Gynecologic Oncology Reports 11 (2015) 7–9

Contents lists available at ScienceDirect

Gynecologic Oncology Reports

journal homepage: www.elsevier.com/locate/gore

Cotyledonoid dissecting leiomyoma with adipocytic differentiation: A case report ☆

Erin A. Blake [a,*], Georgina Cheng [b], Miriam D. Post [c], Saketh Guntupalli [b]

[a] Department of Obstetrics and Gynecology, University of Colorado, Denver, CO, USA
[b] Department of Gynecologic Oncology, Department of Obstetrics and Gynecology, University of Colorado, Denver, CO, USA
[c] Department of Pathology, University of Colorado, Denver, CO, USA

ARTICLE INFO

Article history:
Received 29 August 2014
Accepted 24 October 2014
Available online 11 November 2014

Keywords:
Cotyledonoid
Dissecting
Lipoleiomyoma

Introduction

Cotyledonoid dissecting leiomyoma (CDL) is a rare benign uterine tumor initially recognized in 1975 that presents clinically and radiologically as an apparent malignant growth (Roth et al., 1996). The age distribution ranges from 23 to 73 years (Smith et al., 2012). Typical presenting symptoms are those characteristic for pelvic masses, including abdominal pain, bloating, constipation and weight gain; the most common presentation is abnormal uterine bleeding, as seen in this case (Smith et al., 2012). On ultrasound, these masses appear more lobulated and bulky than typical leiomyoma (Smith et al., 2012). CDL is a leiomyoma variant which shows a dissecting pattern, seen histologically as smooth-muscle tumors with microscopic tongues of tumoral tissue extending at least 5 mm between fascicles of the myometrium adjacent to the dominant mass (Roth et al., 1996). CDL is grossly characterized by a reddish placenta-like appearance and its tendency to expand beyond the pelvic cavity into the abdomen (Smith et al., 2012). To date, there have been fewer than 50 cases of CDL reported in the English literature based on a PubMed search. This case represents only the second reported tumor displaying adipocytic differentiation (lipoleiomyoma) (Fukunaga et al., 2010).

☆ Funding support: None.

* Corresponding author at: Department of Obstetrics and Gynecology, University of Colorado-Denver School of Medicine, Academic Office 1, Room 4010, 12631 E. 17th Avenue, Mailstop B198-1, Aurora, CO 80045, USA.
E-mail address: erin.blake@ucdenver.edu (E.A. Blake).

Case

A 56-year-old was referred to a gynecologic oncologist after an initial work-up for hematuria and postmenopausal bleeding for greater than two years revealed a 30 × 15 cm complex cystic mass in the pelvis. She had no other complaints at presentation, specifically denying abdominal pain, changes in appetite, satiety, and bowel habits. She had a medical history significant for chronic obstructive pulmonary disease but was otherwise uncomplicated. Family history was negative with the exception of an unknown type of cancer in her sister.

On physical exam, she was noted to have a normal cervix and vagina. She had a palpable pelvic mass extending from pelvis to umbilicus. Transvaginal ultrasound revealed a heterogeneous probable soft tissue mass without discernable uterus. Ovaries were not identified, and there was no free fluid in the cul-de-sac. Abdominal and pelvic computerized tomography showed a very large pelvic mass measuring at least 25 × 30 × 15 cm arising from the right adnexa and extending into the abdomen, displacing the surrounding structures. The cephalad extent of tumor was at the L2–L3 disc level. Tumor markers, including CA 125, CEA, and CA 19-9 were within normal limits.

