Broad ligament leiomyosarcoma a diagnostic challenge: Case report and review of literature

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Abstract- Broad ligament leiomyosarcoma is a rare and aggressive soft tissue sarcoma. It is more commonly seen in postmenopausal women. Diagnosis can be difficult preoperatively and often it is confused with adnexal tumors. We herein report a case which was diagnosed as ovarian carcinoma by imaging studies but intraoperative findings and histopathology concluded it to be a broad ligament leiomyosarcoma.

Index Terms- Broad ligament leiomyosarcoma, sarcoma.

Conflict of Interest:
No conflict of interest is involved in this case report for each of the authors

I. INTRODUCTION

Leiomyosarcomas constitute approximately 5-10 % of all soft tissue sarcomas. They are subdivided into 3 groups for therapeutic purpose - somatic soft tissue leiomyosarcoma, cutaneous leiomyosarcoma and leiomyosarcoma of vascular origin. Broad ligament leiomyosarcoma is even more rare sarcoma. [1] As of now only 24 cases of broad ligament leiomyosarcoma have been reported in literature.(Table 1). We report a similar case of huge broad ligament leiomyosarcoma weighing 8.5 kg which was misdiagnosed as ovarian carcinoma and was successfully removed surgically .

Case :

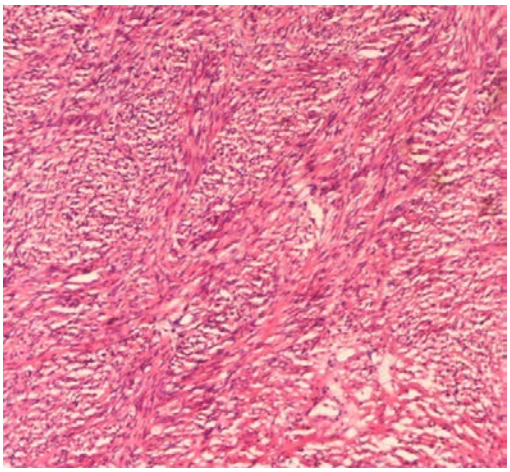
58 years postmenopausal women with average built was admitted for abdominal lump in gynaec ward following total abdominal hysterectomy 15 years back at some private hospital for abnormal uterine bleeding. There was gradual increase in lump size over a span of 10 years. There was no other positive finding in history except for loss of appetite and weight. However the weight loss was not very significant. Ultrasound was suggestive of B/L ovarian tumor with variable echogenecity and multiple cystic lesions. CT scan done showed 24x20x31.6cm right adnexal mass and 12.3x11.8x12.8cm left adnexal mass. Left ureter was seen to be encased between the two masses. CA125 (18 u/ml) CEA 1.2 ng/ml and CA 19-9, 28.5 U/ml done were within normal limits. Total count was also normal however for low Haemoglobin (Hb -6.5gm/dl) 2 units of packed cells were transfused and patient was started on high protein and iron rich diet. After complete workup patient was taken up for surgery. Intraoperatively both tumors were found to be retroperitoneal (Figure 1). They were removed with capsule intact. Lymphadenectomy (paraaortic and iliac group lymph nodes) done and all the tissue material sent for histopathology. Postoperative period was uneventful. Two more units of packed cells were transfused. Histopathology reported it to be malignant smooth muscle tumor - broad ligament leiomyosarcoma. Pathological examination showed -large sheets, nodules which were partially encapsulated and cells were mainly epitheliod, spindled, ovoid forming interlacing fascicles and whorls with 5 mitotic figures/10 high power fields. Extensive myxoid changes, microcystic areas giving reticular appearance and coagulative necrosis were also seen. Immuohistochemistry showed (Figure2 , 3)– smooth muscle actin and desmin diffusely positive, CD34 negative in tumor area. Patient was discharged in stable condition on post operative day nine with advise for follow up.

Figure 1 : Intraoperative photograph of the right tumor which weighed 8.5kg

Figure 2 : Microscopic examination :

