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A rare Case of Uterine Leiomyosarcoma in a Virgin Woman: a case report

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ABSTRACT

Leiomyosarcoma (LMS) compared to other types of uterine cancers is rare uterine malignancy with a very aggressive course and poor prognosis. We report a confirmed histopathologically uterine leiomyosarcoma by IHC study in virgin woman. Case A: 58-year-old woman was admitted with a history of abdominal enlargement with 4-5 month duration of lower abdominal lump. Tumor marker CA-125 was raised. She underwent hysterectomy, bilateral salpingo-oophorectomy and resection of degenerated leiomyoma's. Conclusion: The gold standard of uterine leiomyosarcoma treatment is surgery. The prognosis of uterine leiomyosarcoma depends on the extent of disease at the time of diagnosis and the mitotic index in women.

Key words: Leiomyosarcoma, uterine, Virgin Woman

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1. INTRODUCTION

Uterine leiomyosarcoma (LMS) is an uncommon malignancy. The incidence is 0.67 per 100,000 in above 20 years old (1, 2), it represents 1-1.3% of all uterine malignancies and about 5% of uterine sarcomas (1-4). LMS arises from the smooth muscle cells within the myometrium. Clinically, it is aggressive smooth muscle malignancy. 2 -6% of uterine malignancies have poor prognosis and annual incidence is 1.7 per women (3). Uterine LMS can be usually seen during the fifth or sixth decades of life (5). Incidence of uterine LMS in surgical cases is less than 0.5 %. This risk is increased with age, so that in women older than 60 years is 1.7% (6). The most common clinical presentations in uterine LMS include uterine bleeding, pelvic pain and or pelvic mass (1-3, 7). Preoperatively, diagnose of uterine LMS is very difficult, even if diagnostic imaging and endometrial sample have been performed preoperatively. Frozen section is not always decisive intra operatively (1, 6). First, the diagnosis of LMS is established by a pathologist or after surgical removal of a presumed benign uterine mass (6, 8). . Poorly known responsible factors for the initial neoplastic myometrial transformation is assumed that each leiomyoma arises from a single cell in the myometrium (3,

9). LMS cases that are metastases, particularly to the lungs have been widely described. We report a case of LMS confirmed by biopsy in 58 years virgin woman. This patient did not have any history of uterine bleeding which is the usual in uterine sarcoma. This case was considered due to the rarity of the disease, and so this woman has never ever married and its age is high.

2. The Case

A 58 years old postmenopausal virgin woman with a history of abdominal enlargement and 4-5 month duration of lower abdominal lump had referred to gynecology department of Imam Reza Hospital in kermanshah. She was deaf and not married. So, the history of disease was taken from her brother's wife. Symptoms include nausea, vomiting, constipation, satiety and pressure sensation in urinary tract. She lost 5-6 Kg of weight two month ago. She had no history of vaginal bleeding or abdominal pain. Abdominal examination showed an abdominal mass, As much as 26 weeks of gestational age. Her general physical examination was unremarkable. There was no hepato-splenomegaly. The mass was tough. Ultra sonogram showed a large heterogeneous calcified well defined mass

measuring 20*16 cm in size in the right region of pelvis. We watched malignant tumor in right ovarian (Figure 1).

