# COTYLEDONOID DISSECTING LEIOMYOMA (CDL) OF UTERUS MIMICKING MALIGNANCY: A CLINICAL DILEMMA

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**ABSTRACT**

Cotyledonoid Dissecting Leiomyoma (CDL) or Sternberg tumour is an unusual variant of leiomyoma giving a distinctive malignant gross appearance. We report here a case in 50-year-old woman who presented with Abnormal Uterine Bleeding (AUB) and abdominal mass. Laparotomy showed a large exophytic mass arising from the uterus with irregular contours and variegated appearance suspicious of malignancy.

**KEYWORDS**

Cotyledonoid, Dissecting, Immunohistochemistry, Leiomyoma, Uterus.

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## INTRODUCTION

Leiomyomas are one of the most commonly encountered smooth muscle neoplasms of the uterus. Several variations in the pattern of growth known to occur in them include multinodular appearance and an infiltrative behaviour.[1] One of the most unusual and least common infiltrative variant of leiomyoma is Cotyledonoid Dissecting Leiomyoma (CDL) or Sternberg tumour. The clinical importance is that it grossly resembles cotyledons of placenta with a fleshy sarcomatoid appearance.[2] It exhibits infiltrative pattern resembling malignant tumour on both macro and microscopic evaluation.[3] Worrisome appearance of this lesion can lead to overtreatment by hysterectomy, especially in females of reproductive age group.[4] Here, we report a case of infiltrative cotyledonoid leiomyoma presenting as a malignant exophytic growth in the uterus.

## CASE REPORT

A 50-year-old multiparous lady (Gravida 3, Para 3) with previous two lower segment caesarean sections presented with chief complaints of dysmenorrhea and AUB for the past 4 months. General physical examination showed pallor with normal vital signs. Chest and Cardiovascular examination revealed no abnormality. Per abdomen examination revealed a large intrapelvic mass. Per speculum examination showed cervix to be pulled up with minimal erosions. Per vaginal examination revealed 12 weeks size uterus with an irregular mass not separate from the uterus. Laboratory investigation revealed moderate anaemia with haemoglobin of 7.3 gm% and mildly raised CA-125 of 67IU. Other laboratory parameters and X-ray chest were within normal limits. Ultrasonography (U/S) abdomen showed bulky uterus with presence of heterogeneous mass measuring 7 x 5 x 4 cms, showing increased vascularity in the left posterolateral wall. Right ovary showed a retention cyst of size 3.0 cms, while left ovary was cystic, enlarged of size 10.6 x 4.9 cms and appeared to be adherent to the left side of the uterine wall. Magnetic Resonance Imaging (MRI) showed a large Space Occupying Lesion (SOL) in the Pouch of Douglas (POD) with peripherally arranged haemorrhagic cysts suggestive of enlarged left ovarian mass with torsion along with intramural fibroid, uterus and simple cyst, right ovary. Clinical diagnosis of fibroid uterus with left ovarian mass was made.

Laparoscopic visualization showed a big mass in the POD pushing the uterus anteriorly and having a variegated appearance suggestive of malignancy. Laparotomy performed thereafter showed a large 10 x 10 x 3.0 cms mass arising from left lateral posterior uterine wall inseparable from the uterus. Both the adnexa were normal with no deposits on liver, omentum or peritoneum. No free fluid was present in the pelvic cavity. Intraoperative diagnosis of leiomyosarcoma was suspected due to large size, variegated appearance and inseparable plane of the mass. Total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH and BSO) was done along with biopsy from omentum and peritoneum, all of which were sent for histopathological examination.

Gross examination of the hysterectomy specimen showed an irregularly contoured exophytic, congested, partly cystic mass measuring 10.1 x 5.8 x 3.2 cms on the serosal aspect of lower left lateral posterior wall of the uterus (Fig. 1a & 1b). Cut surface of the mass showed greyish-white to greyish-brown firm solid areas with multiple thin walled cysts containing haemorrhagic and mucoid material (Fig. 1c). The mass had poorly defined borders with the myometrium (Fig. 1d). Myometrium showed trabeculations and an intramural fibroid measuring 2 x 2 x 1.8 cms. Both the fallopian tubes and ovaries were unremarkable.

