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Case Report

Management for a Rare Occurrence of Low Grade Endometrial Stromal Sarcoma (LGESS): A Case Report and Literature Review

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Abstract

According to the American Cancer Society 2017 facts and figures, an estimated 4910 cases of uterine sarcomas are anticipated. According to a population-based analysis from 1988-2003, the overall incidence of ESS among all the uterine malignancies is only 0.2-1%, making it a rather rarer subtype of uterine sarcomas. The median age was 52 years (range: 17-96 years). Surgical treatment was associated with better survival outcomes than other therapy, and especially followed by adjuvant therapy of hormone treatment should be offered, especially in recurrent cases of LGESS. Due to paucity of understanding on the management of this rarer subtype of uterine sarcoma, a case report with review of literature is done which suggests that after surgical debulking, starting patients of maintenance hormonal treatment is the best approach as per current scenario.

Keywords: ***LGESS, Uterine Sarcoma, low grade, endometrial, aromatase inhibitors***

Introduction

According to the American Cancer Society 2017 facts and figures, an estimated 4910 cases of uterine sarcomas are anticipated [1]. A 2012 systemic review of data from 1970-2011, reported uterine leiyomayosarcoma (uLMS) was the most common subtype (63%) followed by endometrial stromal sarcoma (ESS) (21%) of the uterine sarcomas and then other rarer histologies like undifferentiated uterine sarcomas, rhabdomyosarcoma, adenosarcoma, or perivascular epithelioid cell neoplasms (PEComas) [2,3]. According to a population-based analysis from 1988-2003, the overall incidence of ESS among all the uterine malignancies is only 0.2-1%, making it a rather rarer subtype of uterine sarcomas. The median age was 52 years (range: 17-96 years) [4].

The ESS are subdivided based on histopathology and mitotic count in to Low grade ESS (LGESS) and High-grade ESS (HGESS). LGESS have mitotic count <3 per 10 high power field with lack of hemorrhage and necrosis. [5,6] LGESS has a rather rarer occurrence and indolent course with <1% of all uterine malignancies but ranked second among uterine mesenchymal tumors [7,8]. According to the Gynecologic Cancer Intergroup (GCIG) consensus review, early and complete resection is the complete treatment [9]. The five-year disease-specific survival rates for stage I or II disease are 90% and for stage III or IV disease are 50% [10,11]. Surgical treatment was associated with better survival outcomes than other therapy, and especially followed by adjuvant therapy of hormone treatment should be offered, especially in recurrent cases of LGESS [12].

Case Presentation

A 38-year-old female presented with complaints of blood in the urine for the last 1 month. She had no comorbidity, no addiction, and no history of malignancy in the family. She had a history of total abdominal hysterectomy done 3 years ago for complaints of per vaginal bleeding in a local hospital, however, she had no histopathology report or any other document. The patient was evaluated at the time of presentation, her Karnofsky Performance Status (KPS) was 50. Per abdominal examination revealed a palpable suprapubic mass nearly 6 cm in size with restricted mobility. On per vaginal examination, irregular growth of 9 x 8cm involving the vaginal vault, bilateral parametria reaching up to right-side lateral pelvic wall was palpated. USG whole abdomen showed urinary bladder containing an irregular echogenic mass of size 75x 65mm arising from the posterior wall with bilateral gross hydronephrosis. A CT Urogram was done which showed Post hysterectomy status with a large heterogeneous mass lesion 10 x 9.5 x 11cm involving the vaginal vault and grossly infiltrating into the urinary bladder with protrusion into its lumen with encasement of bilateral ureters causing gross hydronephrosis on the right side and mild hydroureteronephrosis on the left side. The lesion was extending into the right pelvic sidewall with a loss of fat planes with the rectum. Few enlarged lymph nodes in the right external iliac chain were noted. A vaginal vault biopsy was done suggestive of a spindle cell tumor. On IHC, tumor was positive for PanCK, Vimentin, Desmin, ER, PR, Bcl-2, CD 99, WT-1 and negative for Cyclin D1, CK7, CK 20, P40, s-100, SMA, MyoD1, HMB45, CD34, Caldesmon, LCA, Synaptophysin, ALK-1, Inhibin. The Ki 67 labeling index was only 1-2% which favored the diagnosis of a low-grade variety of endometrial stromal sarcoma. On further evaluation

