

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

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Cotyledonoid dissecting leiomyoma (Sternberg tumour) is a rare type of smooth muscle tumor. A 38-year-old woman presented with vaginal bleeding and pelvic pain. On ultrasound multiple solid nodular lesions myomas on serosal surface and in the myometrial wall of the uterus were observed. A tumor resembling cotyledons of the placenta with a size of 15x10x5 cm was observed in the operation. Total abdominal hysterectomy and salpingectomy were performed.

The frozen section and pathology results were benign with absence of atypia and mitotic activity. Intraoperative frozen section was an important and helpful procedure to avoid overtreatment of patients in these unusual cases.

Key words: Leiomyoma, Cotyledonoid dissecting leiomyoma, Fibroid, Sternberg tumor

Gynecol Obstet Reprod Med;14:3 (205 - 207)

Introduction:

The most of the uterine smooth muscle tumors are leiomyomas. Asymptomatic fibroids may be present in 40% to 50% of women older than 35 years of age. There are uncommon types of leiomyomas. One of them is called as Sternbergas Sternberg tumor or ‘cotyledonoid dissecting type leiomyoma’; because of its appearance resembles placental cotyledons.¹ They generally arise in the subserosal myometrium of the uterus and tend to go extend into the broad ligament and pelvic cavity. The protruding component is usually red and appears congested. This way of infiltration may also been seen in intravenous leiomyomatosis.²

Here we discussed a case with cotyledonoid dissecting type leiomyoma in the lightwith contributions of the literature.

Case Report

A 38-year-old woman with gravidity 5, parity 3 and abortus 2, presented with abnormal vaginal bleeding and pelvic pain for 2 months. On examination, a large semimobile semi mobile abdominal mass, extending from the pelvis to the umbilical border, was found. On ultrasound, multiple solid nodular lesions were observed myomas on serosal surface and in the myometrial wall of uterus.were observed Both Bilateral

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Submitted for Publication: 28.06.2008

Accepted for Publication: 17.09.2008

ovaries were normal.

The hemoglobin level was 10,5 g/dl and Ca-125:2,22 (U/ml). The other laboratory parameters were normal.

Laparotomy was performed. During the operation, a multinodular red mass, protruding from the anterior part surface of the uterus to the pelvic cavity, was observed. There was no connection between this mass and with ovaries and or salpinxuterine tubes.

The tumor resembled cotyledons of the placenta with a total size of 15x10x5 cm. Except this unusual tumor appearance of tumormultiple, multiple intramural myomas leimyomas of the uterus were also observed. Bilateral ovaries were normal.

A small piece of the tumor was sent for frozen section. The pathologist reported this sample as “benign”. Benign smooth muscle was reported by pathologist. Total abdominal hysterectomy was performed andsalpengectomy because of the atipic appearance of the tubes and existence of severe adhesions between tubes and uterus salpingectomy were also performed. The patient was discharged at the 3rd day of the operation and the pathology result was dissecting type of cotyledonoid leiomyoma, including vascular proliferation, congestion and edema. neither withThere was no atypia, of cellularity nor or increasing of mitosis significant mitotic activity, but including congested vessels.

Macroscopical examination

Uterus was 17x12x7 cm in dimensions. External anterior surface of the uterus was covered with exophytic leafy projections. These uniting lesions were red - purple colored, soft and they resembled cotyledons of placenta (Figure 1). On the cut-surface of uterine wall, numerous contiguous nodular lesions

were seen. These nodules had different diameters; varying between 2mm and 2cm. Nodules were cream colored and soft. Myometrium was almost totally filled with those nodules. Overall tumor diameter was 15 cm. There was no invasion to the endometrium, uterine cervix or bilateral adnexial structures.

