J Korean Med Sci 2002; 17: 840-4
ISSN 1011-8934



# Cotyledonoid Dissecting Leiomyoma of the Uterus
## : A Case Report and Review of the Literature

"Cotyledonoid dissecting leiomyoma" or "Sternberg tumor" is a very rare variant of smooth muscle tumors with a distinctive gross appearance. We describe a similar lesion, probably the sixth reported case, comparing its clinicopathological features with those of previous cases. A 26-yr-old nulliparous woman underwent laparotomy for a large pelvic mass replacing the postero-lateral aspect of the uterus with extension into the left pelvic cavity in the form of numerous exophytic congested small nodules. The tumor was removed by resection without hysterectomy after frozen section examination. Histologically, there were variable sized micronodules of benign smooth muscle fascicles, which were separated by fibrous connective tissue with a marked hydropic change and rich vascularity. Immunohistochemical and ultrastructural studies were helpful for confirmation of the smooth muscle nature, but not useful for the definitive diagnosis. Due to bizarre, sarcoma-like gross appearances, this type of lesion should be subjected to frozen section examination in order to avoid overtreatment and preserve the fertility in young women.

Key Words : *Uterus; Leiomyoma; Frozen Sections; Immunohistochemistry; Microscopy, Electron*

**Mi-Jin Kim, Yoon-Ki Park*, Jae-Ho Cho†**

Department of Pathology, Obstetrics and Gynecology*, and Radiology†, Yeungnam University College of Medicine, Daegu, Korea

Received : 28 August 2001
Accepted : 18 December 2001

**Address for correspondence**
Mi-Jin Kim, M.D.
Department of Pathology, Yeungnam University College of Medicine, 317-1 Daemyung-dong, Nam-gu, Daegu 705-717, Korea
Tel : +82-53-620-3334, Fax : 82-53-656-1429
E-mail : mjkap@med.yu.ac.kr

## INTRODUCTION

Leiomyoma is the most common neoplasm of the female genital tract and conventional leiomyomas are usually easy to diagnose. Some variant forms with unusual infiltrative growth pattern have been known including intravenous leiomyomatosis, leiomyoma with vascular invasion, diffuse leiomyomatosis, and disseminated peritoneal leiomyomatosis (1). Recently, cotyledonoid dissecting leiomyoma was proposed for a new form of uterine leiomyoma with histologically benign, but distinctive gross features resembling cotyledon of the placenta (2). It is indicated to perform frozen sections because of the sarcomatoid operative appearance of the lesion. The recognition of this condition by surgeons and pathologists will prevent radical hysterectomy and preserve fertility in young female patients. This report describes a case of cotyledonoid dissecting leiomyoma of the uterus, comparing the gross, microscopic, immunohistochemical, and ultrastructural findings of the lesion with those of previously reported cases.

## CASE REPORT

A 26-yr-old nulliparous woman presented with an uterine mass, incidentally detected on ultrasound examination for the confirmation of pregnancy, associated with a recent onset of vaginal bleeding. Her pregnancy was terminated by spontaneous abortion 2 months before admission. Ultrasonogram and pelvic magnetic resonance imaging (MRI) showed a large, 12-cm sized, irregular contoured mass in the left lateral aspect of the uterus, which was presumed to be a subserosal leiomyoma (Fig. 1). She underwent laparotomy for the huge pelvic mass. At operation, there were numerous exophytic congested small nodules, resembling placental cotyledons, extending from the uterine wall posteriorly into the left broad ligament and pelvic cavity, as well as two small typical intramural leiomyomas, one of which seemed to be in continuity with the extrauterine lesion. An operative impression of sarcoma by surgeon turned out to be a histologically benign smooth muscle tumor based on frozen section. The extrauterine component was incompletely resected to save the uterus.

