpable in the abdomen, which we were unable to determine the origin of the mass, the lower pole of the tumor was palpable from the vaginal examination.

The patient underwent laboratory and radiological evaluation. Tests revealed normal kidney function. The alpha-fetoprotein level was average (AFP = 3.3), and carcinoembryonic antigen (CEA) was low, with slightly elevated CA-125 levels (CEA < 0.5; CA-125 = 58.1). An abdominal computed tomography (CT) scan revealed a contrastenhanced cystic mass with a solid component suspected to originate from the adnexa and measuring 19.9 \times 27.3 \times 19.9 cm. The mass displaced the uterus anteriorly with ill-defined borders, and bilateral hydronephrosis was also found on the CT scan (Fig. 2). Considering the rapidly expanding tumor mass and the characteristics suggestive of malignancy, we planned a unilateral salpingo-oophorectomy and surgical staging while preserving the patient's fertility.

During surgery, a large solid non-friable mass was found originating from the uterus, 40 cm in diameter (Fig. 3). An adenomyomectomy followed by uterine repair was performed (Fig. 4A), which was decided during surgery due to the difference between our initial management plan, which mainly because our preoperative diagnosis was a malignant





Fig. 1. Grossly enlarged abdomen (pre operative).

ovarian tumor. Upon evaluation, active bleeding in the infundibulopelvic ligament was observed. Ligation was performed but failed to control the bleeding, and a unilateral salpingo-oophorectomy on the hemorrhage side was performed. The surgery was performed by a senior gynecology oncologist consultant accompanied by a second-year trainee resident. The total blood loss volume was 1800 cm³ with the surgery lasted for 3 h and 25 min. The patient received treatment and blood transfusion for a few days postoperatively. She was treated for five days and then was discharged in good condition. There were no complications even until subsequent visits to our oncology outpatient clinic.

Postoperative pathology results were consistent with leiomyosarcoma and confirmed by immunohistochemistry (IHC) evaluation, which was positive on the examination of antibody caldesmon, cyclin D1, SMA, and vimentin (Fig. 5). The patient then was decided to undergo additional surgery to optimize the previous procedure and to maximize a free surgical margin. Further counseling and discussion with the patient's husband were held due to the patient being nulliparous and still hoping for future pregnancy. However, after counseling, both agreed to undergo surgery and a hysterectomy. We managed to achieve a free surgical margin that was confirmed by histopathological examination. The patient has undergone re-surgery, and we performed a total hysterectomy, salpingo-oophorectomy dextra, and omental resection because the tumor was attached to the omentum (Fig. 4B). The patient was discharged from the hospital in good condition. Currently, the patient is well and still visiting our oncology clinic routinely. Currently, she is on a 3-monthly evaluation. Further evaluation with a PET scan to evaluate metastatic sites is ideally needed. However, due to the unavailability of the facility in our institution, we could not perform the procedure.

3. Discussion

Leiomyosarcoma is a sarcoma that originates from the smooth muscle of the uterus. It is an extremely rare uterine cancer, found in less than one case per 100,000 people in the United States. Jayasri et al. reported that 15 % of leiomyosarcoma cases occurred in individuals under 40 years of age [4]. Ozcan et al. said that even adolescent leiomyosarcoma is possible, although very rare [6]. In our institutions, leiomyosarcoma is rare and only reports 2-3 cases yearly. Occult cases occur when leiomyosarcoma surgery is performed initially because of presumed benign disease [7].

Leiomyosarcoma sometimes grows to form an irregular mass and enlarges towards the abdominal cavity. However, in some cases, a solid mass can be found, as in our case. Due to its unusual characteristics, the patient was misdiagnosed twice. The first was at the preoperative diagnosis, where, despite imaging with an abdominal CT scan, the mass was so large that it filled the pelvic cavity, making it difficult to distinguish it from an ovarian mass. The second misdiagnosis occurred at the time of surgery, based on the characteristics of the mass, which were unlike leiomyosarcoma and led us to suspect it to be benign. Only adenomyomectomy was performed, and no frozen sections were obtained. When the pathological results showed leiomyosarcoma, resurgery with a total abdominal hysterectomy and bilateral salpingooophorectomy minimized recurrence.