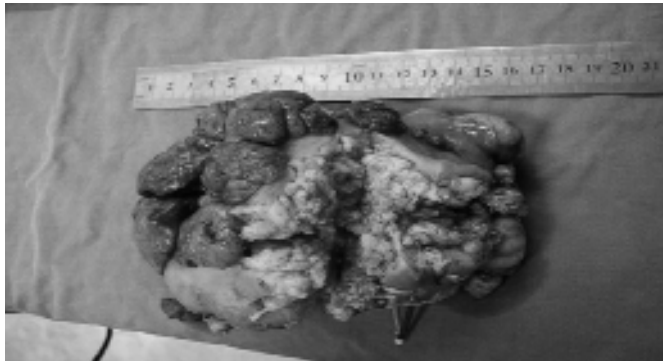


Figure 1: Macroscopic view of the exophytic bulky growth of tumor resembling cotyledons of placenta.

Microscopical examination

The tumor was composed of proliferating smooth muscle fascicles, without any significant mitotic activity. There was no significant nuclear pleomorphism, necrosis or cellularity. Neoplastic smooth muscle cells conjugate to form numerous nodules. These nodules were covered with edematous - loose stroma. Tumor was highly vascular. Muscular nature of tumoral cells was also clarified by immunohistochemical stain: Actin (Muscle specific Ab-4, Mouse Monoclonal antibody, Clone HHF35 -Neomarkers) (Figure 2). Neoplastic lesion was totally in the myometrium. There was no invasion to adjacent endometrium, cervix or adnexial structures. Due to the presence of typical smooth muscle tissue architecture without any sign of malignancy, the lesion was diagnosed as leiomyoma. Distinctive macroscopical and microscopical properties of the tumor resulted in diagnosis of a rarely seen subtype: dissecting cotyledonoid leiomyoma.

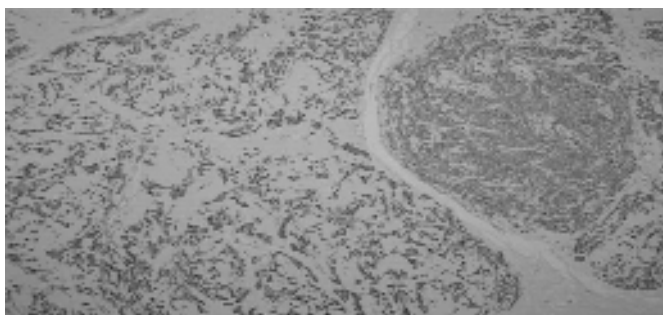


Figure 2: Actin positivity in neoplastic cells. 100x

Discussion:

In 1975 David et al first published two cases of grapelike

leiomyomas of the uterus.³ Cases of grapelike uterine tumors were collected by Dr. William Sternberg for 30 years from 1940 to 1970 so ‘Sternberg tumor’ was given as the other name of this kind of leiomyoma.⁴

Roth et al reported four cases and called them as ‘cotyledonoid dissecting’ leiomyoma of the uterus.⁴

Sternberg tumor is a rare form of the leiomyoma of the uterus. From 1996 to date, nearly 30 cases were found in the medline research. In almost all of the cases, exophytic component of the lesion was described macroscopically as cotyledonoid smooth muscle resembling placental tissue. The patients’ ages ranged from 23 to 67 years. Abnormal uterine bleeding and pelvic mass were the most common symptoms of these patients. Generally, size of the tumor was measured more than 10 cm and except for 2 patients all of them had intrauterine dissecting components. In literature, nearly 80 % of the cases were treated with total hysterectomy and one patient with subtotal hysterectomy. The other patients were treated with tumor excision.^{5,6}

In our case, the tumor diameter was 15 cm and the tumor had extrauterine cotyledonoid and intrauterine dissecting components, just like most of the other cases in the literature.

Although our patient’s age was under 40 and the frozen was benign, because of the tumor’s size, existence of the multiple intramural nodules and absence of the patient’s fertility desire, total abdominal hysterectomy was performed.

Roth, Brand and Kim et al reported that they excised only the tumor, by conserving the uterus, and they did not report any recurrence.^{2,4,7}

Jordan et al reported 3 cases of their series, which demonstrated the features of intravenous leiomyomatosis, and they regarded this tumor as cotyledonoid hydropic intravenous leiomyomatosis.⁸ But according to Norris and Pamley, ‘leiomyomatosis’ term was applied to tumors in which vascular extension was detected on gross inspection. If intravascular growth was seen microscopically, this was considered as leiomyoma.⁹

In our case we did not find the intravascular growth macroscopically, therefore we reported the case as leiomyoma.