A. Showing a cellular tumor arranged in interlacing bundles of spindle cells

B. Elongated hyperchromatic nuclei and nuclear pleomorphism



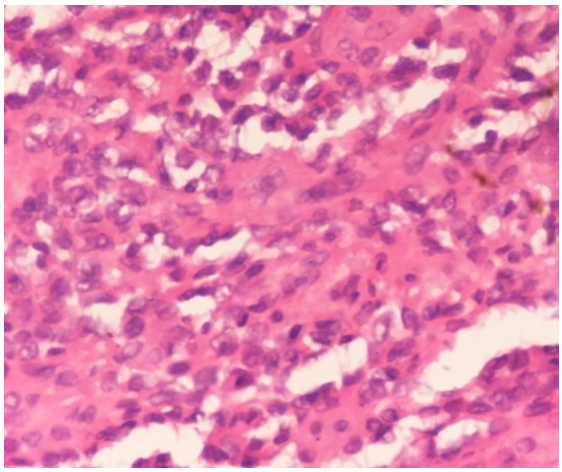
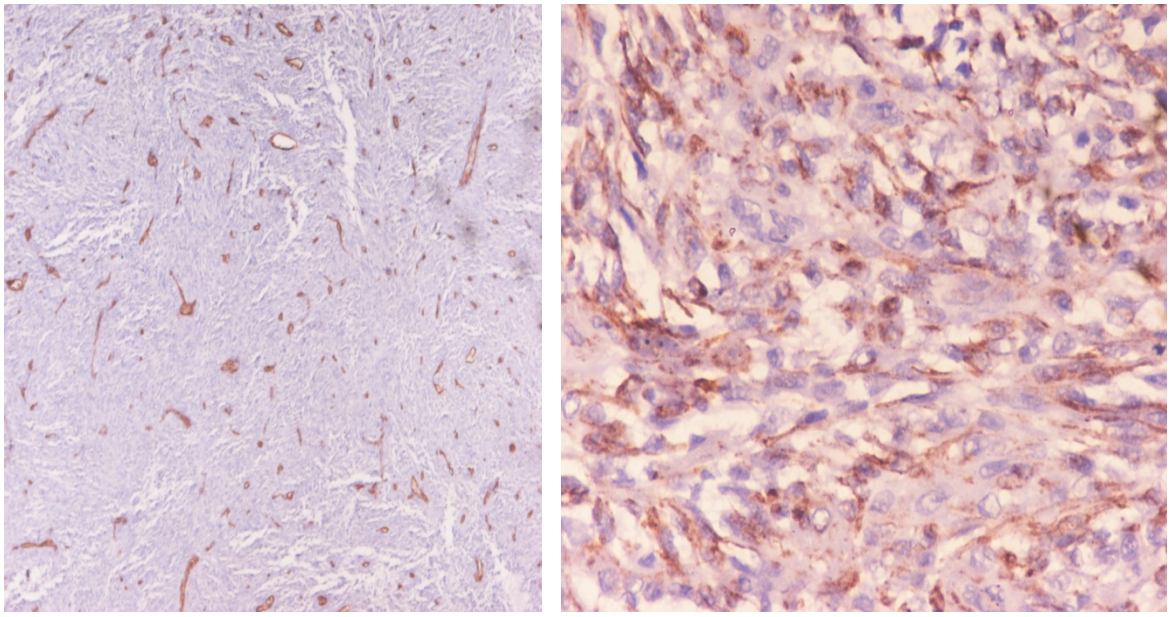


Figure 3 : Immunohistochemistry :

A. Negative staining for CD117

B. Positive staining for smooth muscle actin

C.



II. Discussion

Broad ligament tumors are quite rare; leiomyoma is the most common amongst it with incidence<1 %.[2] Broad ligament leiomyosarcoma is a extremely rare tumor tumor which arises in /on the broad ligament and is in no way connected with the uterus or the ovary as per Gardner et al definition. [3] It is challenging to both clinician and radiologist to diagnose it preoperatively.

Leiomyosarcoma and leiomyoma can be differentiated microscopically by the following three features:[2]

- Five or more mitotic figures per 10 high-power field (HPF)
- Nuclear atypia
- Hypercellularity

Other rare tumors of broad ligament are : Ewing sarcoma family of tumors (ESFT), steroid cell tumor, papillary cystadenoma of the broad ligament, oncocytic adrenocortical adenoma arising from adrenal rest in the broad ligament. [2]

The NCI (United States National Cancer Institute) system and the FNCLCC (French Fédératio Nationale des Centres de Lutte Contre le Cancer) system are the two most important grading systems for soft tissue sarcomas. The NCI system uses a combination of histological type, cellularity, pleomorphism and mitotic rate for attributing grade 1 or 3. The FNCLCC system is based on a score obtained by evaluating three parameters: tumor differentiation, mitotic rate (0-9, 10-19 and ≥ 20 mitotic figures/10HPF) and the amount of tumor necrosis (<50% tumor necrosis and ≥ 50% tumor necrosis). According to both of them leiomyosarcoma is classified into low, intermediate and high grade.[4,5]Due to anatomical close proximity, broad ligament tumors may be confused clinically as well as radiologically with adnexal neoplasms. The differential diagnosis for broad ligament leiomyosarcoma includes masses from ovarian origin – benign or malignant, broad ligament cyst, lymphadenopathy and tubo-ovarian masses. Likewise in our case imaging studies showed it to be a ovarian carcinoma. Lee et al. proposed the "ovarian vascular pedicle" sign as a way of differentiating ovarian neoplasms by single-detector helical CT . When the ovarian vascular pedicle sign on helical CT confirmed the ovarian origin, the sensitivity, specificity, positive predictive value, negative predictive value, and diagnostic accuracy were 92% (99/108), 87% (20/23), 97% (99/102), 69% (20/29), and 91% (119/131), respectively. However origin of the tumor cannot be diagnosed correctly when the tumor is huge as cystic degeneration resembles it to ovarian cancer and it is impossible to detect feeding vessels with MRI and CT as in our case. [6]