The specimen consisted of innumerable fragments of nodules weighing up to 150.0 g. The nodules were rubbery reddish and solid, ranging from 0.5 to 5.5 cm in diameter, and these nodules were closely packed and connected by thin fibrous tissue containing engorged vasculature (Fig. 2). Microscopically, the tumor showed variable sized micronodules of muscle fascicles, which were separated by fibrous connective tissue with a marked hydropic change and rich vascularity, resulting in a multinodular appearance (Fig. 3). The fascicles of smooth muscle cells showed not only the ordinary features of leiomyoma but also a disorganized, swirled appearance with slightly enlarged nuclei. There were neither significant nuclear atypia nor mitotic figures. No coagulative tumor cell necrosis

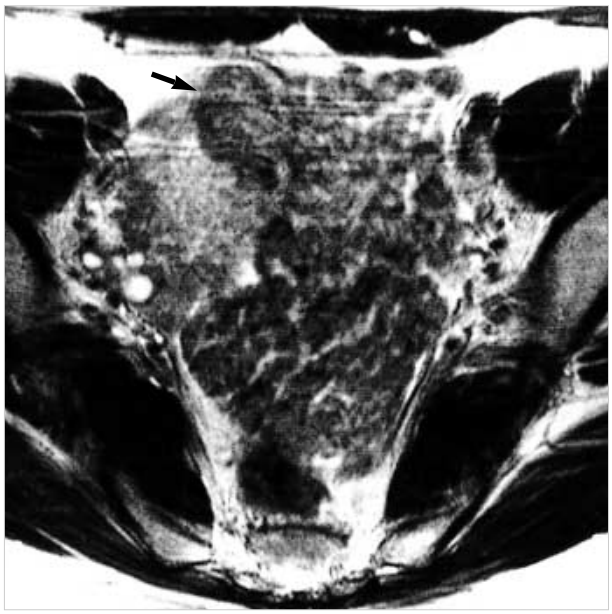

Fig. 1. Pelvic MRI shows an exophytic heterogeneous mass lesion with low signal intensity in the left pelvic cavity as well as a small homogeneous intramural leiomyoma (arrow) in the uterine fundus.

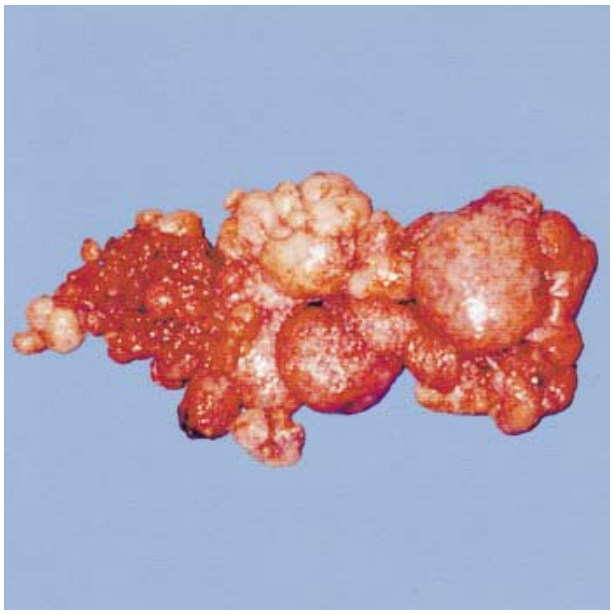

Fig. 2. Gross appearance of the resected exophytic mass shows innumerable congested, bulbous (cotyledonoid) nodules, varying from 0.5 to 5.5 cm in their greatest dimension.

