Case report

A 60-year-old woman presented with the complaint of a 1-year history of lower abdominal pain and third-degree uterovaginal descent. Her previous menstrual history was unremarkable. A vaginal examination revealed a bulky uterus with an ulceration on the anterior cervical lip. Ultrasonography revealed that the uterus was 7.5×3.4 cm in size with an endometrial thickness of 4.9 mm. Multiple uterine fibroids 2–4 cm in diameter were also noted. Both adnexa were unremarkable. A hysterectomy was performed and the specimen was sent for histopathological examination. Grossly, the cervix appeared hypertrophied and epidermidized. The endometrial thickness was 1.6 cm, while the endometrial thickness was 0.4 cm. Multiple fibroids up to 4×3.5 cm were noted. In addition, 2 subserosal fibroids 2 cm and 3.5 cm in diameter were seen. A cut section of the largest intramural fibroid and 1 subserosal fibroid revealed a solid, grayish-white, homogenous, and whorled appearance. The cut section of another subserosal fibroid revealed the presence of multiple grayish-white nodules (Fig. 1A). Microscopy revealed nodules of varying sizes of uniform smooth muscles arranged in interlacing and whorling fascicles with few prominent blood vessels (Fig. 1B–D). However, no signs of significant mitotic activity, nuclear atypia, or necrosis were seen. Based on the gross features and microscopic findings, a final diagnosis of CDL was made. Immunohistochemistry performed
on CDL sections revealed diffuse positivity for vimentin, desmin, and smooth muscle actin, confirming the histopathological diagnosis of CDL (Fig. 2). In the present case, this rare variant of leiomyoma was associated with multiple intramural and subserosal classical leiomyomas showing features of hyaline degeneration (Fig. 3A). The endometrium was in the proliferative phase and adenomyosis was noted in the myometrium (Fig. 3B). The cervix showed features of acute and chronic cervicitis with surface ulceration and keratinization.