III. Conclusion

Primary broad ligament leiomyosarcoma is difficult to diagnose clinically and with radiology studies, histopathology plays an important role for making the diagnosis.

Table 1 : Review of literature of Broad ligament leiomyosarcoma cases :

No	Author	year	Ref No	Age Years	Diameter	Preop diag	Mitoses	Surgery	Adjuvant therapy	Status
1	Devika Gupta et al	2015	3	41	10.3x8.6cm	Adnexal tumor	12-14/10HPF	TAH+BSO	CT	AWD 2 months
2	Akhavan A	2013	4	60	10cm	?	10/10HPF	TAH +BSO	RT	Metastases to abdominal wall 5years later
3	Kolusari et al.	2009	6	35	18	pelvic mass	>20/10HPF	TAH+BSO+OM+PLN+PAND	CT+RT	NED>12 month
4	Duhan et al.	2009	6	45	24	NA	>10/10HPF	Resection+BSO	CT	NED>15 months
5	Papachatzopoulos et al.	2009	6	38	20	fibroid	>10/10HPF	TAH+BSO	-	DOD 8 months
6		2008	7	45	NA	Broad ligament leiomyosarcoma	NA	Resection	CT	NED >15 months
7	Falconi et al.	2006	6	52	NA	NA	NA	TAH+BSO	NA	AWD 117 months
8	RMurialdo etal	2005	5	53	?	Broad ligament leiomyosarcoma	<10/10HPF	TAH+BSO	None	NEM 13 months
9	Ben Amara et al.	2005	6	49	23	ovarian cancer	7/10HPF	TAH+BSO+OM	-	DOD 5 months
10	El-Idrissi & Fadli	2004	6	52	12.5	NA	NA	TAH+BSO	-	DOD 3 months
11	Kir et.	2003	6	35	17	NA	15-20/10HPF	TAH+BSO+PLN	-	NA
12	Shah et al.	2003	6	87	20	ovarian cancer	30-40/10HPF	TAH+BSO+OM	-	DOD 2M
13	Agarwal et al	2003	6	55	14	NA	>10/10HPF	TAH+BSO	CT	NED>12 months
14	Pekin et al.	2000	6	56	11	ovarian tumor	14/10HPF	TAH+BSO	-	NED>25 months
15	Cheng et al.	1995	6	59	7	NA	>10/10HPF	TAH+BSO	-	NED>12 months
16	Lee et al.	1991	6	65	16. 4	fibroid	>10/10HPF	STH+BSO	CT	AWD >26 months
17	Lee et al.	1991	6	36	35	ovarian cancer	>10/10HPF	TAH+BSO	CT	AWD >33 months
18	Shimm & McDonough	1987	6	31	9	NA	8/10HPF	Resection	RT	AWD 30 months
19	Herbold et al.	1983	6	73	15	NA	21/10HPF	TAH+BSO	-	DOD 1 month
20	Raj-Kumar	1982	6	70	10	NA	<10/10HPF	Resection	-	NED>24 months
21	DiDomenico et al.	1982	6	48	11	NA	10.5/10HPF	TAH+BSO	-	NED>21 months
22	Weed & Podger	1976	6	50	11	NA	NA	TAH+BSO	-	DOD 19 months
23	Ullman & Roumell	1973	6	50	11	NA	NA	TAH+BSO	NA	NA
24	Lowel & Karsh	1968	6	50	11	NA	0-4/HPF	TAH+BSO	-	NED>12 months

Ref: Reference; NA: Not Applicable; TAH: Total Abdominal Hysterectomy; BSO: Bilateral Salpingo-Oophorectomy; OM: Omentectomy; PLN: Pelvic Lymphadenectomy; PAND: Paraaortic Lymphnode Dissection; CT: Chemotherapy; RT: Radiotherapy; NED: No Evidence of Disease, DOD: Dead Of Disease; AWD: Alive With Disease

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