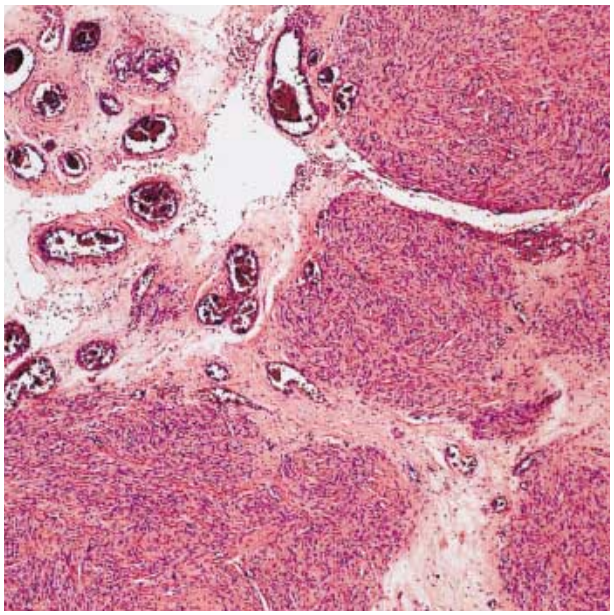

Fig. 3. The tumor is composed of nodules of smooth muscle fascicles separated by hydropic connective tissue containing congested vessels (H&E stain, ×40).

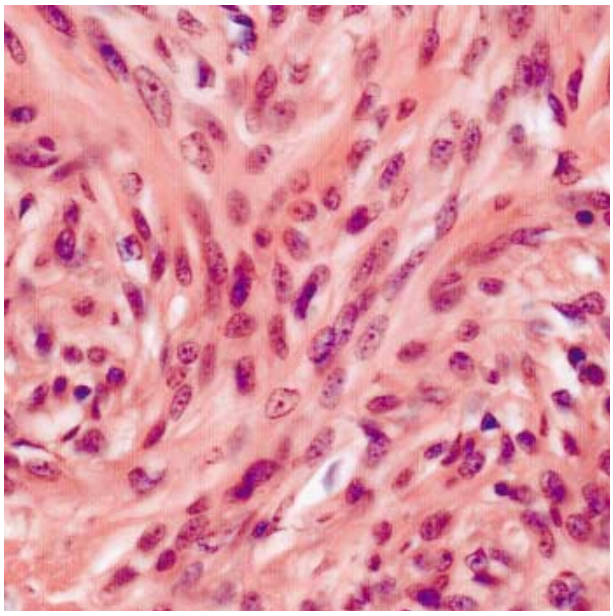

Fig. 4. The tumor cells show a swirled fascicular growth pattern with slightly enlarged nuclei. Significant cellular atypia, mitoses or necrosis are not present (H&E stain, ×200).

was observed (Fig. 4). Masson's trichrome and elastic stains revealed accentuated proliferation of muscular and non-muscular vessels in the fibrous connective tissue between muscle fascicles and its covering as well as prominent nodularity (Fig. 5). The patient was well with no recurrent disease at 2 yr after operation.

Immunohistochemically, the tumor showed a strong staining for both desmin and muscle-specific actin. Factor VIII-related antigen was positive in the endothelial cells of the blood vessels, with no evidence of intravascular growth of the tumor (Fig. 6). Vimentin was positive in intervening fibrous septa including blood vessel walls and was only focally positive in the

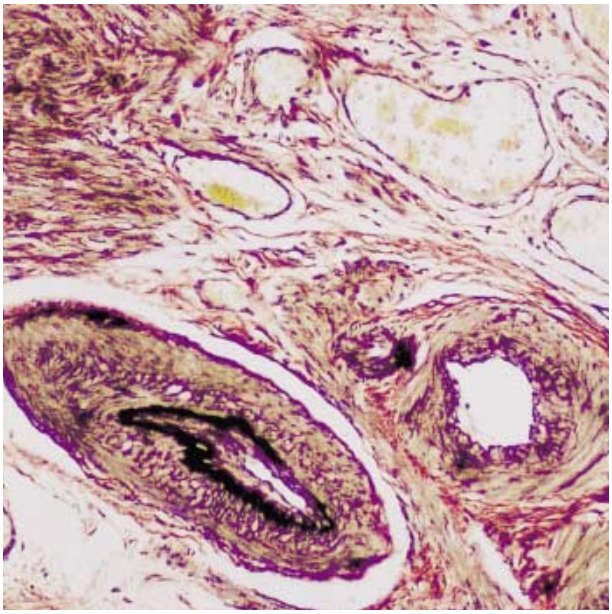

Fig. 5. Cotyledonoid processes show proliferation of both round thick-walled muscular and dilated thin-walled non-muscular vessels in the fibrous connective tissue between muscle fascicles and covering the process (elastic stain, ×100).

