e - ISSN - 2349 - 8005

INTERNATIONAL JOURNAL OF ADVANCES IN CASE REPORTS

Journal homepage: www.mcmed.us/journal/ijacr

AGGRESSIVE COURSE IN A WELL MANAGED EARLY STAGE UTERINE LEIOMYOSARCOMA-A CASE REPORT

Ritu Sharma[1*], Ila Tyagi[2], Gaurav Jain[3], Sudha Salhan[4]

[1]Assistant professor, [4]Professor (Obstetrics and gynecology); [2]Associate professor, [3]Senior resident (Pathology), NDMC Medical College, New Delhi, India.

Corresponding Author:- **Ritu Sharma**
E-mail: drritu661@gmail.com

Article Info
Received 15/03/2015
Revised 27/04/2015
Accepted 23/05/2015

Key words:
Leiomyosarcoma, leiomyoma, Degenerative leiomyoma, Post menopausal bleeding, fibroid.

ABSTRACT
Uterine leiomyosarcoma is a rare but highly notorious tumor. Even if diagnosed early the prognosis remains poor. Here we describe the case of a 52 year old postmenopausal female who presented with stage 1B of uterine leiomyosarcoma and was managed aggressively. Despite all, the tumor recurred within 6 months postoperatively resulting in mortality within one year. We present this case because of its rarity and to emphasize the fact that since uterine leiomyosarcomais a highly malignant tumor with poor prognosis, the target of management should be to improve the quality of life by early diagnosis, aggressive treatment and close follow up.

INTRODUCTION

Uterine leiomyoma, arise from smooth muscles of myometrium; and is the most common benign tumor of uterus whereas, uterine leiomyosarcoma, the malignant counterpart is very rare comprising of only 1-2% of uterine malignancies. However, it remains the most common uterine sarcoma subtype as per new FIGO staging representing 25-36% of them [1-3]. Postmenopausal females with uterine leiomyosarcoma mostly present as postmenopausal bleeding and pelvic mass followed by pelvic pain [2]. They are notorious because of associated high risk of recurrence and mortality despite timely diagnosis and management. Here we present a case of a postmenopausal female with stage 1B uterine leiomyosarcoma with postoperative recurrence within 6 months, eventually resulting in mortality within 1 year.

CASE REPORT

This is a case report of a 52 year postmenopausal female, P_4L_4, who reported to gynecology OPD with complaints of abdominal pain for 2 days and postmenopausal bleeding for 2 weeks. The pain was dull and continuous involving lower abdomen without any aggravating or relieving factors. There was history of abdominal discomfort off and on for one month. She attained menopause five years back and started with bleeding per vaginum 2 weeks back. Bleeding was intermittent without any relation with pain abdomen. She was using one to two pads per day. There was no history of vaginal discharge, weight loss, loss of appetite or any abnormal bowel and bladder habits. Before menopause she had normal menstrual cycles. There was nothing significant in her past history except that she had undergone cholecystectomy twenty years back for cholelithisis. Her family history was also not suggestive of any chronic medical disease or malignant disease. All her deliveries were normal with last delivery 24years back. She was average built with normal general physical and systemic examination. On abdominal examination, a large

