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■ CASE STUDY ■

Cotyledonoid Dissecting Leiomyoma of the Uterus with Intravascular Luminal Growth: A Case Study

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Here, we report the case of a 43-year-old female who was diagnosed with a cotyledonoid dissecting leiomyoma (CDL) of the uterus. CDL is a recently described and extremely rare variant of a benign leiomyoma that can grossly masquerade as a malignancy. The 13-cm sized tumor was located primarily on the extrauterine surface as an intrauterine continuity, which showed dark red, congested, bulbous protuberances. It was multinodular appearance, encasing the bilateral adnexae and the left iliac vein. Microscopically, the nodules were separated by extensive hydropic degeneration. The nodules were composed of cigar-shaped spindle cells with no mitosis, cellular pleomorphism or coagulation necrosis. They also showed an intravascular luminal growth pattern. CDL with intravascular growth was diagnosed after excluding intravascular leiomyomatosis, disseminated peritoneal leiomyomatosis, and benign metastasizing leiomyoma. The present case is the second reported case of CDL in Korea. Recognition of this rare and bizarre, malignancy-mimicking leiomyoma is crucial to prevent inappropriate treatment.

Key Words: Cotyledonoid; Leiomyoma; Intravascular; Uterus

Cotyledonoid dissecting leiomyoma (CDL), also known as a Sternberg tumor or "grape-like" leiomyoma, is an extremely rare variant that mimics malignant gross features but has a benign histology and biological behavior.[1] The name "cotyledonoid" leiomyoma originated from the characteristic gross morphology, which includes extrauterine dark red, spongy, bulbous protuberances, also resembling a placenta.[2] To date, 41 cases of CDLs have been reported in the English literature.[1-9] Here, we report an additional case of CDL. To the best of our knowledge, the present case is the second reported case of CDL in Korea.[3] Our case also showed intravascular luminal growth, which is very uncommon in CDLs of the uterus.

Here, we emphasize that clinicians should be aware of this rare entity to prevent unnecessary aggressive treatment due to its warning gross morphology.

CASE REPORT

A 43-year-old Korean female (gravida 2, para 2) with a three-year history of a palpable abdominal mass was admitted to the gynecology outpatient department. She had a history of two caesarian sections. The patient had been followed up for 12 months. A recent evaluation showed a marked increase in tumor size (up to approximately 8.0 cm) and the presence of abdominal pain. Abdominopelvic computed tomography showed a large enhancing mass measuring 13.0 cm that encased the left adnexa, the uterus, and the left iliac vein (Fig. 1). Omental haziness and small seeding nodules suggested the possibility of seeding carcinomatosis peritonei. Intraoperatively, no collection of pelvic cavity fluid was found. Total abdominal hysterectomy and bilateral salpingo-oophorectomy were performed with removal of the parametrially extended tumor. Grossly, the multinodular mass was dark red, congested and spongy with bulbous protuberances over the extrauterine surface and bilateral broad ligaments (Fig. 2A). Contrary to the dark red color of the external surface, the cut surface showed a homogeneous, grayish tan-colored, multinodular, firm trabeculated appearance (Fig. 2B). No intravascular growth or invasion was identified on gross examination. Leiomyosarcoma was initially suspected. Microscopically, the excised mass was composed of variably sized multiple nodules,

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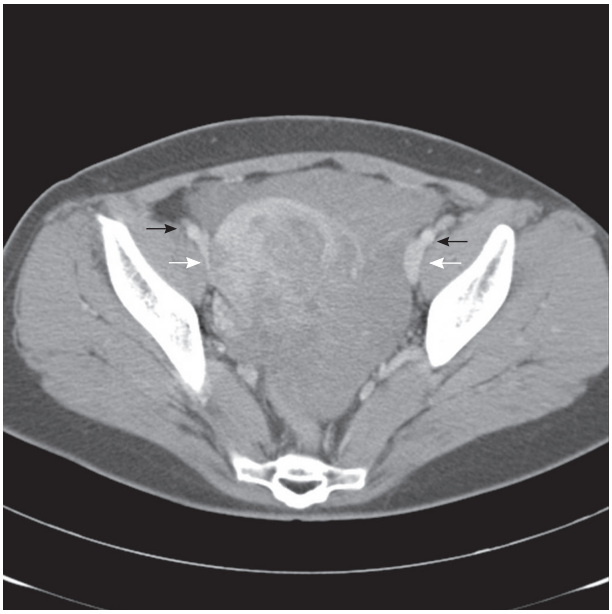