**Discussion**

Leiomyomas are the most common benign smooth muscle neoplasms of the uterus. A number of patterns of leiomyomas have been described. CDL, a very rare variant that is also
| SN  | Reference             | Age | Clinical presentation       | Tumor size (cm) maximum dimension | Tumor location                      |
|-----|-----------------------|-----|----------------------------|----------------------------------|-------------------------------------|
| 1   | David et al. [1]      | 65  | Abnormal uterine bleed     | 15                               | Uterine fundus and cervix           |
|     |                       | 48  | Uterine prolapse            | 12                               | Uterine fundus                      |
| 2   | Roth et al. [2]       | 39  | Pelvic mass                 | 10.3                             | Uterine cornua                      |
|     |                       | 41  | Abnormal uterine bleed      | 10                               | Uterine cornua                      |
|     |                       | 23  | Pelvic mass                 | 25                               | Uterine cornua                      |
|     | Unknown               |     | Pelvic mass                 | 24                               | Uterine cornua                      |
| 3   | Brand et al. [3]      | 24  | Abdominal mass              | NA                               | Uterine fundus                      |
| 4   | Roth and Reed [4]     | 46  | Pelvic mass                 | 34                               | Uterine cornua                      |
| 5   | Kim et al. [5]        | 26  | Incidental                  | 12                               | Posterior uterine wall              |
| 6   | Cheuk et al. [6]      | 55  | Abnormal uterine bleed      | 10                               | Uterine cornua                      |
| 7   | Stewart et al. [7]    | 58  | Abdominopelvic mass         | 16.4                             | Uterine fundus                      |
| 8   | Jordan et al. [8]     | 46  | Right adnexal mass          | 22                               | Uterine with extraterine extension  |
|     |                       |     | Pelvic mass                 | 20                               |                                     |
|     |                       |     | Pelvic mass                 | 10                               |                                     |
|     |                       |     | Pelvic mass                 | 18                               |                                     |
|     |                       |     | Abnormal uterine bleed      | 13                               |                                     |
|     |                       |     | Uterine mass, infertility   | 18                               |                                     |
| 9   | Saeed et al. [9]      | 27  | Pelvic mass                 | 41                               | Uterine fundus                      |
| 10  | Maimoon et al. [10]   | 40  | Urinary retention           | 10                               | Uterine isthmus                     |
| 11  | Shelekhova et al. [11]| 73  | Uterine mass                | 8                                | Uterine fundus                      |
| 12  | Gurbuz et al. [12]    | 67  | Incidental                  | 10                               | Uterine cornua                      |
| 13  | Weissferdt et al. [13]| 52  | Abnormal uterine bleed      | 6.2                              | Uterine fundus                      |
| 14  | Raga et al. [14]      | 33  | Abdominal pain              | 6                                | Lateral part of uterus              |
| 15  | Driss et al. [15]     | 47  | Pelvic mass                 | 25                               | Uterine with extraterine extension  |
| 16  | Preda et al. [16]     | 41  | Uterine mass                | 9                                | Left and posterior uterine wall     |
| 17  | Fukunaga et al. [17]  | 56  | Constipation                | 30                               | Posterior uterine wall              |
|     |                       |     | Abdominal pain              | 26                               | Posterior uterine wall              |
|     |                       |     | Abnormal uterine bleed      | 4                                | Posterior uterine wall              |
|     |                       |     | Abdominal pain              | 18                               | Lateral uterine wall                |
| 18  | Gezginç et al. [18]   | 57  | Pelvic pain                 | 2.5, 4.5                         | Intrauterine, lateral uterine wall  |
| 19  | Agarwal et al. [19]   | 52  | Abnormal uterine bleeding   | 10                               | Uterine cornua                      |
| 20  | Ersöz et al. [20]     | 51  | Abnormal uterine bleeding   | 8.5                              | Subserosal                          |
| 21  | Roth et al. [21]      | 33  | Abnormal uterine bleeding   | 6.5, 13.5                        | Posterior uterine wall              |
| 22  | Tanaka et al. [22]    | 36  | Incidental                  | 10                               | Posterior & lateral uterine wall    |
| 23  | Onu et al. [23]       | 50  | Incidental                  | 10                               | Uterine fundus                      |
| 24  | Kim et al. [24]       | 43  | Abdominal mass              | 13                               | Uterine with extraterine extension  |
| 25  | Blake et al. [25]     | 56  | Abnormal uterine bleeding   | 30                               | Uterine with extraterine extension  |
| 26  | Shimizu et al. [26]   | 40  | Abnormal uterine bleeding   | 10                               | Posterior uterine wall              |
| 27  | Rocha et al. [27]     | 38  | Abnormal uterine bleeding   | 25                               | Uterine isthmus                     |
| 28  | Xu et al. [28]        | 55  | Pelvic mass                 | 6                                | Posterior uterine wall              |
|     |                       | 43  | Pelvic mass                 | 3                                | Body of uterus                      |
|     |                       | 37  | Pelvic mass                 | 30                               | Peri-uterine                        |
| 48  | Lower abdominal pain  | 6.7 |                             |                                  | Right wall of uterus                |
commonly known as Sternberg tumor, was first reported by Roth et al. [3] Menolascino-Bratta et al. [4] coined the term “angionodular dissecting leiomyoma”. These tumors are frequently seen in the third to sixth decades of life. The most common complaints are lower abdominal pain and abnormal uterine bleeding. The apex case also presented with complaints of lower abdominal pain; however, no vaginal bleeding was revealed. Tumor size was typically 2–15 cm [4,5]. Three types of CDL have been described in the literature. The first appears as an exophytic mass of multinodular tissue protruding from the lateral surface of the uterine cornua; resembling the placenta is called CDL. The second type is an intramural dissecting tumor that is confined to the uterus. These 2 types share similar histopathological features. The last type is pure cotyledonoid leiomyoma, which is not associated with either a parent intramural mass or intramural dissection [6]. This case met the criteria for exophytic CDL. Microscopically, it is characterized by nodules of various sizes of uniform smooth muscles arranged in interlacing and whorling fascicles. Many blood vessels are also prominent with focal hypercellular areas. However, in contrast to malignant lesions, signs of mitotic activity, nuclear atypia, cellular pleomorphism, and necrosis are absent. Vascular invasion, capsular infiltration, and metastasis are not seen. In a few cases, perinodular hydropic changes may be prominent [7]. A variety of other unusual patterns of uterine leiomyoma have been described, such as parasitic leiomyoma, cellular leiomyoma, symplastic or bizarre leiomyoma, epithelioid leiomyoma, intravenous leiomyomatosis, and leiomyoma with secondary changes. Some CDL appear as large fungating masses with widespread extension into the broad ligament and pelvic cavity. Due to its rarity and a clinician’s lack of familiarity, such tumors are sometimes misdiagnosed as malignancies [8]. Gurbuz et al. [9] reported a case of cotyledonoid leiomyoma that had no intrauterine portion but had extraterine extensions. A comparative analysis of various CDL cases reported in the literature is given in Table 1 and Supplementary Data 1. In conclusion, CDL is a unique and rare variant of leiomyoma with a characteristic gross nodular appearance and microscopic features. Increasing awareness among clinicians and pathologists regarding this rare entity will prevent inappropriate diagnosis and treatment.

Conflict of interest
No potential conflict of interest relevant to this article was reported.

Ethical approval
The study was done in adherence to the Institutional Ethics committee guidelines (All India Institute of Medical Sciences, Patna, Bihar, India). As the study is purely based on tissue sample and slides which were used after routine reporting and no clinical/drug trial was done it was not registered in ethics committee as per Institutional Ethics committee guidelines, hence Institutional Review Board (IRB) number was not obtained. The study performed in accordance with the principles of the Declaration of Helsinki. Written informed consents were obtained.

Patient consent
The present work was performed after the patient provided informed consent, and a sincere effort has been made to uphold patient confidentiality.

Supplementary material
Supplementary Data 1 associated with this article can be found online at https://doi.org/10.5468/ogs.2019.62.5.362.

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