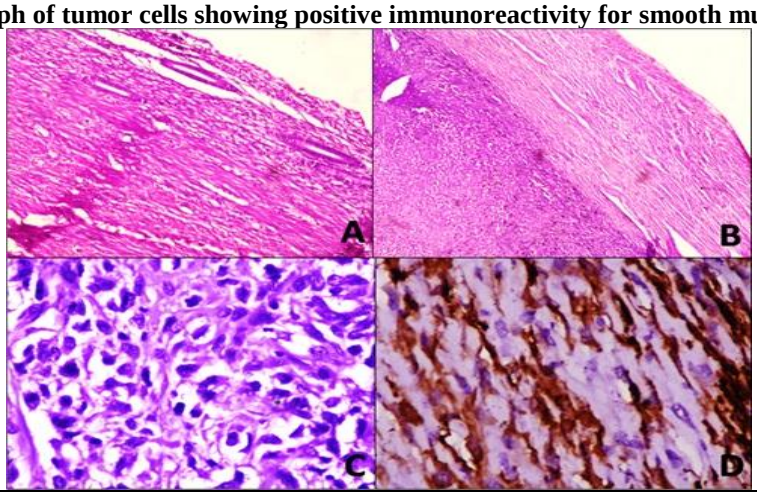
well defined mass of about 16weeks fundal height was palpated which appeared to be arising from pelvis as its lower limit could not be reached. The mass was firm in consistency, non tender with smooth surface, mobile from side to side and dull on percussion. No other mass, organomegaly or ascites was appreciated clinically. On pelvic examination the cervix was mid position and firm. The characteristics of mass observed abdominally were confirmed on pelvic examination and it appeared to be of uterine origin. There was no induration, tenderness or any other mass palpable through any of the fornices. Rectal mucosa was found to be free on per rectal examination. Patient was admitted and her baseline investigations were found to be within normal limits. Ultrasonography done one month back outside our hospital showed bulky uterus with 5cm×6cm fundoposterior fibroid. Repeat ultrasonography done in our hospital showed a bulky uterus with an echogenic area of 8cm×5cm in the fundal region and thickened endometrium with ill defined endomyometrial junction suggestive of either uterine fundoposterior intramural fibroid with degenerative changes or malignant uterine tumor. Rest of abdominal scan was normal without any evidence of metastasis or ascites. Magnetic resonance imaging study was in favor of malignant uterine tumor without any adenexal involvement or metastasis. Her PAP's smear report was inconclusive. The endometrial biopsy showed cells with features suggestive of malignancy most likely high grade stromal lesion. As per new FIGO staging, patient was kept in stage IB of leiomyosarcoma and the treatment was planned accordingly. She was prepared for surgery after taking written informed consent and arranging adequate amount of blood and components within three weeks of her admission. During her hospital stay the mass was found to be rapidly growing from 16 weeks to 20 weeks size. Considering her age, clinical and radiological findings, radical hysterectomy was done involving removal of uterus, fallopian tubes, ovaries and upper one third of vagina along with para aortic and pelvic lymph node sampling. The intraoperative findings included enlarged uterus with posterior wall intramural fibroid of about 8cm×6cm size with overlying serosa being grossly intact. Both ovaries were atrophic. There were no intra abdominal adhesions, palpable lymph nodes, abnormal nodules or ascites. On cut section the endometrium was hypertrophied and unhealthy looking, the myometrium was also hypertrophied and fibroid showed areas of necrosis and calcifications (Fig 1). All samples were sent for histopathological examination. She received one blood transfusion during surgery. Her postoperative period was uneventful. She was discharged on 10^{th}potoperative day after stitch removal. The histopathological report showed leiomyosarcoma of the uterus with tumor cells showing positive immunoreactivity for smooth muscle actin (Fig 2), however the lymph nodes were not involved. The patient was referred to higher centre for adjuvant therapy where she received radiotherapy and chemotherapy. Postoperatively within six months she developed local recurrence as well as distant metastasis to the liver and expired within 1 year.

DISCUSSION

Irrespective of the stage of presentation and time of initiation of treatment uterine leiomyosarcoma has a fulminant course. Over and above that, no definite risk factors have been identified till now although African-American race, age >40years, pelvic irradiation, retinoblastoma gene, exposure to certain chemicals (vinyl chloride, dioxins, herbicidal) and obesity have been found to be associated with it [4]. Again, it is very difficult to differentiate between leiomyoma and leiomyosarcoma clinically as well as radiologically. However, certain clinical signs like rapid growth of fibroid in postmenopausal female are highly suspicious of leiomyosarcoma which is known to have a doubling time of as less as 4 weeks [5]. Also, no screening method is available for this rare tumor. Leiomyosarcoma diagnosis is usually made by high index of suspicion aided with various investigations like endometrial biopsy, radiological investigations (ultrasonography, CT scan, Magnetic resonance imaging, PET scan, Chest X ray) and hysteroscopy. Since it is limited to myometrium, endometrial biopsy mostly does not yield positive result in early stage tumor. In one series of women with uterine leiomyosarcoma, only 35 percent were diagnosed preoperatively [6]. The diagnosis is mostly made retrospectively post operatively. Ours is one of the few cases where diagnosis was made preoperatively by endometrial biopsy. Probability of pre operative diagnosis may be increased using gadolinium enhanced MRI and serum LDH [7]. No doubt in 60% of cases, leiomyosarcoma is limited to uterus at the time of presentation; but the matter of concern is that even these patients despite adequate treatment have poor prognosis due to high rate of local and distant recurrences ranging from 45-80% [8, 9]. The most common sites of metastasis include lungs, liver followed by bowel, heart, brain and thyroid [1, 10]. Patients with recurrent disease after initial resection have a median survival of less than 1year [11]. Long term survival rates vary from 53% for stage 1 [12, 13] to 8% for stage 2-4 [12]. Prognostic factors include size of tumor, mitosis, stage of disease, grading and aneuploidy; the first two being the major prognostic factors according to some studies [4]. Treatment includes total abdominal hysterectomy with bilateral salpingo oophrectomy with lymph node sampling with or without adjuvant therapy for early stage tumor. In late stages debulking along with radiotherapy and chemotherapy is required. For local recurrence radiotherapy is said to be effective while chemotherapy has shown efficacy in distant and advanced disease. Since leiomyosarcoma behaves like a silent volcano which erupts abruptly and aggressively, there should be every possible attempt for its early diagnosis and management.