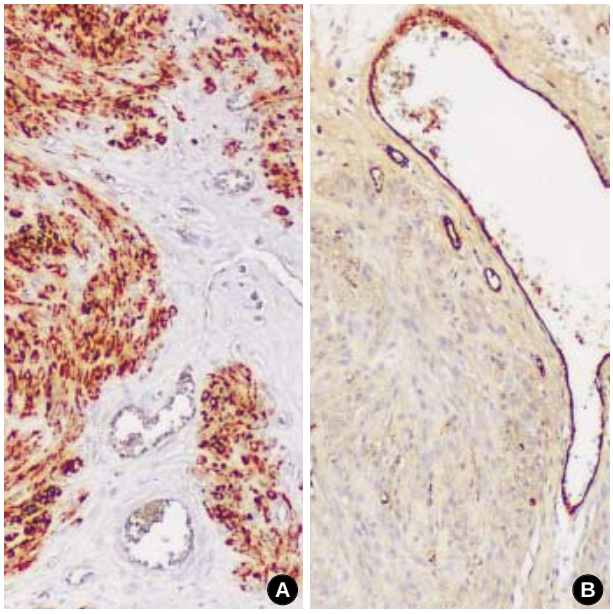


Fig. 6. Immunohistochemical stains. (A) desmin is positive in muscle fibers (×40). (B) Factor VIII shows marked proliferating vessels without intravascular growth (×40).

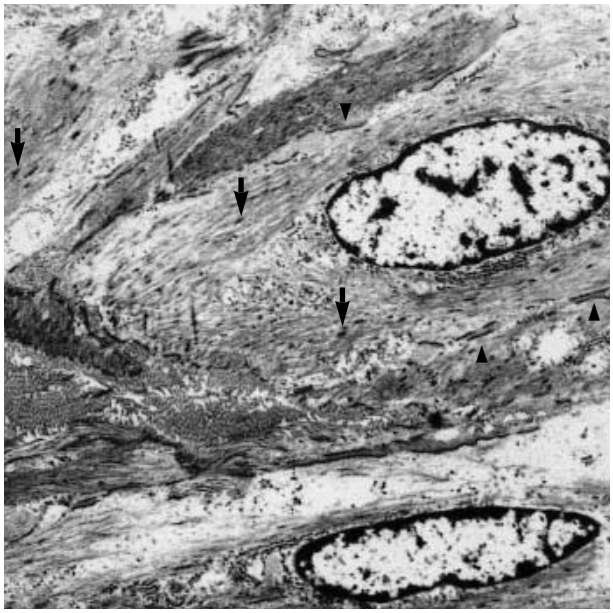

Fig. 7. Electron micrograph of the tumor cells shows elongated nuclei and cytoplasms packed with myofilaments with dense plaques (arrows). Intermediate junctions (arrowheads) on the plasma membrane are also seen (×3,000).

tumor cells. Ultrastructurally, the tumor cells had elongated nuclei and myofilaments with dense plaque in their cytoplasm that can be seen in usual leiomyoma (Fig. 7).

