CASE REPORT

Epithelioid cotyledonoid leiomyoma of uterus

Indu Chawla,¹ Minakshi Bhardwaj,² Neetu Sareen,¹ Nikhil Khattar³¹Department of Obstetrics and Gynecology, Dr RML Hospital, New Delhi, India²Department of Pathology, Dr RML Hospital, New Delhi, India³Department of Urology, Dr RML Hospital, New Delhi, India**Correspondence to**Dr Indu Chawla,
induchawla2009@gmail.com**SUMMARY**

A 42-year-old para 2 women underwent laparotomy with a preoperative diagnosis of leiomyoma uterus and ovarian endometrioma. At laparotomy, uterus was 12-week size and bilateral tubes and ovaries were normal. There was a large friable reddish-brown mass dissecting the leaves of broad ligament with extension into pelvic cavity and retroperitoneal spaces. Total abdominal hysterectomy with bilateral salpingo-oophorectomy with resection of tumour was performed as there was concern about the possibility of malignancy. Histopathological examination revealed a cotyledonoid leiomyoma epithelioid variant. These tumours, though benign, raise the possibility of malignancy at the time of surgery due to their bizarre and sarcoma-like appearance. It is important for the pathologist and gynaecologist to be aware of this rare variant of leiomyoma to avoid an overtreatment

BACKGROUND

Cotyledonoid leiomyomas are rare and pose a significant problem when encountered at the time of laparotomy due to fungating appearance and infiltrative growth pattern. Awareness and recognition of this benign and unusual variant of leiomyoma by the gynaecologists will prevent an overtreatment.

CASE PRESENTATION

A 42-year-old para 2 woman presented with a history of pain in the lower abdomen and dysmenorrhoea for 2 months. Her menstrual cycles were regular and there was no history of menorrhagia. General systemic and abdominal examination was normal. On per speculum examination, cervix appeared normal and was pushed towards the right side. Bimanual examination revealed uterus of 12-week size and an ill defined, non-tender fixed mass of approximately 10–12 cm size, felt close to uterus on the left side.

INVESTIGATIONS

Transvaginal ultrasound scan showed an enlarged uterus of 13×7×6 cm in size with three round to oval fibroids, largest one measuring 4×3 cm. Another round to oval echogenic mass of 13×12 cm with a small cystic area was seen in the pelvis near cervix on the left side. The right ovary was normal and the left ovary was not seen separately from the mass. There was no evidence of free fluid in the pouch of Douglas. As the nature of mass was not clear on ultrasound MRI was performed.

MRI confirmed an enlarged uterus with multiple fibroids. A well-circumscribed lesion of 12×10×11 cm inseparable from the left ovary was

noted abutting the uterine wall and bladder. The right ovary was normal. The diagnosis of the left ovarian cystic lesion with blood as content with the possibility of endometrioma was made on MRI. CA-125 was 9.6 U/mL (ref 0–35). Biochemical and hematological investigations were normal.

DIFFERENTIAL DIAGNOSIS

In the present case, provisional preoperative diagnosis of multiple uterine myomas with the left ovarian endometrioma was made based on the clinical and MRI findings.

TREATMENT

The patient was taken for laparotomy. At laparotomy, there was no evidence of free fluid in the abdomen. The uterus was enlarged due to intramural fibroids. The tubes and ovaries appeared normal. A mass dark red to brown in colour was seen dissecting the leaves of left broad ligament and was attached to uterus near the isthmus (figure 1). The mass was approximately 10×11 cm in size, soft, friable, reddish in colour with a look of the placenta. It was extended into pelvic cavity and retroperitoneal space. The mass surrounded the left ureter and the left iliac vessels but there was no infiltration of these structures. Total abdominal hysterectomy along with removal of the mass was performed. The mass could not be removed en bloc and a few smaller pieces surrounding the ureter and iliac vessels were removed separately. As the malignant nature of mass could not be ruled out on the operation table, bilateral ovaries and tubes were also removed. The patient made an uneventful recovery.