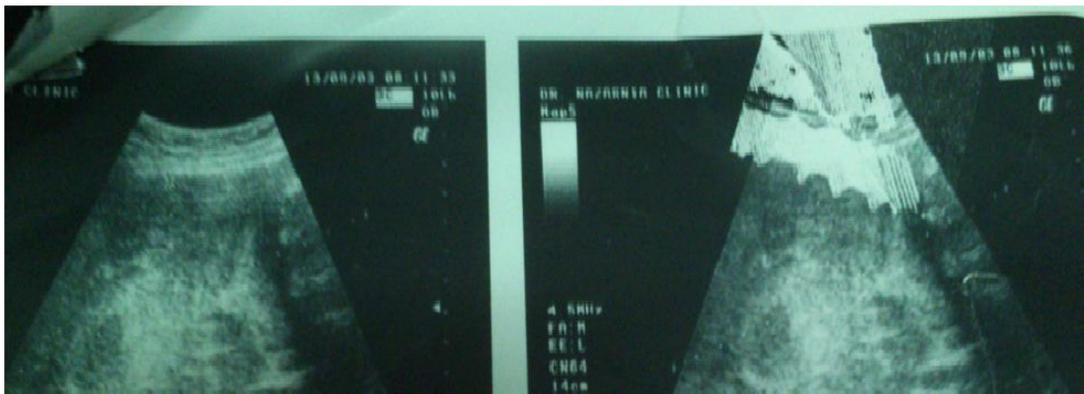


Figure 1. Trans abdominal sonogram of a pelvis revealing a large heterogeneous calcified well defined mass in the right region of pelvis

Other organs such as liver, spleen, and kidney were normal in sonography. Ascites was not found. CA-125 was 43ng/ml (0-35 ngr/ml). Other laboratory findings were normal. Clinical diagnosis was made for ovarian neoplasm and she was advised exploratory laparotomy (total abdominal hysterectomy and bilateral salpango-oophorectomy (TAH+BSO). Bronchovascular marking was prominent in chest x ray. Total abdominal hysterectomy with BSO was performed by general anesthesia. Abdomen was opened by vertical midline incision.

After opening the abdomen a hemorrhagic mass measuring 14cm in diameter attached to left posterior aspect of uterine fundus with a 4cm pedicle was found. Extending to the right side of abdomen was observed. Both ovaries and the fallopian tubes were normal. The degenerated subserosalmyoma was resected. In another intra mural huge myoma (9cm) in diameter in the anterior body of uterus was also resected (figure2- 5).



Figure2. Uterus pedunculated



Figure3. Lobulated mass arise from the



Figure4. Resection of myoma.



Figure5. Gross specimen of intramural myoma.

Both resected myoma sent for immediate pathological study. A total abdominal hysterectomy with bilateral salpango-oophorectomy was performed by successive clamping. On the omentum it did not show any tumor deposit. No suspected Para

-aortic lymph-node was palpated. The patient hospitalized for a week without any immediate post-operative complication. Ten days after operation consequent results were shown; IHD study confirmed the diagnosis (Fig 6-9).

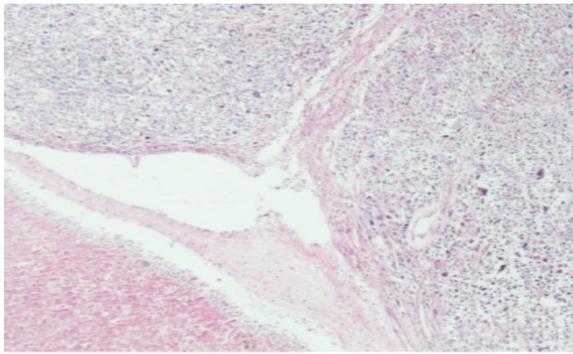


Figure6. LMS showing hypercellularity , pleomorphism and necrosis (X100)

