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Case Report

INVASIVE LEIOMYOSARCOMA OF UTERUS IN A 22 YEAR OLD FEMALE FROM EASTERN INDIA

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ABSTRACT

Invasive uterine leiomyosarcoma is a very rare tumour mainly affecting peri-menopausal women. It presents with profuse vaginal bleeding and pelvic mass. These are highly aggressive and often show recurrence. Such tumours are rarely reported in young age group.

We here present a case of invasive uterine leiomyosarcoma in a 22 year old female. She presented with a pelvic mass without bleeding manifestations. Despite early surgery and resuscitative measures, she succumbed to the disease. The clinical profile of the disease and relevant treatment options are also discussed.

KEYWORDS: Leiomyosarcoma, uterine, menorrhagia, surgery

INTRODUCTION

Leiomyosarcoma is a rare malignancy of soft tissues of the body[1]. It can arise from any tissue containing smooth muscles like gut, uterus or blood vessels. The incidence of uterine leiomyosarcoma varies around 0.4 cases per 100 000 and it comprises around 1% of all uterine malignancies[2]. It is mostly found around menopause and the incidence increases with age[3]. These are aggressive tumours and long term prognosis is dismal. However, this type of tumour is rarely reported in young age groups. We here report a case of invasive uterine leiomyosarcoma in a 22 year old female without any risk factor. This is probably the first reported case of this type of tumour at such a young age from Eastern India.

Case Report

A 22 year old unmarried female from Kolkata presented with progressive lower abdominal swelling for one month with significant loss of weight (6 kg in last one month). She also had significant anorexia and fatigue. She complained of increased frequency of micturition. The menstrual cycles were normal and there was no fever or pain. On examination, a firm variegated fixed mass was palpable in lower abdomen with upper border at level 4 cm above umbilicus (28 weeks' pregnant uterus). Per vaginal examination did not reveal any fornix mass. Clinically there was no ascites, pedal swelling or lymphadenopathy.

Laboratory investigations revealed haemoglobin of 7 gm% with total leukocyte count of 4600/µL and platelet count of 140000/ µL. Blood urea/ creatinine and liver function tests were normal. Ultrasonographic study of abdomen revealed a lobulated heterogeneous mass in pelvis with invasion of surrounding structures. Magnetic Resonance imaging scan of abdomen revealed a huge heterogeneous mass arising from uterus with invasion of sigmoid colon (Figure 1). Retroperitoneal lymphadenopathy and minimal ascites were present. Serum CA 125 level was 118 IU/ml (N<35) and alpha fetoprotein level was normal.

At this point of time, it was decided to operate the patient for palliative purpose after 2 units of blood transfusion. Her abdominal discomfort was becoming more severe. At laparotomy under general anaesthesia, we found a huge bosselated mass arising from the fundus of uterus with invasion of sigmoid colon (Figure 2). The surface was highly vascular with areas of necrosis. We did a subtotal hysterectomy with ascitic fluid and lymph node sampling. Also, end sigmoid colostomy was done.

Sadly, post operatively the patient developed refractory hypotension with dyspnea. She had to be put on mechanical ventilator and died the next day.

Later, the ascitic fluid report came positive for malignant cells by Pap smear and biopsy of the mass revealed invasive leiomyosarcoma with vascular necrosis and areas of dedifferentiation (Figure 3).

DISCUSSION

Uterine leiomyosarcoma is an invasive malignancy arising from myometrium. They usually present with rapidly growing lower abdominal mass associated with menorrhagia[4]. In some studies, this type of tumour has been found to be associated with shoe and leather workers, although the pathophysiological connection is still debated[2]. Our patient did not have any such exposure history.

In 2008, a similar case of leiomyosarcoma in a 25 year old female was reported from south India[5]. That was probably the first reported case of leiomyosarcoma at such a young age. Like our patient, that patient also did not have menorrhagia. Thus, presenting symptoms of this malignancy may vary depending upon the age of onset. Usually these tumours are very aggressive and being rare, a proper treatment algorithm is yet undecided[5]. But whatever the treatment, chance of recurrence is very high.

A study from Pakistan showed increasing age and multiparity to be associated factors with this malignancy[6]. Usually, the preoperative imaging shows uteromegaly. Diagnosis of the type of tumour is usually possible only after pathological study and preoperative diagnosis is possible through curettage of an accessible mass[6]. However, necrosis and/or haemorrhage may be found in imaging in case of a leiomyosarcoma.

The surgical specimen is usually a vascular mass with surrounding invasions. Hepatic metastases may also be found[6].

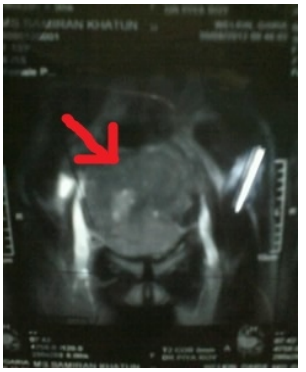

Figure 1: MRI of pelvis of the patient with the mass arising from uterus [red arrow]

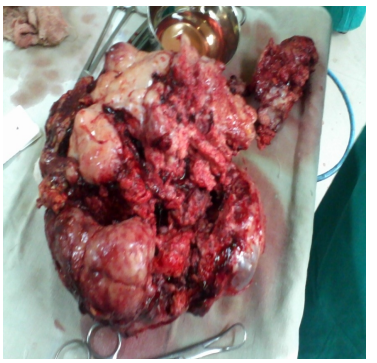

Figure 2: Surgical specimen with superficial necrosis and haemorrhage

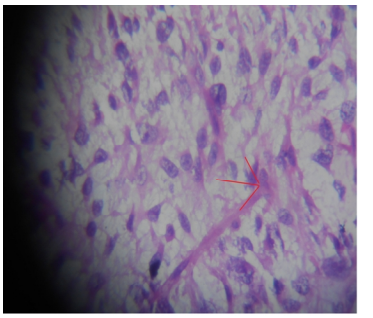

Figure 3: Histopathological appearance of the tumour with spindle shaped cells and pleomorphic nuclei and one multinucleated cell [Red Arrow]

In our case, the mass was invading into the colon. In these cases, cytoreductive surgery followed by chemoradiation may be needed.

A special problem arises in leiomyosarcomas of young patients like ours. In these cases, retaining of fertility may be an issue. Considering the aggressive nature of the tumour, conservation of female organs is often not possible. But still, ablative therapy and hormonal suppressive therapies can be considered[6].

Leiomyosarcomas in young patients are very rare. Besides uterus, they can also arise from stomach or heart[7]. However, compared to uterine variety, these other varieties often have better prognosis and often respond to surgical resection[7].

The main clinical problem in leiomyosarcoma of young persons is the threshold for suspicion. Often, these tumours are thought of as myomas and non-surgical measures like danazol are used initially[8]. Since treatment options are limited, early diagnosis is essential for favourable outcome. Often, in case of multiple myomas, one or two tumours may show malignant changes with the others benign. Thus, pathological study of all the enucleated masses is essential to detect a focus of malignancy[8]. At surgery, difficulty in enucleation, excessive bleeding and irregular surface may be the predictors of malignant change in uterine masses.

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