OUTCOME AND FOLLOW-UP**Pathological findings**

On gross pathological examination, the uterus along with cervix measured 13×9×5 cm. Multiple intramural leiomyomas were seen in the uterus measuring 0.5–3 cm. The cervix was unremarkable with the exception of few Nabothian cysts. The ovaries and tubes were normal. The tumour removed from broad ligament consisted of a dark brown, friable and lobulated soft tissue mass measuring 7×7×5 cm with a few more small friable tissue fragments. On cut section, the mass was grey white with areas of haemorrhage. Microscopic examination sections revealed well-circumscribed tumour composed of cells arranged in sheets, intersecting fascicles and nodules separated by fibrous connective tissue. Cells were spindle shaped with vesicular nuclei and inconspicuous nucleoli. Some areas showed epithelioid cell morphology characterised by plump cells with abundant eosinophilic

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To cite: Chawla I, Bhardwaj M, Sareen N, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-202434

Figure 1 Preoperative photograph showing tumour mass in the broad ligament.

cytoplasm and central vesicular nuclei (figure 2). There was no cellular atypia, mitosis or necrosis. The stroma was vascularised, showed oedema and foci of hyalinisation. Features were consistent with those of epithelioid leiomyoma. No intravascular tumour was noted. The tumour did not have an intrauterine component or extension in the cervix.

FOLLOW-UP

The patient is doing well and has no evidence of recurrence when examined 5 months after surgery.

DISCUSSION

Cotyledonoid dissecting leiomyoma (Sternberg tumour), a rare tumour of smooth muscle fibres of the uterus, was first described by Roth *et al.*¹ David *et al.*² coined the term 'grapelike leiomyoma' based on the gross description of tumour. Roth *et al.*¹ analysed four cases and found that in all cases, at laparotomy, tumour extended from the uterus into the broad ligament and had a striking resemblance to the placenta, hence the term cotyledonoid dissecting leiomyoma was used. They stated that poorly supported vessels in the broken down stroma are markedly congested giving rise to a characteristic reddish-brown colour observed in the tumour. The most important reason to recognise this unusual variant is to avoid mistaking it for a malignant neoplasm. Cheuk *et al.*³ reported a case of cotyledonoid tumour, which was discovered during a surgery for prolapse uterus. A fungating mass with an appearance of sarcoma was discovered and total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. They remarked that several factors are required to produce the

gross appearance of cotyledonoid leiomyoma, namely origin close to serosal surface, dissecting growth and perinodular hydropic degeneration.

Saeed *et al.*⁴ reported a case of cotyledonoid dissecting leiomyoma in a 27-year-old woman, for whom total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. He remarked that these tumours have a distinctive gross appearance raising the suspicion of sarcoma. Intraoperative frozen section is helpful in these cases to avoid an overtreatment. He analysed 20 cases reported in the literature and found that the age of patients ranged from 23 to 65 years (mean 40.3 years). Fifteen cases were treated with abdominal hysterectomy, one with subtotal hysterectomy and four with tumour resection. Intraoperative frozen section performed in four cases revealed a benign leiomyoma and only tumour resection was performed in three of them.

In the literature, two types of cotyledonoid leiomyoma have been described, namely, cotyledonoid dissecting leiomyoma, where exophytic mass is in continuity with intramural tumour^{3 5 6} and cotyledonoid dissecting leiomyoma which lacked the intramural component.^{7 8} The present case did not have any intramural component. In most of the cases reported in the literature, tumour was composed of spindle-shaped smooth muscle cells with areas of hyalinisation and myxoid change.^{3–8} However, in the present case, in addition to the spindle-shaped smooth muscle cells, there were areas showing epithelioid cell morphology. No such case showing an admixture of spindle cells and epithelioid cells has been reported until date. An epithelioid cell variant of cotyledonoid dissecting leiomyoma was reported by Majd *et al.*⁹ with no adverse outcome. This case consisted of sheets of only epithelioid cells.

Learning points

- ▶ Cotyledonoid leiomyomas are rare.
- ▶ May be mistaken for malignant growth because of appearance.
- ▶ Awareness of this benign entity can prevent an overtreatment.

Contributors IC was involved in operating surgeon and wrote the manuscript; MB and NS searched the literature; and NK reviewed the manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Figure 2 Histopathology showing admixture of smooth muscle cells and epithelioid cells.

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