Microscopic examination revealed a tumour comprised of oval-to-spindle shaped cells with mildly pleomorphic nuclei, inconspicuous nucleoli and moderate-to-abundant eosinophilic cytoplasm. The cells were seen arranged in interlacing fascicles, whorls and bundles dissecting into the myometrium at places (Fig. 2a). Focal areas showed peri-nodular hydropic degeneration with thick-walled ectatic and congested blood vessels (Fig. 2b), interspersed haemorrhage, extensive hyalinization and mucoid degeneration. No

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intravascular growth, increase in cellularity, mitosis or Coagulative Tumour Necrosis (CTN) seen. Few entrapped endometrial glands were also seen amidst the tumour cells (Fig. 2c). Tumour cells stained positive for Masson Trichrome stain (MT). Immunohistochemistry done showed tumour cells showing positivity with h-Caldesmon and Smooth Muscle Actin (SMA) (Fig. 3a, b); Oestrogen Receptor (ER) positivity was seen in a few entrapped endometrial glands (Fig. 3c). CD10 was negative in tumour cells (Fig. 4a), thus ruling out endometrial stromal origin of tumour. The cells showed low proliferative index (<2%) on Ki 67 MIB (Fig. 4b). Hence diagnosis of Cotyledonoid Dissecting Leiomyoma (CDL) was made. There was marked adenomyosis in the myometrium and a small intramural leiomyoma. Both the fallopian tubes showed chronic salpingitis. Right ovary showed cystic follicles and haemorrhagic cystic corpus luteum, while left ovary showed corpora albicantia. Histology from peritoneum and omentum showed congestion only.

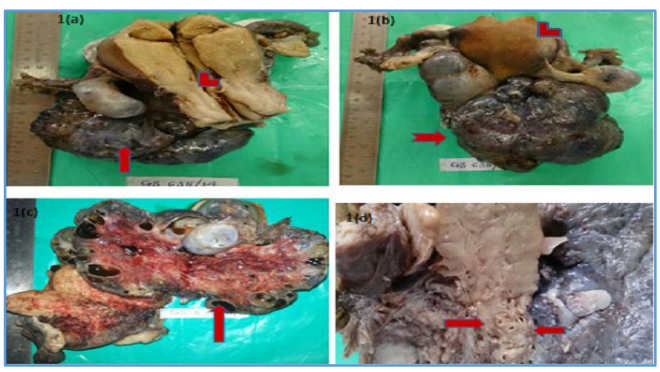


***Fig. 1: (a, b) Gross Finding of Uterus (Arrow Head) shows Subserosal Exophytic Congested Mass on the Posterior Aspect (Arrow),***

***(c) Cut Section showing Greyish Brown Solid Areas with a Few Thin Walled Cysts containing Haemorrhagic and Mucoid Material (Arrow),***