for metastasis, CECT thorax showed few heterogeneously enhancing soft tissue lesions scattered in the bilateral lung parenchyma, the largest measuring 1.1x 1cm in the left lower lobe, likely metastatic. The patient then underwent bilateral percutaneous nephrostomy (PCN) followed by palliative radiation to the pelvic mass to a total dose of 20 Gy in 5 frac-tions by 3D- CRT technique. The Patient was symptomatically improved. Further, the patient was given 6 cycles of systemic chemotherapy containing Gemcitabine and docetaxel with palliative intent. Gemcitabine was given in the dose of 1000mg/m2 intravenously on day 1 and day 8, and Docetaxel in the dose of 75mg/m2 intravenously on D1, and the cycle was repeated every 21 days. After 2 cycles of Chemotherapy, Ultrasonography of whole abdomen was done for PCN evaluation which showed resolution of left side mild hydroureteronephrosis, and the PCN tube on the left side was removed. After completion of 6 cycles of chemotherapy, a CECT abdomen and pelvis was done for response assessment which showed a decrease in the size of the lesion involving the vaginal vault to 6.2 x 5.9 x 4.5cm. The tumor anteriorly infiltrates the posterior aspect of the bladder along with the bilateral vesicoureteric junction and right distal ureter. Laterally it infiltrates the right pelvic wall. Multiple well-defined lung nodules were seen in the bilateral visible lung fields with the largest measuring 9 x 8mm and the Right side PCN tube in situ. As per RECIST 1.1 criteria, the patient had stable disease after completion of chemotherapy and KPS improved to 90. In view of ER & PR positivity, the patient was started on oral tablets of Letrozole 2.5 mg, once daily as maintenance therapy. The patient is under follow-up for the last 6 months and is stable. Her general condition has improved significantly with KPS of 80 on last visit to the hospital.

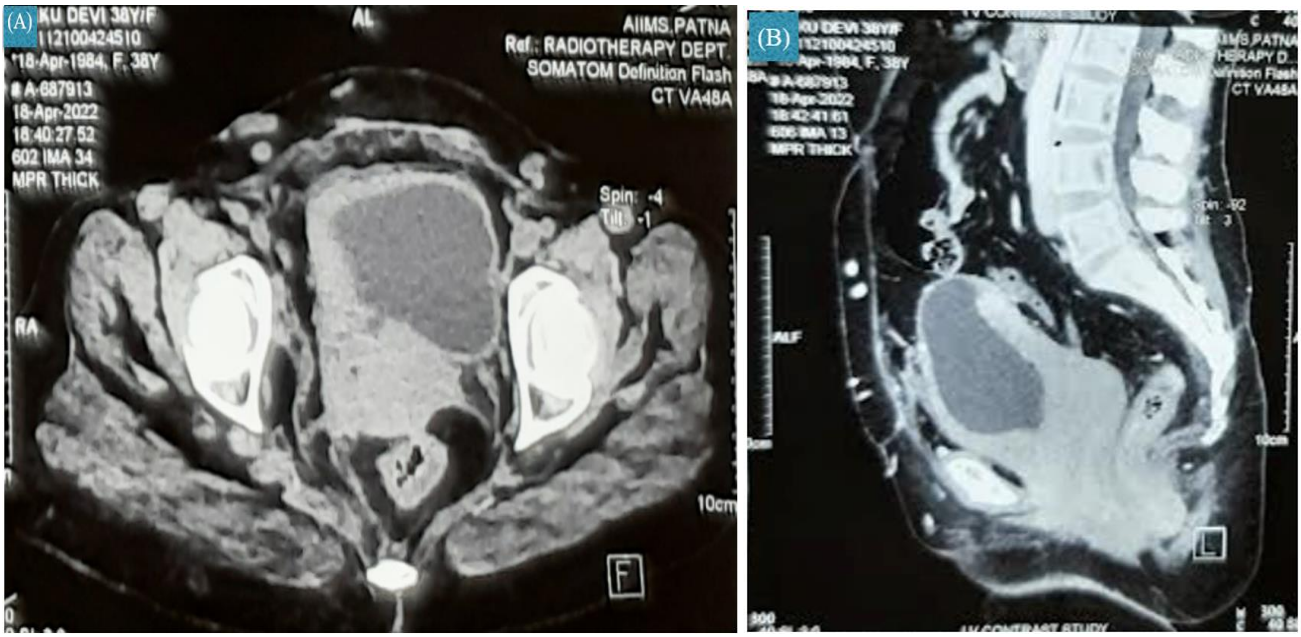

Figure 1: CT-scan images depicting the extent of the disease in, (A) Axial section, and (B) Coronal section