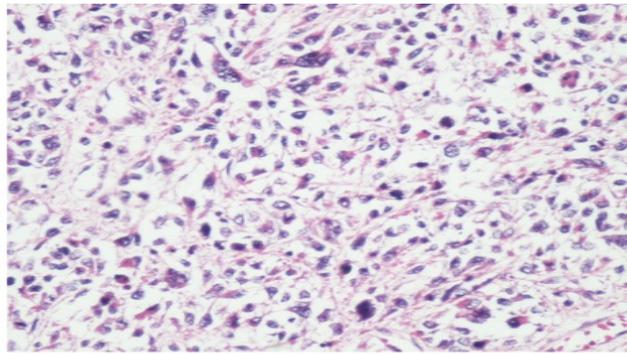


Figure7. Hyper cellularity, pleomorphism

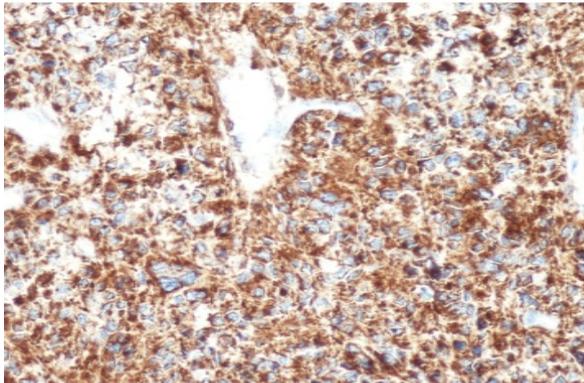


Figure8. SMA immunostaining showing positive staining in tumoral

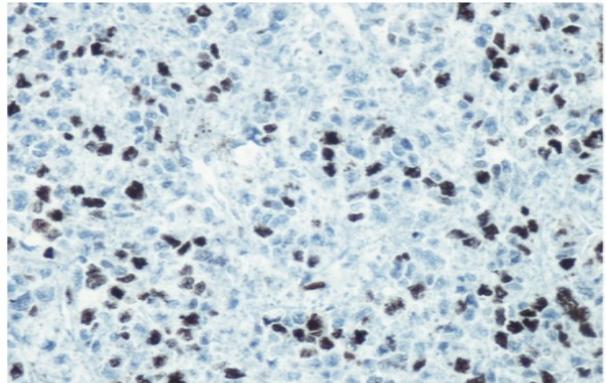


Figure9. Ki-67 immunostaining showing positive nuclear staining in

3. DISCUSSION

Uterine (LMS) compared to other types of uterine cancers is rare uterine malignancy. LMS is an aggressive tumor associated with a high risk of recurrence and death, regardless of a stage at presentation(3, 10). In most cases the diagnosis of uterine (LMS) is made following hysterectomy(11, 12). In our case, hysterectomy by permanent section was diagnosed. In rare cases it is diagnosed with endometrial sampling preoperatively or with frozen section. Frozen section did not suggest in the present case. Prognostic factor include tumor size > 5cm and a high mitotic index. These tumors are highly aggressive, with mitotic count of less than 2 per mm². The most common mode of spread is hemotogenous, and lymphatic spread is rare. Recurrence is reported up to 70% in stage 1 and 2. Commonly, place of recurrence is lungs or upper abdomen (10, 13, 14). Liver, Abdomen, Pelvis and Pelvic or Para- aortic lymph nodes are other sites of metastases (11, 15). Total hysterectomy is done for patient with LMS that is confined in to the uterus at time of surgery (16, 17). Often, bilateral salpingo-oophorectomy (BSO) is performing at the time of total hysterectomy particularly for menopausal or premenopausal women. In women with confined disease to pelvis (stage 2) or the abdomen (stage 3), surgical cytoreduction is also performed. An optimal cytoreduction is associated with improved overall survival (12, 17). In women with metastatic disease extending beyond the peritoneal cavity there is no benefit to surgery (18). Pelvic lymphadenectomy is performed in women with enlarged pelvic nodes and extra uterine disease (15, 19). In the present case palpable pelvic

nodes were not found and omentum was free of tumor. Therefore, pelvic lymphadenectomy and omentectomy were not performed. Adjuvant radiation therapy had no impact on survival outcomes for woman with early stage LMS (20). In stage 3 or distant metastases (stage 4) that have undergone complete resection of disease; there is high risk of disease progression following surgery. Therefore adjuvant chemotherapy is offered by docetaxel and gemcitabine on a GOG trial (21). Due to the high risk of relapse, regardless of stage, for all women with early diagnosed LMS, surveillance examinations and imaging is required. Survival rates are dependent on the stage of disease at diagnosis (10, 22). Five-year survival rate is 50-55% for stage 1 year is 8-12% for stage 2-4 (13).The present case left the hospital after 1 week with no immediate post-operative complication.

4. CONCLUSION

Uterine (LMS) compared to other types of uterine cancers is rare and aggressive uterine malignancy. Total hysterectomy and BSO is recommended for women with confined disease to the uterus pelvic. Lymphadectomy is limited to women with enlarged pelvic nodes. Prognosis for women with uterine (LMS) primarily depends on the extent of disease at the time of diagnosis and the mitotic index. Non randomized studies have reported improved survival after adjuvant chemotherapy. The value of pelvic radiation therapy has not been established.

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AUTHORS CONTRIBUTION

This work was carried out in collaboration between all authors.

CONFLICT OF INTEREST

The authors declared no potential conflicts of interests with

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