Magnetic resonance imaging (MRI) with contrast can diagnose a leiomyosarcoma preoperatively. In our case, only a contrast-enhanced CT scan of the abdomen was performed, and the results suggested an ovarian tumor [2]. Tantitamit et al. reported that preoperative diagnosis of leiomyosarcoma by clinical examination is complicated, and a rapidly growing mass is not pathognomonic of leiomyosarcoma. It is not easy to distinguish a leiomyosarcoma from a degenerating uterine leiomyoma [8,9]. Although MRI is not always performed for every leiomyoma case, it may be considered in patients with large and rapidly growing tumors [10]. Other options include tumor markers such as CA-125 and lactate dehydrogenase (LDH) have poor sensitivity and specificity. CA-125 is more specific for ovarian cancer and also needs to improve by



Fig. 2. Imaging CT scan abdomen-pelvic with contrast (pre operation first surgery) 1.



Fig. 3. During the first surgery, the tumor mass was obtained from the uterus.



Fig. 4. A uterus after myomectomy – cytoreduction. B surgical findings second surgery.



Fig. 5. A. IHC caldesmon positive. B. IHC cyclin D1 positive. C. IHC SMA positive. D. IHC vimentin.

combining with other marker [2,11,12].

The primary management of leiomyosarcoma, diagnosed or suspected preoperatively, involves total abdominal hysterectomy, bilateral salpingo-oophorectomy, and debulking all masses outside the uterus. The preservation of ovaries can be performed in young women, but there is no place for fertility-preserving surgery. Generally, in the era of the covid pandemic, our hospital services are slightly hampered and affect the patient's psychology related to the operation queue and the decision to undergo a hysterectomy. Achieving R0 resection is challenging because of the large tumors and involvement of the retroperitoneal spaces [13,14]. Leiomyosarcoma generally has metastases in the surrounding area and the lung. Surgery patients generally continue with a palliative approach because leiomyosarcoma is neither chemosensitive nor radiosensitive, and the recurrence rate is still high [4].

Lawlor et al. reported that in patients who underwent inadequate surgery due to misdiagnosis of leiomyosarcoma, the prognosis was poor (12–25 % 5-year survival rate) [2]. The problem in our case was that due to its dense characteristics, which highly resemble uterine leiomyoma, a myomectomy procedure was previously performed, resulting in the morcellation of the cancerous tissue. Therefore, it is important to perform follow-up surgery immediately to remove all tissues that have the potential to cause spread [8].

The diagnosis of uterine leiomyosarcoma based on histopathological examination is difficult. In IHC, 50 % of leiomyosarcoma cases are positive for an anti-desmin antibody with good specificity. The other markers expressed were global muscle actin (HHF35), smooth muscle actin (SMA), and h-caldesmon, which have good specificity and are expressed in 85 % of leiomyosarcoma cases. Here, the diagnosis of leiomyosarcoma was confirmed by IHC, which was positive on the examination of IHC caldesmon, cyclin D1, SMA, and vimentin (Fig. 5a–d).

4. Conclusion

Leiomyosarcoma is a very rare malignant tumor of the uterus, sometimes seen in young women. When exophytic growth develops in the uterine cavity, it isn't easy to make a definite diagnosis preoperative, even with adequate imaging. Diagnosis at the time of surgery is essential. Any mass with a history of rapid enlargement should be suspected as a malignancy, even if the clinical findings are not supported, so that an appropriate and efficient surgical decision can be made. Counseling regarding postoperative diagnosis, further management plans, and a fertility-sparing approach is also essential and poses a challenge.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Consent

Written informed consent was obtained from the patient to publish this case report and accompanying images. On request, a copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethical approval

This study has been approved by the hospital ethic board, and certification is available should there is a need to provide it.

Funding

This study was funded solely by the researcher.

Guarantor

Brahmana Askandar Tjokroprawiro.

Research registration number

None because this report is not the first in man.

CRediT authorship contribution statement

The first and the second author perform the surgery and analyze and discuss this report. We also note the valuable expertise of Grace Ariani, MD, for the pathology specimen analysis and interpretation, which was essential in determining the patient's diagnosis and management.

Declaration of competing interest

We declare that there is no conflict of interest with any parties involved in this study.