Fig. 1. Radiologic finding. Abdominopelvic computed tomography reveals a huge pelvic mass encasing the ovaries and salpinges as well as the iliac vein (white arrows) on the left side. Black arrows indicate the iliac arteries.

which were surrounded by perinoudular widened hydropic degeneration (Fig. 3A). The nodules consisted of fascicles of spindle cells with no cellular pleomorphism, mitotic activity, or coagulation necrosis (Fig. 3B, C). The mass encased the bilateral ovaries and salpinges, which were free from tumor invasion. Intravascular protruding growth was also found (Fig. 3D). Immunostaining for CD34 highlighted the endothelial cells of the blood vessels surrounding the foci of the intravascular growth of the leiomyoma (Fig. 3E). The spindle cells were positive for muscle-specific actin (IA4, prediluted, Dako, Glostrup, Denmark) and desmin (D33, prediluted, Dako). They were negative for CD10 (56C6, prediluted, Dako). CDL with an intravascular growth pattern was diagnosed.

DISCUSSION

CDL, the least variant of benign uterine leiomyomas, is characterized by extrauterine bulbous expansile growth continuous to the intrauterine location of the mass, mimicking a grape-like appearance.[1-3] Due to its bizarre gross morphology (including a large size, widespread invasive growth, etc.) and status as a rarely encountered, unfamiliar variant, an intraoperative impression of malignant neoplasm, especially leiomyosarcoma, was initially suspected. However, uterine CDL can be distinguished from leiomyosarcoma in that the former has a consistency that is not soft but firm instead. In addition, macroscopic differential diagnoses, such as sarcomas (e.g. leiomyosarcomas), can be easily excluded after microscopic observation.[1,4] Microscopic differential diagnoses included intravascular leiomyomatosis, disseminated peritoneal leiomyomatosis, and benign metastasizing leiomyoma. Intravascular leiomyomatosis is characterized by a grossly visible vascular invasion of a histologically proven benign smooth muscle tumor.[10] Disseminated peritoneal leiomyomatosis shows numerous small (usually less than 2 cm in size) smooth muscle tumorlets spread across the peritoneum.[11] Benign metastasizing leiomyoma is a morphologically benign leiomyoma occurring at distant extrauterine sites, such as the lung or regional lymph nodes.[12]

The exact nature of CDL remains unclear. The exophytic dark red portion of the CDL may be caused by venous obstruction and congestion, and the endophytic intramyometrial portion with poorly defined borders may arise from the parent leiomyoma between the myometrium.[2]

Ultrasonographic findings are not distinct from other conventional leiomyomas. Magnetic resonance images of CDLs show heterogeneous intensity with patchy isosignal nodules and stromal components with a high signal on T2-weighted images.[5] The high signal intensity reflects perinodular hydropic degeneration.[6]

After reviewing the 42 previously reported cases of CDLs (including the present one), all the cases showed no recurrence or metastases during the follow-up period.[1] Clinically, most cases occur in women of reproductive age, ranging from 23 to 73 years (mean, 44 years). Grossly, CDLs are composed of an exophytic subserosal mass and an intramural endophytic portion with continuity. The size of the reported CDLs ranged from 4 to 41 cm (mean, 14.2 cm). Microscopically, the nodules are separated by a fibrovascular connective tissue with marked hydropic changes or congestion filled with numerous vascular tissues.[2] The spindle cells show no cellular atypia, mitosis or coagulation necrosis with a dissecting growth pattern due to perinodular extensive hydropic changes.[1,2] Intravascular growth is rare, and only 9 cases (21.4%) have been reported in the 42 published cases of CDLs, which includes the present case.[1,4,7-9] Foci of intravascular luminal growth pattern may cause misdiagnosis of the condition as a malignancy when present. Intravenous leiomyomatosis behaves in a benign fashion although vascular invasion may extend as far as the extrauterine pelvic veins, the inferior vena cava, and even up to the right side of the heart.[10] Similarly, CDLs with intravascular luminal growth have been known to pursue a benign nature but with uncertain bio-

