Primary leiomyosarcoma of the broad ligament: A case report with review of literature

Devika Gupta a,⁎, Giriraj Singh b,1, Pooja Gupta b,2, Satish Mendonca c,3, Kamlesh Kumar Singh a,4

a Armed Forces Medical College, Department Of Pathology and Laboratory Science, Command Hospital, Pune, 411040, India
b Armed Forces Medical College, Department of Radiodiagnosis and Imaging, Command Hospital, Pune, 411040, India
c Armed Forces Medical College, Department Of Medicine, Command Hospital, Pune, 411040, India

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Abstract Primary leiomyosarcoma of the broad ligament is a very rare and highly malignant tumour with very few cases being reported in literature. They have an aggressive course and occur usually in postmenopausal women. We herein report a rare case of primary leiomyosarcoma of the broad ligament which was diagnosed based on imaging, the classical histomorphological and immunohistochemistry features and subsequently treated by complete surgical resection. Primary leiomyosarcoma of the broad ligament is a rapidly progressive and highly malignant gynaecological tumour. It requires complete surgery (hysterectomy with bilateral salpingoophorectomy) for all cases. Adjuvant chemotherapy or radiotherapy is used in selected cases. Low to intermediate grade tumours with no evidence of metastasis are treated surgically only, with advice for a close follow-up.

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★ Authors’ Contributions:
[Devika Gupta]
Group 1 — Conception and design, acquisition of data, analysis and interpretation of data
Group 2 — Drafting the article, critical revision of the article
Group 3 — Final approval of the version to be published

[Giriraj Singh]
Group 1 — Acquisition of data, analysis and interpretation of data
Group 2 — Critical revision of the article
Group 3 — Final approval of the version to be published

[Pooja Gupta]
Group 1 — Acquisition of data, analysis and interpretation of data
Group 2 — Drafting the article, critical revision of the article
Group 3 — Final approval of the version to be published

[Satish Mendonca]
Group 1 — Conception and design, acquisition of data
Group 2 — Critical revision of the article
Group 3 — Final approval of the version to be published

[Kamlesh Kumar Singh]
Group 1 — Conception and design, acquisition of data
Group 2 — Critical revision of the article
Group 3 — Final approval of the version to be published

⁎ Corresponding author at: Armed Forces Medical College, Department Of Pathology and Laboratory Science, Command Hospital, Pune, 411040, India. Tel.: +91 58984335.
E-mail addresses: devikalives5h@gmail.com (D. Gupta), girigujral@gmail.com (G. Singh), pooja1306@gmail.com (P. Gupta), satishmendonca@yahoo.co.in (S. Mendonca), kamulyses@gmail.com (K.K. Singh).

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1. Introduction

Primary leiomyosarcomas of the broad ligament are very rare, aggressive tumours with about only 20 case reports in literature [1]. The definition of leiomyosarcoma of broad ligament as proposed by Gardner et al. is that “these tumors occur on or in the broad ligament, but are completely separated from and in no way connected with either the uterus or the ovary” [2]. The histological criteria for the diagnosis of leiomyosarcoma are high mitotic index, cytological atypia and areas of necrosis [3]. We hereby report a rare case of this highly malignant tumour in a premenopausal female.

2. Case report

A 41 year old female Para 2 presented to our tertiary care hospital with symptoms of dull pain and swelling in lower abdomen of eight month's duration. There was no significant gynaecological or past medical history. There were no complaints of menorrhagia, metrorrhagia or dysmenorrhoea. General physical examination revealed a vague ill-defined, nontender lump palpable in the left side of lower abdomen. All her haematological and biochemical parameters were within normal limits. An ultrasound examination of the abdomen and pelvis showed a heterogenous space occupying lesion of size 10.3 × 8.6 cm in the left adnexa with increased vascularity of probable neoplastic origin (Fig. 1). The uterus was normoechogenic with normal thickness of endometrium. Computed tomography (CT) scan of the abdomen and pelvis showed a well defined, rounded heterogenous space occupying lesion arising from left adnexa. It measured 10.4 × 9.6 cm. On post contrast study, there was moderate heterogenous enhancement with nonenhancing necrotic component within (Fig. 2). It was causing mass effect in the form of displacement of uterus to the right side and also anteriorly. It was also causing an indentation over posterior surface of urinary bladder. Fat planes were maintained between the mass, uterus and ovary. The serum CA-125, Alpha Fetoprotein and Beta HCG levels were within normal limits. After a complete work up, patient was taken up for surgery and she underwent total abdominal hysterectomy with bilateral salpingoophorectomy.