## DISCUSSION

Cotyledonoid dissecting leiomyoma was initially described by Roth et al. (2) in 1996. They reported four cases of an unusual form of leiomyoma, naming it "cotyledonoid dissecting leiomyoma" due to its gross similarity with placental cotyledons. It could also be called "Sternberg tumor" because their study was dedicated to the late Dr. William H Sternberg who had originally studied the tumor as "a red seaweed lesion". Recently, an additional case of cotyledonoid dissecting leiomyoma was subsequently reported by Menolascino-Bratta et al. (3), who preferred to call this tumor "angionodular dissecting leiomyoma".

In the report by Roth et al. (2), the age range of the patients was 23-41 yr. All patients had a pelvic mass or an enlarged uterus. Menolascino-Bratta's case was a 26-yr-old woman with colic abdominal pain due to acute appedicitis. In the case described here, the mass was incidentally detected on ultrasound examination in a 26-yr-old nulliparous woman. The patient reported by Menolascino-Bratta et al. (3) also had preoperative ultrasonogram, which suggested leiomyomatosis. All patients were treated by hysterectomy, except one of Roth's series who underwent partial resection of the uterus with excision of the tumor. Our patient also underwent resection of exophytic mass with consequent preservation of the uterus, according to the frozen sections that demonstrated a histologically benign smooth muscle tumor. This is the first reported case in which a frozen section was taken.

In the previous reports, the tumor size ranged 10-25 cm.

Table 1. Summary of clinico-pathologic features of cases of cotyledonoid dissecting leiomyoma

| Case | | Age (yr) | Presentation | Size (cm) | Side | Frozen section | Operation |
|---|---|---|---|---|---|---|---|
| Roth's series | Case 1 | 39 | Pelvic mass | 10, 3 | Bilateral | None | TAH-BSO |
| | Case 2 | 41 | AUB,enlarged uterus | 10 | Left | None | TVH |
| | Case 3 | 23 | AUB, pelvic mass | 25 | Left | None | Resection |
| | Case 4 | Unknown | Pelvic mass | 24 | Right | None | TAH-BSO |
| Menolascino-Bratta's case | | 26 | Abdominal pain | 16 | Right | None | TAH-BS Appendectomy |
| This case | | 26 | Incidental | 12 | Left | Done | Resection |

AUB, abnormal uterine bleeding; TAH-BSO, total abdominal hysterectomy-bilateral salpingo-oophorectomy; TVH, total vaginal hysterectomy

The tumor was on the right side in two patients, left side in two, and bilateral in one in which the smaller measured 3.2 cm in maximum dimension. In all cases, the exophytic component was in continuity with a lobulated, multinodular myometrial tumor with an irregular border. Typical intramural leiomyomas were identified in two cases. Gross findings of the present case were similar to those previously described and illustrated, although only the extrauterine component was examined.

Microscopically, Roth et al. described that the main tumor growing in dissecting pattern in the intrauterine component and cotyledonoid extrauterine extension was composed of variable sized micronodules of muscle fascicles with a marked hydropic change and rich vascularity. The case documented herein confirmed this observation, although details of the microscopic pictures of the myometrial component of the tumor remain unknown. Whereas all other cases reported had thin-walled, dilated, and congested vessels in the extrauterine nodules and round, muscular, and sometimes hyalinized vessels in the intramural portion, our case showed both round, thick-walled muscular and dilated, thin-walled non-muscular vessels in the extrauterine component, without evidence of hyalinization of the vessel walls. The vascular proliferation was more prominent in the fibrous septa than within the fascicles of smooth muscle.

Menolascino-Bratta et al. (3) noticed that immunohistochemical and ultrastructural studies were not useful for the diagnosis. Our case also showed that those ancillary techniques were not useful in evaluation of the tumor outside of the confirmation of smooth muscle nature. The whorling, disorganized myofilaments observed ultrastructurally in Menolascino-Bratta's case were not observed in the present case.

Roth's patients were followed for up to 41 yr and none had experienced recurrence, and Menolascino-Bratta's patient had been asymptomatic for 1 yr. In our case, the patient has been well without recurrent disease 2 yr after operation.