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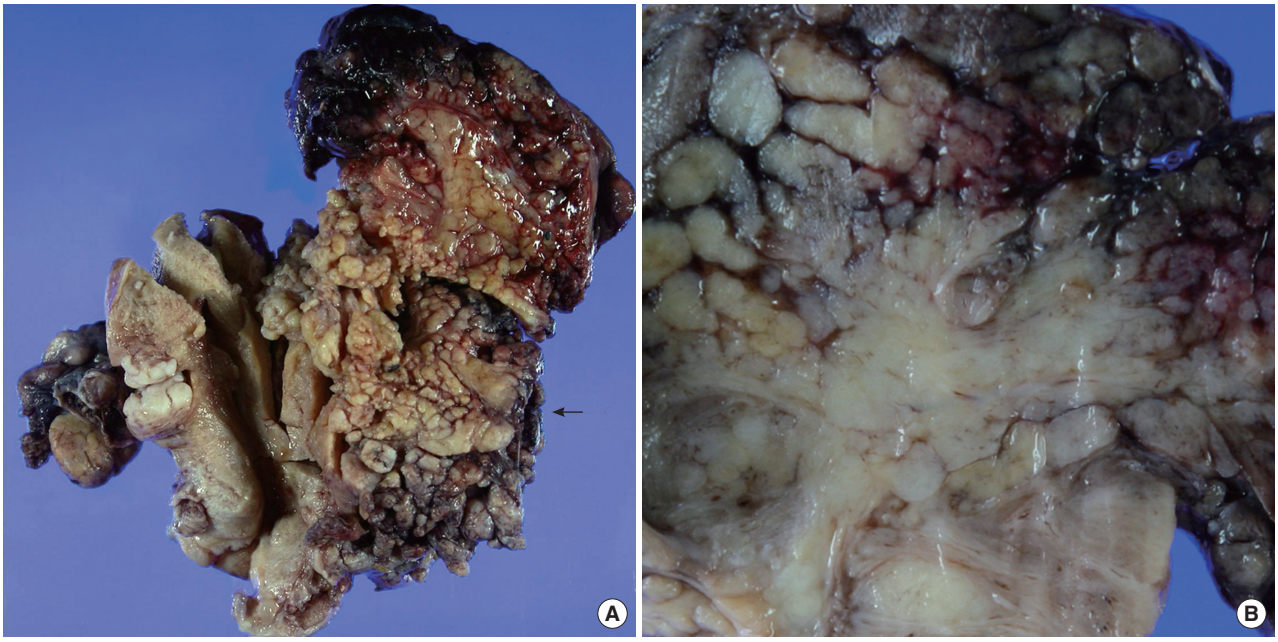

Fig. 2. Gross finding. (A) The resected uterus shows an exophytic bosselated mass with a dark red and firm appearance. Note the multinodular exophytic part (arrow) of the mass extending into the broad ligament. (B) The cut surface is a mass of rubbery reddish and solid multiple nodules, measuring from 0.4 to 3.0 cm in their greatest dimension.