Gross examination of the specimen revealed a large, encapsulated greyish white firm mass arising from left broad ligament. The mass was separate from the uterus and left ovary. Cut surface was solid, grey brown with areas of necrosis and haemorrhage. Cut surface of uterus and both ovaries was unremarkable. Microscopic examination of the mass showed a hypercellular tumour composed of malignant spindle cells arranged in intersecting fascicles and focally forming a herringbone pattern. The tumour cells had pleomorphic, vesicular coarse chromatin with conspicuous nucleoli. Mitosis was brisk at about 12–14/10 HPFs. There were interspersed areas of necrosis (Fig. 3a & b). Sections from both ovaries and uterus were unremarkable. Immunohistochemistry (IHC) studies showed the tumour cells to be positive for Alpha smooth muscle actin (Fig. 3c), Desmin and vimentin. The cells were negative for PanCK, CD117, S-100 and CD68.

Based on these morphological and IHC analyses a diagnosis of primary leiomyosarcoma of left Broad Ligament—Grade II (intermediate) as per FNCLCC (French Federation of Cancer Centre Sarcoma Group) grading system was made.

Post operative recovery was uneventful and a repeat abdomino-pelvic CT scan did not show any residual tumour. Because of an intermediate grade leiomyosarcoma with no residual disease, the patient was advised against adjuvant chemotherapy. The patient is presently...
3. Discussion

Primary leiomyosarcoma originating in the broad ligament is a rare neoplasm. The broad ligament is a double layered peritoneal fold that encloses the parametrium. The ligament extends from the sides of the uterus to the pelvic side walls and the pelvic floor. Broad ligament tumours are generally asymptomatic. If they are large they may be palpable and cause symptoms related to compression of pelvic organs. On applying the stringent criteria laid down in 1977 by Gardner et al. for the primary sarcomas of broad ligament, the tumours arising primarily from broad ligaments became extremely rare. These tumours occur commonly in the postmenopausal women and only three cases in younger age group has been reported [4,5].

Leiomyosarcoma is the most common solid broad ligament malignant mesenchymal tumour. Amongst the reported cases of primary malignant tumour of broad ligament are leiomyosarcoma, GIST, Endometrial Stromal Sarcoma, Malignant Fibrous Histiocytoma and Carcinosarcoma [6,7].

The clinical manifestations of the cases are non specific signs and symptoms which include abdominal pain, distension, nausea, constipation and malaise. Occasionally there were cases with progressive symptoms of acute retention of urine. However no case has been diagnosed before surgery and the final diagnosis is based on microscopic examination, supported by IHC studies [8,9].

As there are very few case reports of primary leiomyosarcoma the histological diagnostic criteria, the staging and management used are the same as for uterine leiomyosarcomas [10].

The microscopic diagnosis of leiomyosarcoma has evolved gradually over the years. Earlier diagnosis of sarcoma was related to mitotic count. Over the years the diagnosis of leiomyosarcoma relies on the presence of three criteria: coagulative tumour cell necrosis, cytologic atypia and mitotic activity. Zaloudek and Hendrickson [11] proposed that the presence of coagulative necrosis itself is enough for diagnosis of leiomyosarcoma. The Stanford study [12] was the first to appreciate that necrosis in a uterine smooth muscle tumour was of crucial importance. In the absence of cell necrosis the diagnosis of leiomyosarcoma needs diffuse moderate to severe cellular atypia and more than 10 mitoses/10 HPFs [13]. Authors have refined and redefined the diagnostic criteria that have been used for years and have made attempts to correlate these findings with clinical outcome.

The systems used for grading of soft tissue sarcomas are the NCI (National Cancer Institute) and FNCLCC systems. The NCI system uses a combination of histological type, cellularity, pleomorphism and mitotic rate for attributing grade 1 or 3. We used the FNCLCC grading system which is based on a score obtained by evaluating three parameters: tumour differentiation, mitotic rate (0–9, 10–19 & > 20 mitoses/10 HPFs) and the amount of tumour necrosis (<50% tumour necrosis and >50% tumour necrosis). According to both these systems, leiomyosarcoma is classified as low, intermediate and high grade.

There is wide variation in the management practices of this uncommon tumour but initial treatment is same as that of uterine leiomyosarcoma, i.e., total abdominal hysterectomy and bilateral salpingoophorectomy [14]. Pelvic lymph node dissection is debatable. The imaging and microscopic pattern play a crucial role in defining overall prognosis and need for
adjuvant therapy. Pelvic irradiation therapy has been used for adjuvant treatment of uterine leiomyosarcomas as radiation therapy and has been shown to decrease the pelvic locoregional relapse rate. Studies have not demonstrated a significant survival benefit.

Our patient had an intermediate grade tumour and did not receive any adjuvant therapy.

She has had no evidence of recurrence or metastasis after one year of follow-up in Gynae-oncology OPD.

4. Conclusion

This case highlights the fact that high index of suspicion, early diagnosis and prompt surgical intervention can prevent distant metastasis and increase the survival rate.

References