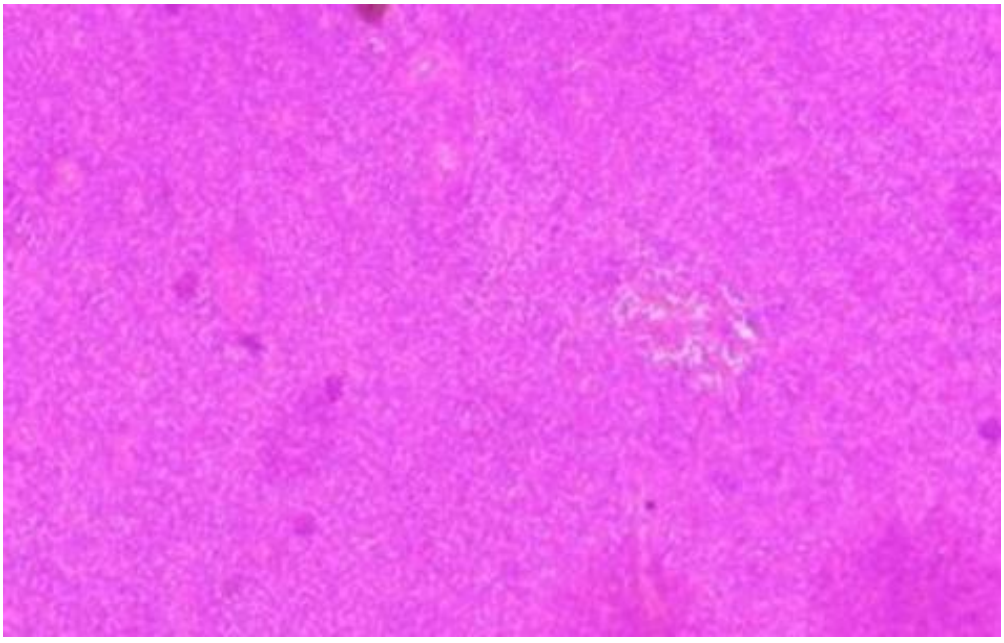

Figure 2: Hematoxylin and Eosin (H & E) Stain (x10)

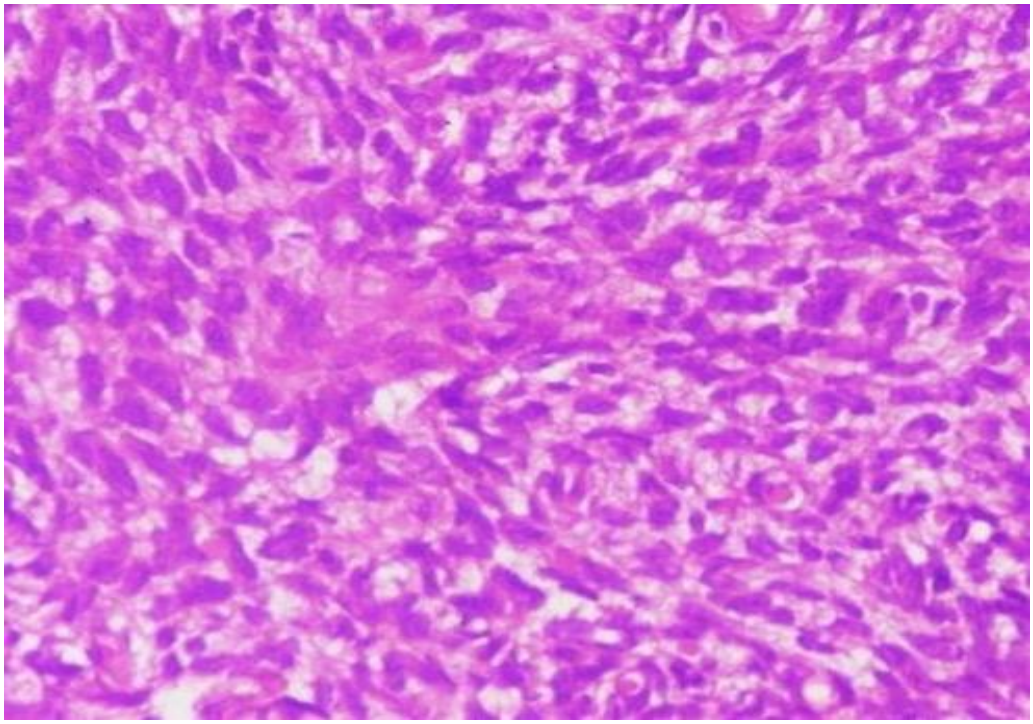

Figure 3: Hematoxylin and Eosin (H & E) Stain (x400)

