

## **Sternberg Tumor of the Uterus (Cotyledonoid Uterine Leiomyoma): A Rare Benign Uterine Smooth Muscle Tumor with Suspicious Appearance**

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**Abstract:** Cotyledonoid leiomyoma is a rare tumor of the growth variants of the uterine leiomyoma. Researchers report a case of cotyledonoid leiomyoma in a 45 years old women presented with abnormal uterine bleeding. On laparotomy, a large multinodular, exophytic, fungating tumor mass adherent to the fundus of the uterus was seen. Microscopic examination revealed cotyledonoid leiomyoma. Researchers report it because of rarity and potential misinterpretation as a leiomyosarcoma.

**Key words:** Cotyledonoid leiomyoma, sternberg tumor, uterus, microscopical, uterine

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### **INTRODUCTION**

Cotyledonoid leiomyoma is a rare variant of uterine smooth muscle tumor with an unusual and alarming gross appearance (Saeed *et al.*, 2006). Because of the resemblance to the placenta, the term cotyledonoid leiomyoma was given for this benign variant of leiomyoma. This type of leiomyoma has been reported under grape-like leiomyoma. It is also known as Sternberg tumor (Cheuk *et al.*, 2002). Here, researchers report a rare case of cotyledonoid leiomyoma.

### **CASE REPORT**

A 45 years old female presented with abnormal uterine bleeding to the hospital. Previous menstrual cycles were irregular. Palpation of abdomen revealed a uterus size of 16 weeks pregnant with irregular surface. Blood tests showed no abnormalities. Computed Tomography (CT) showed lobulated tumor mass measuring 5 cm dimensions. There were few cystic and necrotic areas. Increased vascularity was also noted. Uterus was enlarged with multiple fibroids. Ultrasonography showed 5 cm fibroid, arising from the fundus with multiple fibroids. Laparotomy was performed. Intraoperatively there was a multinodular, exophytic, fungating tumor mass arising from the fundus of the uterus. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done (Fig. 1). There was no intrauterine component of tumor mass. The specimen was sent to the department of pathology for histopathological examination. Macroscopic the uterus was enlarged with multiple fibroids. A large exophytic, firm, multinodular, grayish white tumor mass, measuring 6 cm. Cut surface showed homogeneous, grayish white tumor mass with many irregular grape-like



Fig. 1 : The hysterectomy specimen showing the fundal fibroid on the left side with nodular surface

tan nodules attached to it. Microscopic examination of multinodular tumor mass revealed nodules of uniform smooth muscle cells, arranged in interlacing and whorling fascicles. Many blood vessels were prominent. The cellularity was increased in some areas. However, <1 mitosis per 10 high-power fields was seen. Nuclear atypia or necrosis was not seen. Perinodular hydropic change is prominent. No intrauterine or dissecting component was evident. The endometrium showed proliferative phase. After reviewing the literature and taking into consideration the gross appearance, a diagnosis of cotyledonoid leiomyoma was established.

### **DISCUSSION**

Uterine smooth muscle tumors are known to exhibit a wide variety of growth patterns. One unusual variant is

the cotyledonoid leiomyoma of the uterus or Sternberg tumor first described by Roth *et al.* (1996), Weissferdt *et al.* (2007) and Aggarwal and Arora (2011). A variety of unusual patterns can occur in uterine leiomyoma as parasitic leiomyoma, intravascular leiomyomatosis, dissecting leiomyoma, leiomyoma with perinodular hydropic degeneration (Cheuk *et al.*, 2002). No malignant behavior or recurrence has been described in these lesions with the longest follow-up period amounting to 41 years (Roth *et al.*, 1996).

The tumor grossly resembles placenta. The median age of patients at presentation is 40 years (23-65 years). Because of lack of familiarity with the alarming fungating appearance of cotyledonoid leiomyoma, the large size of the tumor and the apparent wide spread infiltrative growth with extension into the pelvic cavity and broad ligament and even into the peritoneal space, grossly mimics malignancy (Aggarwal and Arora, 2011).

Gurbuz *et al.* (2005) described a case of cotyledonoid leiomyoma which had no intrauterine portion but had extra uterine extensions. The bizarre and unusual shape was in favour of malignancy, the frozen section examination revealed a benign histology. Although, the cotyledonoid leiomyoma is a benign entity, it may suggest a malignant disease owing to its unusual sarcomatoid appearance and its rarity (Gurbuz *et al.*, 2005). Microscopic examination reveals nodules of interlacing fascicles of uniform smooth muscle cells with no atypia or necrosis. Perinodular hydropic change is prominent.

In the reviewed literature, most common presenting symptoms were abnormal bleeding and pelvic mass. The tumor can vary from 10-25 cm. Microscopically, the tumor shows fascicles of uniform smooth muscle cells with no atypia, mitotic activity or necrosis. The nodules of smooth muscle fascicles are separated by marked hydropic change and highly vascular stroma. It is believed that the poor mechanical support in the exophytic part of the tumor may lead to breakdown of hydropic stroma, exposing the neoplastic smooth muscle nodules to produce exophytic nodules. These nodules along with the stromal blood vessels give the characteristic red-brown color to the tumor (Maimoon *et al.*, 2006; Bothale *et al.*, 2013). Awareness of obstetricians and gynecologists regarding this entity will prevent unnecessary or in

appropriate treatment. Intraoperative frozen section is a mandatory and helpful procedure to avoid overtreatment of such cases (Bothale *et al.*, 2013).

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