In conclusion, a gynecological surgeon should always remember the frozen section and should earn a habit of sending the unusual tissue to frozen section during the operation. By this way, unnecessary radical surgical procedures may be prevented. Atypical uterine masses need intraoperative attention, as they can be “benign” or “malignant”, which directly changes the way of the procedure. Macroscopically cotyledonoid morphology and infiltrative pattern of the tumor may arise the suspicion of a probable malignancy. Those tumors can be differentiated from malignant counterparts by absence

of mitotic activity. But on frozen sections, there may be problems with counting mitoses. Also enlarged nuclei may seem as atypical, and may put the pathologist in doubt, preventing a certain diagnosis of leiomyoma. Preventive surgical approach may be possible when malignancy is fully excluded, but this is not practical on most of the cases, because of infiltrating pattern. For cases that cannot be differentiated from malignant neoplasm by frozen sections, the surgeon must better choose a preservative procedure. Routine Hematoxyline – eosin stains of sections obtained from paraffin blocks may be enough for diagnosis. Additional immunohistochemical stains like smooth muscle actin may be needed for some difficult cases.

Uterusun Kotiledonoid Disecting Tip Leiomyomu: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

Kotiledonoid tip leiomyom, (Sternberg tümörü) düz kas tümörünün nadir görülen bir tipidir. Otuz sekiz yaşında kadın hasta, vajinal kanama ve pelvik ağrıyla başvurdu. Ultrasonografide uterusun serozal yüzeyinde ve myometrial duvarında multipl, solid nodüler lezyonlar görüldü. Ameliyatta, yaklaşık 15x10x5 cm boyutlarında plasentanın kotiledonlarına benzeyen tümör izlendi. Total abdominal histerektomi ve salpenjektomi yapıldı. Frozen ve patoloji sonuçlarında mitotik aktivite ve atipi izlenmedi ve benign olarak geldi.

İntraoperatif frozen inceleme, bu vakada olduğu gibi alışılmışın dışında görülen vakalarda, hastaya gereksiz fazla tedavi yapmamak için önemli ve yararlı bir işlemdir.

Anahtar Kelimeler: Leiomyoma, Kotiledonoid tip disecting leiomyoma, Fibroid, Sternberg tümörü

References

1. Weissferdt A, Maheshwari MB, Downey GP, Rollason TP, Ganesan R. Cotyledonoid dissecting leiomyoma of the uterus: a case report. *Diagnostic Pathology* 2007;2:18.
2. Kim MJ, Park YK, Cho JH. Cotyledonoid dissecting leiomyoma of the uterus: a case report and review of the literature. *J Korean Med Sci* 2002;17(6):840-4.
3. David MP, Homonnai TZ, Deligdish L, Loewenthal M. Grape-like leiomyomas of the uterus. *Int Surg* 1975; 60: 238-9.
4. Roth LM, Reed RJ, Sternberg WH. Cotyledonoid dissecting leiomyoma of the uterus: the Sternberg tumor. *Am J Surg Pathol* 1996; 20: 1455-61.
5. Roth LM, Reed RJ. Cotyledonoid leiomyomas of the uterus: report of a case. *Int Jynecol Pathol* 2000;19: 272-5.
6. Saeed AS, Hanaa B, Faisal AS, Najla AM. Cotyledonoid dissecting leiomyoma of the uterus: a case report of a benign uterine tumor with sarcomalike gross appearance and review of literature. *Int J Gynecol Pathol*; 2006;25:262-7.
7. Brand AH, Scurry JP, Planner Rs, Grant PT. Grapelike leiomyoma of the uterus. *Am J Obstet Gynecol* 1995; 173: 959-61.
8. Jordan LB, Al-Nafussi A, Beattie G. Cotyledonoid hydropic intravenous leiomyomatosis : a new variant leiomyoma. *Histopathology* 2002;40:245-52.
9. Norris HJ, Parmley T. Mesenchymal tumors of the uterus.V. Intravenous leiomyomatosis. A clinical and pathologic study of 14 cases. *Cancer* 1975;36(6):2164-78.