

Primary ovarian leiomyosarcoma: a case report with review

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ABSTRACT

Primary ovarian leiomyosarcoma is a very rare tumor. Presenting a case report with review of literature.

Keywords: Primary ovarian leiomyosarcoma, Leiomyosarcoma, Primary ovarian

INTRODUCTION

Primary ovarian leiomyosarcoma is very rare ovarian tumor, with less than 50 cases in the literature.¹ Because of its extreme rarity; a standard treatment isn't yet established. We present this case of ovarian neoplasm in a postmenopausal woman. Literature of primary leiomyosarcoma of the ovary is reviewed.

CASE REPORT

A 59 year old female, P₂L₂A₁ came with complains of post-menopausal bleeding since 1 week, it was mild in amount; there was no history of contact bleeding. No complains of pain abdomen or any urinary complains.

On per abdomen examination, abdomen was soft, no guarding, rigidity or tenderness was present. On per speculum examination cervix appeared to be healthy, bleeding was seen through the os. On per vaginal examination multiparous anteverted uterus palpable, a 3 by 4cm mass was palpable in right fornix, left fornix was free and non-tender.

Routine investigations were normal. CA-125 was 5.5U/ml. On ultrasonography, anteverted uterus with thick ET of 13mm was noted; a 4 by 4 cm solid right adnexal mass was seen, likely to be of ovarian origin, normal left ovary seen. On MRI pelvis solid right ovarian mass likely neoplastic, showing internal cystic changes.

Thick ET with poor definition of cervico vaginal junction, suggestive of endometrial hyperplasia/endometrial carcinoma. No obvious ascites or deposits on serosal surface of pelvis, no enlarged lymph nodes noted. On Fractional Curettage findings were suggestive of simple endometrial hyperplasia along with chronic cervicitis. Ultrasound guided FNAC of the mass showed blood component with occasional cluster of epithelial cells having mild atypia, aspirate was inadequate for a definitive opinion.

On laparotomy, TAH with BSO was done. On imprint cytology of right ovary, it was found to be benign ovarian tumor. On histopathology? Cellular fibroma? fibrosarcoma. On immunohistochemistry, intermediate grade leiomyosarcoma was diagnosed. Microscopic examination of left ovary revealed normal ovarian stroma. No leiomyoma were detected in sections taken from uterus and tubes.

DISCUSSION

Primary ovarian sarcoma are very rare tumors, comprising less than 2% of all ovarian malignancies.¹ They are a heterogeneous group of tumors, consisting of the fibrosarcomas, endometrial stromal sarcomas, rhabdomyosarcoma^{3,4} and the rare leiomyosarcomas, (less than 0.1%).¹

Primary ovarian leiomyosarcomas are usually seen in postmenopausal women, though few cases seen in younger women have been reported.^{1,3} Pathogenesis of ovarian leiomyosarcomas is not clear, though various theories proposed, two of the prominent ones being, malignant degeneration of smooth muscle tissue present in the ovary and malignant transformation of uterine leiomyoma to the ovary.⁴ Leiomyomas are usually seen in superficial tissue, whereas leiomyosarcomas are deep seated tumors,⁶ thus negating the theory of leiomyomas to be the precursor of leiomyosarcoma.

According to the source of origin, ovarian leiomyosarcomas can be divided into three types, teratoid, mesenchymal and müllerian.⁷ Leiomyosarcomas of teratoid origin are usually unilateral and observed in young patients whereas those of mesenchymal origin are seen more commonly in the elderly age group and more likely to metastasize.

Leiomyosarcomas usually present late with vague non-specific symptoms comprising of vaginal bleeding, pelvic pain or pressure, urinary or bowel complaints and awareness of abdominal pelvic mass which on examination appears as a unilateral, solitary, soft and fleshy mass.

On pathological examination, grossly they are indistinguishable from other sarcomas and appear as solid masses with or without cystic degeneration, depending on size of tumor.² Microscopically, uniform spindle shaped cells with abundant eosinophilic cytoplasm and elongated nuclei are seen. They are arranged in whorled pattern with areas of pleomorphism and necrosis.³ They differ from the leiomyomas in being hypercellular and having increased mitotic activity (10 or more mitoses per 10 hpf).³ Coagulative necrosis, cellular atypia and mitotic index greater than 10 are taken to be evidence of malignancy. Immunohistochemical and electron microscopic evaluations may improve diagnostic accuracy.

Due to rarity of the disease and late presentation, a standard optimal treatment does not exist. Different treatment modules have been described in literature. Post-operative radiotherapy and chemotherapy have been recommended in large tumours to prevent local and metastatic spread of the disease.² In a study by Rasmussen et al.³ treatment with repeated cyto-reductive surgery, hormonal treatment and adjuvant ifosfamide has shown to have better prognosis than surgery alone. Pre-operative chemotherapy has also been tried in large tumors to increase chances of operative success. Still there is no conclusive evidence that adjuvant chemotherapy or radiotherapy offers prognostic benefit and surgery has remained the mainstay of treatment.² Ploration.⁹

Ovarian leiomyosarcomas have poor prognosis and patients usually die due to their late presentation and aggressive nature. Patients usually die within one

year,⁴ because of late presentation and aggressive nature of the tumor leading to widespread metastasis and pressure symptoms from the large ovarian tumor.

In our patient radical surgery wasn't done because, imprint cytology suggested a benign ovarian tumor and omentectomy/ omental biopsy / lymphadenectomy was also not done. Post operatively under supervision of oncologist patient is on repeated CT every 6 months. Patient is asymptomatic and healthy. Probably surgical intervention at an early stage has helped in improving her prognosis. Moreover, considering the age of the patient and rarity of a benign tumor at this age, in spite of imprint cytology, she could have been given the benefit of doubt and omental biopsy / lymph node biopsy / peritoneal washings could have been taken.

CONCLUSIONS

Ovarian leiomyosarcomas are very rare ovarian tumors, whose origin is unclear. They are very aggressive tumors with poor prognosis. Optimal debulking surgery has prognostic value in the course of disease. There is need for further research in efficacy of adjuvant chemotherapy, radiotherapy.

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