In 1997, Kiaer (4) and Honore (5) pointed out the similarity of these tumors to "grapelike" leiomyoma of the uterus (6) and suggested that these tumors including those in Roth's series, if not identical, are very closely related variants of an extremely rare form of uterine leiomyoma. To our knowledge, another case reported in 1998 (7) was similar in part to these lesions. Unlike other reports, they performed DNA flow cytometric study, which revealed diploid, benign nature.

Differential diagnosis includes intravenous leiomyomatosis, and leiomyoma with perinodular hydropic change. Although intravenous leiomyomatosis may be grossly multinodular and may involve broad ligament (8), the exophytic component is not congested and intravascular growth is characteristic. Multinodularity of the intrauterine component and hydropic changes in connective tissue are features of both cotyledonoid dissecting leiomyomas and some leiomyomas with perinodular hydropic degeneration (9), but leiomyomas with multinodular hydropic degeneration show usual pattern of leiomyoma lacking distinctive gross features seen in cotyledonoid dissecting leiomyoma.

In summary of the reported cases, the tumor seems to have a "cotyledonoid" or "grapelike" gross appearance of the extrauterine nodules and "dissecting" infiltrative growth pattern in the intrauterine component under microscope. Recently, Roth and Reed (10) described a case of 'cotyledonoid leiomyoma' without an intrauterine component. They stated that the tumor was different from the cotyledonoid dissecting leiomyoma and represented another variant of benign uterine smooth muscle tumors with unusual growth patterns. Anyway, at present, it is mandatory to unify the terminology indicating this unique variant of leiomyoma. Further accumulation of information on these tumors under the name of "cotyledonoid" dissecting leiomyoma will contribute to elucidate the nature of the tumor.

## REFERENCES


1. Clement PB. *Pure mesenchymal tumors. In: Tumor and tumor-like lesions of the uterine corpus and cervix, New York: Churchill Livingstone, 1993: 265-74.*
2. Roth LM, Reed RJ, Sternberg WH. *Cotyledonoid dissecting leiomyoma of the uterus: the Sternberg tumor. Am J Surg Pathol 1996; 20: 1455-61.*
3. Menolascino-Brrata F, Garcia de Barriola V, Navajo de Gomez M, Garcia Tamayo J, Suarez JA, Hernandez Chacon AV. *Cotyledonoid dissecting leiomyoma (Sternberg tumor): an unusual form of leiomyoma. Pathol Res Pract 1999; 195: 435-8.*
4. Kiaer H. *Is cotyledonoid dissecting leiomyoma of the uterus (Sternberg tumor) identical with grape-like leiomyoma of the uterus? [letter] Am J Surg Pathol 1997; 21: 1257-8.*

5. Honore LH. *Is cotyledonoid dissecting leiomyoma of the uterus (Sternberg tumor) identical with grape-like leiomyoma of the uterus? [letter] Am J Surg Pathol 1997; 21: 1257-8.*
6. David MP, Hommonnai TZ, Deligdish L, Lowenthal M. *Grape-like leiomyomas of the uterus. Int Surg 1975; 60: 238-9.*
7. Fukunaga M, Ushigome S. *Dissecting leiomyoma of the uterus with extrauterine extension. Histopathology 1998; 32: 160-4.*
8. Clement PB, Young RH, Scully RE. *Intravenous leiomyomatosis of the uterus. A clinicopathological analysis of 16 cases with unusual histologic features. Am J Surg Pathol 1988; 12: 932-45.*
9. Clement PB, Young RH, Scully RE. *Diffuse, perinodular and other patterns of hydropic degeneration within and adjacent to uterine leiomyomas. Problems in differential diagnosis. Am J Surg Pathol 1992; 16: 26-32.*
10. Roth LM, Reed RJ. *Cotyledonoid leiomyoma of the uterus: report of a case. Int J Gynecol Pathol 2000; 19: 272-5.*