The patient underwent an exploratory laparotomy, total abdominal hysterectomy, bilateral salpingo-oophorectomy, radical tumor debulking, resection of right and left sided retroperitoneal masses, right sided complete ureterolysis, extensive lysis of adhesions, infracolic omentectomy, and diagnostic cystoscopy. At time of the procedure, the mass was noted to be a large approximately 26 cm uterus with the extrauterine component continuous with the corpus. The extrauterine component consisted of large bilateral extensions of the mass from the uterus into the retroperitoneum and pelvic sidewall, the right side being 15 × 20 cm and the left 10 × 8 cm. Extraction of the entirety of the mass took approximately 6 h due to dense adhesions and its large nature. The mass was noted to be soft and spongy. Multiple intraoperative frozen sections were diagnosed as smooth muscle neoplasm without cytologic atypia. The mass was highly vascular, and transfusion of one unit of packed red blood cells was required during the procedure. At the end of the case, no visible tumor remained.

Tissue analysis was performed by a board certified pathologist with specialization in gynecologic pathology. On gross examination, multiple specimens containing tumor measured 84 cm in aggregate. Tumor involving the surface of the uterus had a lobulated, red appearance (Fig. 1a). Microscopically the tumor was composed of cytologically

http://dx.doi.org/10.1016/j.gore.2014.10.004
2352-5789/© 2014 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/3.0/).

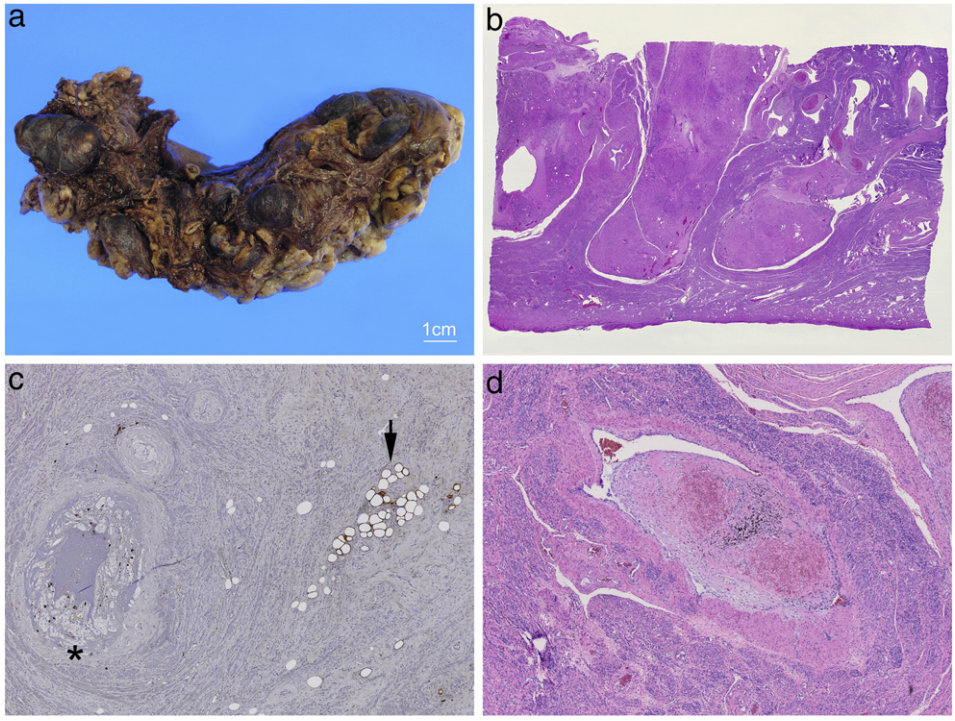

Fig. 1. a. Gross photograph of uterine serosa showing red bulbous protrusions with a "placenta-like" appearance. b. Bland fascicles of smooth muscle dissecting into the background myometrium with a pushing border in multiple areas (1× magnification, hematoxylin and eosin stain). c. Background of smooth muscle cells with foci of embedded adipose tissue (arrow) showing positive staining for S100. To the left, an occluded vessel with prominent atherosis of the wall is indicated (*) (4× magnification, S100 stain). d. Lymphovascular space invasion, as demonstrated by a nodule of smooth muscle tumor with hemorrhage sitting within a vascular space (4× magnification, hematoxylin and eosin stain).