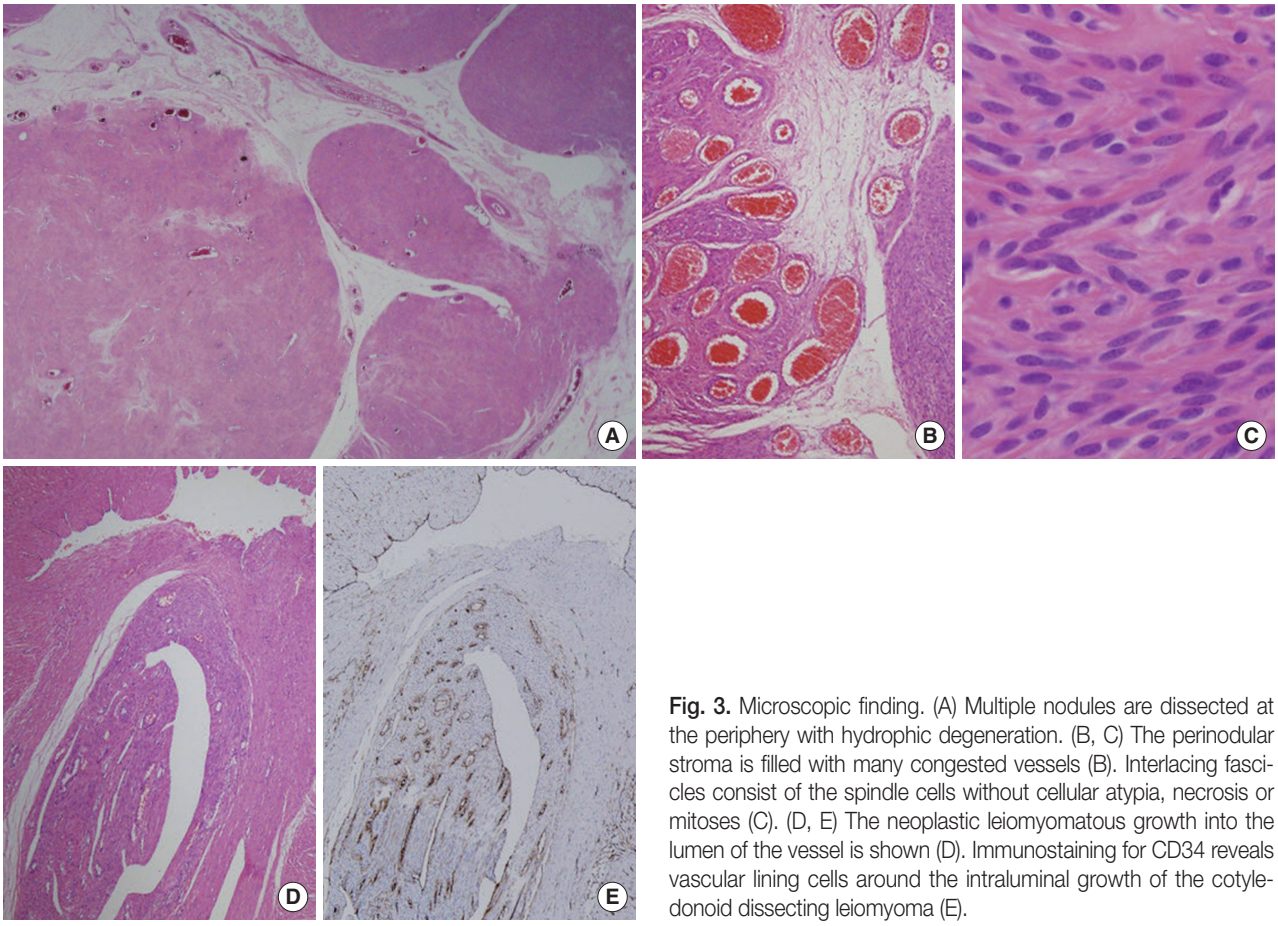

Fig. 3. Microscopic finding. (A) Multiple nodules are dissected at the periphery with hydrophic degeneration. (B, C) The perinodular stroma is filled with many congested vessels (B). Interlacing fascicles consist of the spindle cells without cellular atypia, necrosis or mitoses (C). (D, E) The neoplastic leiomyomatous growth into the lumen of the vessel is shown (D). Immunostaining for CD34 reveals vascular lining cells around the intraluminal growth of the cotyledonoid dissecting leiomyoma (E).

logical behavior.[7] Further accumulated cases will be required to establish the prognostic impact of intravascular luminal growth in these rare entities.

To avoid misdiagnosis, especially in intraoperative frozen diagnosis, acknowledgment of this rare entity is important because it is known to have a benign biological behavior that mimics the gross morphology of a malignant tumor.

Conflicts of Interest

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

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