Discussion

Low grade endometrial stomal sarcoma (LGESS) being a rarer subtype of the uterine sarcomas and indolent nature, not much studies are there delineating definite treatment guidelines for the same. A cohort of 56 patients, which might be the largest population to date study for recurrent LGESS by Dai et. al, suggested that fertility sparing surgeries are not the way forward for the treatment and total abdominal hysterectomy should be done for these cases [12].

SEER analysis of database from 1973 to 2015 survival outcomes of different treatment modalities in patients with low-grade endometrial stromal sarcoma indicated that 35-50 years was the most common age group of diagnosed females, who were primarily treated with total hysterectomy with oophorectomy. The Kaplan-Meier method was used to construct survival and cumulative risk curves, and statistically significant differences between the curves were compared with log-rank tests. Regional lymphadenectomy did not improve 5-year overall survival (OS) and cause specific survival (CSS). Post-operative radiation and chemotherapy had better median survival but p-values were not significant as compared to no radiation and chemotherapy arms [13]. Radiation therapy was offered presumably to patients with larger tumor size and myometrial invasion and is associated with limited benefit of decreasing rates of recurrence but no effect on overall survival [14,15].

A study of 39 patients, from 1984 to 2017, compared aromatase inhibitors (AIs) with progestins as adjuvant hormonal therapy (AHT) for low-grade endometrial stromal sarcomas (LGESSs) for Disease recurrence and recurrence-free survival (RFS). Out of 39 patients, 18 received progestins, 13 received AI, and 8 received no AHT. 38 patients were of stage I disease, and 9 belonged to stage II to IV disease. All patients underwent primarily hysterectomy. Disease recurred in 70% (7/10) of stage I patients in no AHT arm, compared to 14.3% (1/7) receiving AI, and 7.7% (1/13) receiving progestins (P = 0.003) [16].

In our case, we saw a significant improvement in the KPS of patient with stable disease over a period of 6 months of follow-up. The patient had recurrence following the hysterectectomy with large mass, compromising the kidneys and ureters in the form of hydronephrosis, for which PCN was done. As the recurrence was unresectable, she was offered with adjuvant chemotherapy and palliative radiation to the pelvic mass in light of its large size, followed by maintenance endocrine therapy with AI and is being tolerated well.

In conclusion, LGESS is a rarer subtype of uterine sarcoma with very indolent course but higher rates of recurrence. Treatment is limited to total hysterectomy with adjuvant endocrine therapy in the form of aromatase inhibitors. Chemotherapy and Radiotherapy are offered in cases with large tumor burden or invasion with limited benefits.

Ethics approval and consent to participate

Not Applicable

List of abbreviations

LGESS- Low Grade Endometrial Stromal Sarcoma
uLMS- uterine Leiyomayosarcoma
PEComas- Perivascular Epithelioid Cell Neoplasms
KPS- Karnofsky Performance Status
PCN- Percutaneous Nephrostomy
ER- Estrogen Receptor
PR- Progesterone Receptor
AI- Aromatase Inhibitors
AHT- Adjuvant Hormonal Therapy
RFS- Recurrence-Free Survival

Data Availability

Not Applicable

Conflicts of Interest

The author(s) declare(s) that there is no conflict of interest regarding the publication of this paper.

Funding Statement

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Authors' contributions

Dr.Amrita Rakesh took the case history and initiated the treatment after discussion with Dr. Pritanjali Singh. Dr. Amrita Rakesh then followed up the patient for 6 months, keeping her documentation records up-to-date and prepared the manuscript. Dr. Pritanjali Singh reviewed the manuscripts and suggested corrections. Dr. Abhishek Shankar helped finalise the manuscript after keen scrutiny. Dr. Rakesh Ranjan kept tract of all the investigations and histopathological images.

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