***(d) Mass showing Poorly Defined Borders with the Myometrium (Arrows)***

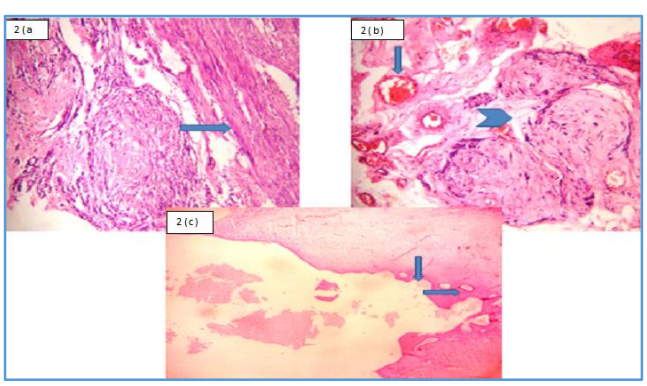


***Fig. 2 (a): Photomicrograph showing the Tumour Dissecting into the Myometrium (Arrow)***

***Fig. 2 (b): Showing Perinodular Hydropic Degeneration (Arrow Head) and Congested Ectatic Blood Vessels (Arrow)***

***Fig. 2 (c) Showing Cyst Lined by Pseudostratified Columnar Epithelium with Embedded Endometrial Glands (Arrows)***

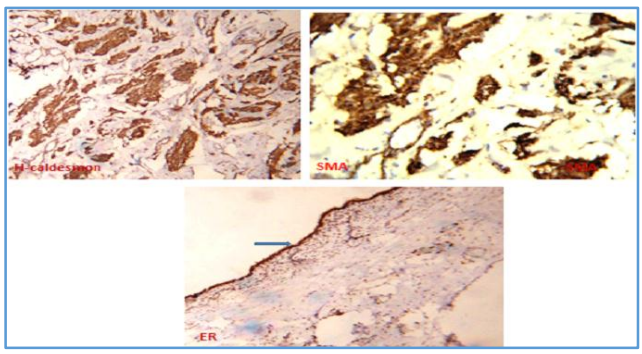


***Fig. 3: (a) IHC Showing H-Caldesmon Positivity in Tumour Cells Groups***

***(b) IHC Showing SMA Positivity in Tumour Cells***

***(c) IHC Showing ER Positivity in Cystic and Dilated Endometrial Glands (Arrow)***

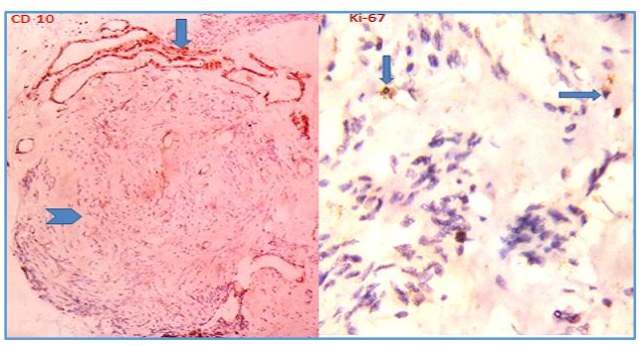


***Fig. 4 (a): Showing CD 10 Positivity around Blood Vessels (Arrow); Tumour Cells were Negative (Arrow Head)***

***(b) Shows Low Proliferative Index (Ki-67) (Arrows)***

**DISCUSSION**

Grape like leiomyoma of uterus, CDL, is an extremely rare and unusual variant of uterine leiomyoma.[5] Thereafter Sternberg in 1979 described it as "proliferating pelvic angio-leiomyomatosis (Red seaweed lesion)."[6] This tumour presents in a wide age range of 23-73 years, mean age 45 yrs. with complaints of AUB, abdominal mass, pain, dysmenorrhea, infertility, prolapse, uterus or even as incidental finding.[1,2,7-9] Our patient was a little older perimenopausal lady with AUB and hence the concern for malignancy. CDL commonly arises from the fundus or posterior aspect of cornu uterus and is characterized by reddish, exophytic, expansile, spongy, bulbous, placenta-like gross appearance by its extrauterine extension into the pelvic cavity raising possibility of malignancy intraoperatively.[1-12] which was seen in our case.

The dark red appearance of exophytic component is due to venous obstruction and congestion with tan white endophytic parent intramural leiomyoma.[2] This intramural continuation is apparent only on diligent sectioning of uterine wall, hence giving appearance of separate adnexal mass.[13] as was diagnosed on ultrasonogram of our patient. The tumour size ranges from 4-41 cms (mean 15.4 cms) with no side predilection and may even be bilateral.[1,2,3,8]

Microscopically, the tumour is composed of variable sized micronodules of muscle fascicles with marked hydropic change. The tumour shows very minimal or no cellular atypia

with no increase in mitotic activity (<1 mitosis/10 HPF) or CTN. The cells infiltrate the myometrium in dissecting pattern.

The tumour cells give positive staining with smooth muscle markers (SMA, Desmin, Vimentin, h-caldesmon), but negative for CD-10 and S-100 protein.[3,7] Similar microscopic features were seen in our patient.

Tumours with hydropic degeneration when present in abundance are close mimics for CDL.

Leiomyoma with Perinodular Hydropic Degeneration (LPHD) show abundant hydropic degenerative change surrounding muscle bundles distorting the architectural pattern and extend outside the uterus.[10,14] Intravenous Leiomyomatosis (IVL) displays intravascular luminal growth outside the main tumour mass, while lesions with vascular invasion show intravascular growth within the tumour body. IVL has similar gross morphological characteristics; however, it usually does not show any congestion.[15] All three- CDL, LPHD and IVL are considered to be related lesions.

Myxoid leiomyosarcoma which has minimal cytological atypia and low mitotic count can also mimic CDL. Lack of gross appearance, infiltrative growth pattern and high Ki67 labelling index of more than 60% help differentiate them from CDL.[3,7,8] To conclude, the authors would like to emphasize that one must be aware of this rare variant of leiomyoma, advocate careful assessment of morphologic features, diligent search for atypia and confirmation of proliferative index by IHC for correct diagnosis to prevent overtreatment, especially in females of reproductive age group.

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