Figure 1. Gross image showing large intramural mass with cut surface showing variegated appearance and areas of necrosis.

Figure 2(a-d). Figure a – photomicrograph showing endometrium with underlying tumour consisting of pleomorphic spindle cells. (H&E X100); Figure b – photomicrograph of tumour with overlying uterine serosa. (H&E X100); Figure c – photomicrograph showing bizarre pleomorphic tumor cells with multiple atypical mitotic figures. (H&E X400); Figure d – photomicrograph of tumor cells showing positive immunoreactivity for smooth muscle actin.



CONCLUSION

In a postmenopausal female with sudden increase in size of fibroid, a diagnosis of leiomyosarcoma should be considered until proved otherwise. Considering the fact that it is a highly malignant tumor with poor prognosis, the target of management should be to improve the quality of life by early diagnosis, aggressive treatment and close follow up.

DECLARATIONS

All the authors have contributed significantly in this manuscript. There is no conflict of interest among the authors.

REFERENCES

1. Echt G, Jepson J, Steel J, Langholz B, Luxton G, et al. (1990). Treatment of uterine sarcomas. *Cancer*, 66, 35-39.
2. Norris HJ ZC. (1993). Mesechymal tumors of the uterus. 4th edn. Springer-Verlag, New York, USA.
3. D'Angelo E, Prat J. (2010). Uterine sarcomas: a review. *Gynecol Oncol*, 116,131.
4. Giuntoli II RL, Metzinger DS, DiMarco CS, Cha SS, Sloan JA, Keeney GL, et al. (2003). Retrospective review of 208 patients with leiomyosarcoma of the uterus: prognostic indicators, surgical management, and adjuvant therapy. *Gynecol Oncol*, 89, 460–9.
5. Venkata Sujatha Vellanki, Meghana Rao, Chinna Babu Sunkavalli, Rao N Chinamotu, Shailaja Kaja.(2010). A rare case of uterine leiomyosarcoma: a case report. *Journal of Medical Case Reports*, 4, 222.

6. Sagae S, Yamashita K, Ishioka S, et al. (2004). Preoperative diagnosis and treatment results in 106 patients with uterine sarcoma in Hokkaido, Japan. *Oncology*, 67, 33.
7. Goto A, Takeuchi S, Sugimura K, Maruo T. (2002). Usefulness of Gd-DTPA contrast-enhanced dynamic MRI and serum determination of LDH and its isozymes in the differential diagnosis of leiomyosarcoma from degenerated leiomyoma of the uterus. *Int J Gynecol Cancer*, 12, 354–61.
8. Major FJ, Blessing JA, Silverberg SG, et al. (1993). Prognostic factors in early-stage uterine sarcoma. A Gynecologic Oncology Group study. *Cancer*, 71(suppl 4), 1702–1709.
9. Dinh TA, Oliva EA, Fuller AF Jr., et al. (2004). The treatment of uterine leiomyosarcoma: Results from a 10-year experience (1990-1999) at the Massachusetts General Hospital. *Gynecol Oncol*, 92, 648–652.
10. Rose PG, Piver MS, Tsukada Y, Lau T. (1989). Patterns of metastasis in uterine sarcoma. *Cancer*, 63, 935-938.
11. Hensley ML, Maki R, Venkatraman E, et al. (2002). Gemcitabine and docetaxel in patients with unresectable leiomyosarcoma: Results of a phase II trial. *J Clin Oncol*, 20, 2824–2831.
12. Salazar, O.M., Bonfiglio, T.A., Patten, S.F., Keller, B.E., Feldstein, M., Dunne, M.E., and Rudolph, J. (1978). Uterine sarcomas: natural history, treatment and prognosis. *Cancer*, 42, 1152-1160.
13. Abeler VM, Royne O, Thoresen S, Danielsen HE, Nesland JM, Kristensen GB. (2009). Uterine sarcomas in Norway.A histopathological and prognostic survey of a total population from 1970 to 2000 including 419 patients. *Histopathology*, 54, 355–64.