bland interlacing fascicles of smooth muscle, which showed infiltration into the surrounding myometrium (Fig. 1b). Cytologic atypia, increased mitotic activity and tumor-cell necrosis were not seen despite extensive sampling. Immunohistochemical staining showed lesional cells to be positive for SMA and negative for HMB-45, with a Ki-67 index of <1%. Multiple areas of the tumor showed collections of cells with adipocytic differentiation, confirmed by an S100 stain (Fig. 1c). Extensive atherosis and thrombosis of intratumoral blood vessels were noted, as well as focal lymphovascular space invasion (Fig. 1d). The final diagnosis was cotyledonoid dissecting lipoleiomyoma with focal vascular invasion.

The patient was discharged from the hospital without complications 5 days following surgery and was healing well without evidence of recurrence 2 months post-surgery (Fig. 2).

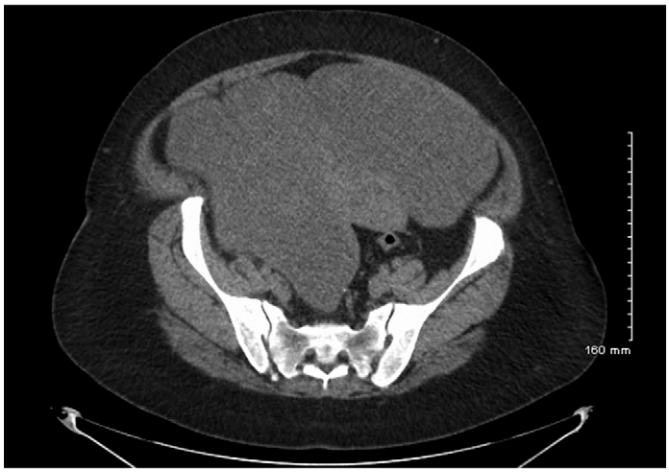

Fig. 2. Pre-operative CT scan.

Discussion

CDL presents in a manner that causes concern for malignancy based on gross and radiological appearance; however, it is a benign smooth muscle neoplasm with a growth pattern characterized by infiltrative intramural dissection within the uterine corpus and often a placental-like appearance macroscopically in its extrauterine component (Roth et al., 1996). The tumors typically present as a multinodular dissecting uterine mass with or without extrauterine extension, including to the broad ligament, adnexa and retroperitoneum. In most cases, there is some connection found between the uterine and extra-uterine disease. Due to the aggressive growth and extension of these masses, the differential diagnosis may include hydropic leiomyoma, leiomyoma with perinodular hydropic degeneration, myxoid leiomyoma, intravenous leiomyomatosis, low-grade endometrial stromal sarcoma, perivascular epithelioid cell tumors, and leiomyosarcoma (Smith et al., 2012).

Histologically, CDL is composed of benign smooth muscle. Characteristics observed in typical leiomyomas such as edema, hydropic change, focal hemorrhage, hyalinization and occasional abnormal nuclei without associated mitotic activity can also be seen in CDL, as was observed in our case. Approximately 20% of cases are reported to have an intravascular component, including vascular invasion present in the myometrial mass; however, this is not believed to change the benign clinical course (Smith et al., 2012). Over 90% of cases include a dissecting component consisting of infiltrating, sinus-like tongues extending for at least 5 mm into the surrounding tissue (Smith et al., 2012). All cases reported to date have been clinically benign despite concerning pathological prognostic factors such as lymphovascular invasion (Smith et al., 2012; Roth et al., 2013).

While the vast majority of reported cases are composed of purely smooth muscle cells, there is one reported case of a tumor displaying adipocytic differentiation (lipoleiomyoma) in the literature (Smith et al., 2012; Fukunaga et al., 2010). In addition to this finding, our case was also notable for extensive atherosis and thrombosis of intratumoral vessels. This finding has not been previously reported and is of uncertain

significance. More frequently noted in the context of the pregnant uterus, atherosis refers to lipid-filled macrophages that accumulate in the spiral arteries of the uteroplacental circulation. Atherosis in pregnancy is associated with preeclampsia and is considered to be a sign of defective vascular remodeling (Staff et al., 2013).