References

- [1] A.S. Brohl, L. Li, V. Andikyan, S.G. Običan, A. Cioffi, K. Hao, et al., Age-stratified risk of unexpected uterine sarcoma following surgery for presumed benign leiomyoma, Available from, The Oncologist [Internet]. 20 (4) (2015 Apr 1) 433–439, https://academic.oup.com/oncolo/article/20/4/433/6399011.
- [2] H. Lawlor, A. Ward, A. Maclean, S. Lane, M. Adishesh, S. Taylor, et al., Developing a preoperative algorithm for the diagnosis of uterine leiomyosarcoma, Diagnostics [Internet]. 10 (10) (2020) 735. Available from: https://www.mdpi.com/ 2075-4418/10/10/735.
- [3] J. Mishra, S. Bhattacharya, A. Pandia, A. Padhy, M. Mahapatra, J. Mohapatra, Leiomyosarcoma of uterus in a nulliparous female: mimicking as ovarian malignancy, Available from, Asian Journal of Oncology [Internet]. 07 (03) (2021 Sep 29) 156–159, http://www.thieme-connect.de/DOI/DOI?10.1055/s-004 1-1730098.
- [4] S. Jayasri, B.N. Devi, A rare case report of uterine leiomyosarcoma in young age, IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) [Internet]. 18 (3) (2019) 1–3. Available from: https://www.iosrjournals.org/iosr-jdms/papers/Vo l18-issue3/Series-7/A1803070103.pdf.
- [5] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, et al., The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, Available from, International Journal of Surgery [Internet]. 84 (2020 Dec) 226–230, https://linkinghub.elsevier.com/retrieve/pii/S1743919 120307718.
- [6] J. Özcan, Ö. Dülger, L. Küpelioğlu, A.İ. Gönenç, A. Erşahin, Uterine sarcoma in a 14year-old girl presenting with uterine rupture, Available from, Gynecologic Oncology Reports [Internet]. 10 (2014 Dec) 44–46, https://linkinghub.elsevier. com/retrieve/pii/S2211338X14000192.
- [7] A. Ludwin, I. Gawron, K. Pityński, Occult uterine leiomyosarcoma in women undergoing abdominal and minimally invasive surgeries for myomas, Available from, Ginekologia Polska [Internet]. 89 (10) (2018 Oct 31) 546–552, https://jo urnals.viamedica.pl/ginekologia_polska/article/view/58931.
- [8] T. Tantitamit, K.-G. Huang, M. Manopunya, C.-F. Yen, Outcome and management of uterine leiomyosarcoma treated following surgery for presumed benign disease: review of literature, Gynecology and Minimally Invasive Therapy [Internet]. 7(2): 47 (2018). Available from: http://www.e-gmit.com/text.asp?2018/7/2/47/ 231754.
- [9] D. DeMulder, S.M. Ascher, Uterine leiomyosarcoma: can MRI differentiate leiomyosarcoma from benign leiomyoma before Treatment?, Available from, American Journal of Roentgenology [Internet]. 211 (6) (2018 Dec) 1405–1415, https://www.ajronline.org/doi/10.2214/AJR.17.19234.
- [10] C.M. Yeni, C.R. Maharani, H. Supriadi, Unexpected leiomyosarcoma after total abdominal hysterectomy with indication uterine myoma in nullipara: a case report, Bali Medical Journal [Internet]. 10 (2) (2021) 775. Available from: https://www. balimedicaljournal.org/index.php/bmj/article/view/2395.
- [11] P. Perbowo, B. Askandar, KLK6 as a predictor of ovarian malignancy, Available from, Medicina Moderna - Modern Medicine [Internet]. 26 (4) (2019 Dec 30) 189–195, https://medicinamoderna.ro/wp-content/uploads/2019/12/RMM_art-5-KLK6-as-a-Predictor-of-Ovarian-Malignancy.pdf.
- [12] Yildiz G, Gundogdu EC, Basol G, Mat E, Kale A. Uterine Sarcomas: 10 Years of Experience at a Single Institution/Uterin Sarkomlar: Tek Merkezin 10 Yillik Deneyimi. Southern Clinics of Istanbul Eurasia (SCIE) [Internet]. 2020 Oct 4;31:8 . Available from: https://link.gale.com/apps/doc/A648842637/AONE? u=googlescholar&sid=googleScholar&xid=f5ac7b33.
- [13] S. George, C. Serrano, M.L. Hensley, I. Ray-Coquard, Soft tissue and uterine leiomyosarcoma, Available from, Journal of Clinical Oncology [Internet]. 36 (2) (2018 Jan 10) 144–150, https://ascopubs.org/doi/10.1200/JCO.2017.75.9845.
- [14] Pratiktio NB, Tjokroprawiro BA, Utomo B, Muhdi N. Psychosocial perspective of patients with gynecological cancer towards medical services during the coronavirus disease pandemic at RSUD Dr. Soetomo, Surabaya, Indonesia. International journal of health sciences [Internet]. 2022 Jul 30;6(March): 3192–204. Available from: https://sciencescholar.us/journal/index.php/ijhs/ article/view/11295.