Although widely reported to be without potential for metastasis or recurrence, there has been one recently reported case of recurrent CDL following myomectomy (Roth et al., 2013). While this case was classified as a recurrence, incomplete resection with regrowth of persistent disease should always be taken into consideration. Literature review indicates that 24 patients were treated with total hysterectomy accompanied by unilateral or bilateral salpingo-oophorectomy, and 13 cases received a hysterectomy with ovarian preservation, all without evidence of recurrence (Smith et al., 2012; Chawla et al., 2014; Geynisman et al., 2014). Eight patients were treated with tumor resection alone with one documented recurrence (Smith et al., 2012; Roth et al., 2013; Tanaka et al., 2013). The one patient with recurrent disease underwent a subsequent total abdominal hysterectomy and right salpingo-oophorectomy without evidence of disease 2.5 years after the second surgery (Roth et al., 2013).

While CDL is a rare clinical entity, it is important that both surgeons and pathologists be aware of its characteristics in order to avoid overtreatment. In addition to the typical macroscopic appearance described above, radiologic characteristics of CDL include T2-weighted MRI images that are less heterogenous than sarcomatous lesions, and images isointense to myometrium on T1-weighted imaging. Ultrasound images, however, are not specific (Smith et al., 2012). Both macroscopic and radiological features are distinctive enough to suggest CDL in the initial evaluation or the operating room, and, despite the aggressive appearance of the tumor, a benign process should be considered in the differential when confronted with a lesion of this nature. While recurrence is exceedingly rare, treatment decisions should be informed by the fact that there has been a likely recurrence documented after conservative treatment.

This case presentation adds to the literature by documenting an extremely rare case of an aggressive appearing but benign entity. Although rare, cotyledonoid dissecting lipoleiomyoma should be in the differential when a suspected malignancy of the uterine corpus is encountered.

Conflict of interest statement

The authors declare that there is no conflict of interest to the study.

References

Chawla, I., Bhardwaj, M., Sareen, N., Khattar, N., 2014. Epithelioid cotyledonoid leiomyoma of uterus. BMJ Case Rep.

Fukunaga, M., Suzuki, K., Hiruta, N., 2010. Cotyledonoid dissecting leiomyoma of the uterus: a report of four cases. Apmis 118, 331–333 Denmark.

Geynisman, J., Pagan, C., Pirog, E., Holcomb, K., 2014. Cotyledonoid dissecting leiomyoma. Int. J. Gynaecol. Obstet. 125 (3), 284.

Roth, L.M., Reed, R.J., Sternberg, W.H., 1996. Cotyledonoid dissecting leiomyoma of the uterus. The Sternberg tumor. Am. J. Surg. Pathol. 20 (12), 1455–1461.

Roth, L.M., Kirker, J.A., Insull, M., Whittaker, J., 2013. Recurrent cotyledonoid dissecting leiomyoma of the uterus. Int. J. Gynecol. Pathol. 32 (2), 215–220.

Smith, C.C., Gold, M.A., Wile, G., Fadare, O., 2012. Cotyledonoid dissecting leiomyoma of the uterus: a review of clinical, pathological, and radiological features. Int. J. Surg. Pathol. 20 (4), 330–341.

Staff, A.C., Dechend, R., Redman, C.W., 2013. Review: preeclampsia, acute atherosis of the spiral arteries and future cardiovascular disease: two new hypotheses. Placenta 34, S73–S78 (Suppl.).

Tanaka, H., Toriyabe, K., Senda, T., Sakakura, Y., Yoshida, K., Asakura, T., et al., 2013. Cotyledonoid dissecting leiomyoma treated by laparoscopic surgery: a case report. Asian J. Endosc. Surg. 6 (2), 122–125.