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Case Report

Uterine angioleiomyoma with extra-gonadal arterial supply from the splenic artery - case report and literature review

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Summary

Angioleiomyoma (AL) is a benign tumor, composed of smooth muscle cells and thick-walled vessels. It occurs very rarely in female genital tract. The following is a case report of 45-year-old woman diagnosed with AL of the uterine with aberrant arterial supply from extra-gonadal vessels. Preoperative computed tomography examination revealed a large 30-cm tumor of the uterine, which was supplied by branches from the splenic artery. The woman was treated radically with hysterectomy and bilateral salpingo-oophorectomy. A review of the literature of AL diagnosed in the female genital tract was done.

Key words: Angioleiomyoma (AL); Benign tumor; Aberrant arterial supply from extra-gonadal vessels

Introduction

Uterine fibroid is the most common benign neoplasm of the female genital tract. It is estimated that myomas occur in about 80% of black and 70% of Caucasian women [1]. Development of fibroids depends on both (1) genetic damage - chromosomal abnormalities occur in at least 40% of myomas - and (2) overexpression of growth factors that lead to angiogenesis and fibrosis [2, 3]. Also sex hormones play an important role in the formation of fibroids [4]. Most of them are found in women in reproductive age. Fibroids rarely occur in adolescence and after menopause they tend to regress.

Angioleiomyoma (AL) is an uncommon benign tumor that originates from the vascular smooth muscle. The most common localization of AL are lower extremities, upper extremities, and head and neck [5]. It occur very rarely in the uterine - only 19 cases (18 in English literature) were presented until now.

The uterine and ovarian arteries are the main arterial supply of the female reproductive system. Extra-gonadal branches to the uterine occur in about 1,5 % of women [6]. Misdiagnosed collateral arterial flow can result in ineffective fibroid embolization or excessive bleeding during surgical procedures.

The authors present a case of angioleiomyoma with collateral arterial supply from branches of the splenic artery.

Case Report

A 45-year-old woman was admitted to 1st Department of Obstetrics and Gynecology, Medical University of Warsaw in August 2016 due to a large tumor in abdominal cavity, that probably originated from the ovary. Her medical history was: two natural childbirths, regular 30 day periods, no history of any chronic disease, and smoker for ten years.

Since January 2016, the patient observed abdominal distention, that intensified over the past three months. Two weeks before admission to the department she was in another hospital because of ascites. At the time paracentesis reveled 3,600 ml bloody fluid. Cytologic and microbiologic examination of the peritoneal fluid were negative.

Physical examination on the admission to the clinic revealed large, fixed tumor that filled the entire abdominal cavity and reached the xiphoid process. Transabdominal and transvaginal ultrasound examination detected the uterus of size 35×59 mm and large 300×480 mm solid-cystic tumor; ovaries were not seen. Computed tomography confirmed the presence of large tumor of diameter 400 mm, without infiltration to surrounding structures. No metastases in abdomen and chest were observed. The tumor was well-vascularized from the uterine arteries and extra-gonadal collaterals from spleen artery. In the right pleural cavity, a small amount of fluid was found.

Laboratory tests' outcomes (complete blood count, coagulation tests, biochemical analysis) were within normal range except for D-dimer = 7095 ng/ml and Ca125 = 202.1 U/ml.

Laparotomy revealed large tumor deriving from the fundus of the uterine, which filled the entire abdominal cavity (Figure 1). A cystic tumor of approximately 50 mm in diameter was found in the right ovary. The left ovary was normal. During surgery many extra-gonadal vessels of diameter 5-7 mm were found. They originated from the vessels supplying the greater omentum and penetrated into the tumor (Figure 2) Hysterectomy with bilateral salpingo-oophorectomy and omentectomy was performed. Moreover bilateral ilio-obturator lymphadenectomy was performed, because of enlarged lymph nodes. Blood loss during surgery was 1,500 ml. Post-operative period was complicated by fever (38.8°C), anemia (HGB 6.2 g/dl) and elevated markers of inflammation. The patient was transfused with five units of FFP and five units of RBC concentrate. Broad-spectrum antibiotic therapy has been used. The women was discharged three weeks after the operation.

Result of the histopathological examination of the uterine tumor

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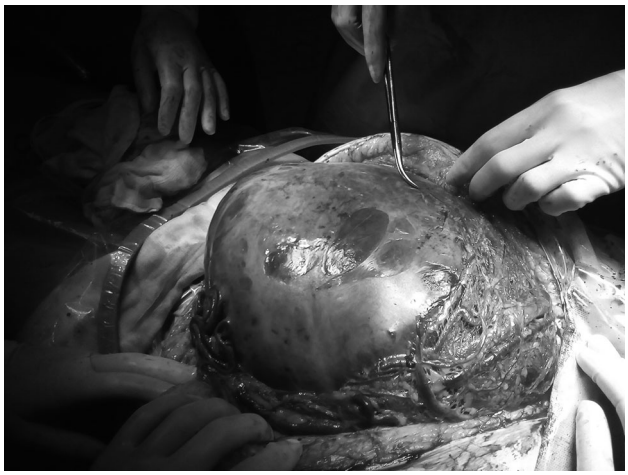

Figure 1. — Large tumor of the uterine, that filled the entire abdominal cavity. Extra-gonadal vessels of diameter 5-7 mm pass through the greater omentum and penetrate into the tumor. The vessels originate from the splenic artery.

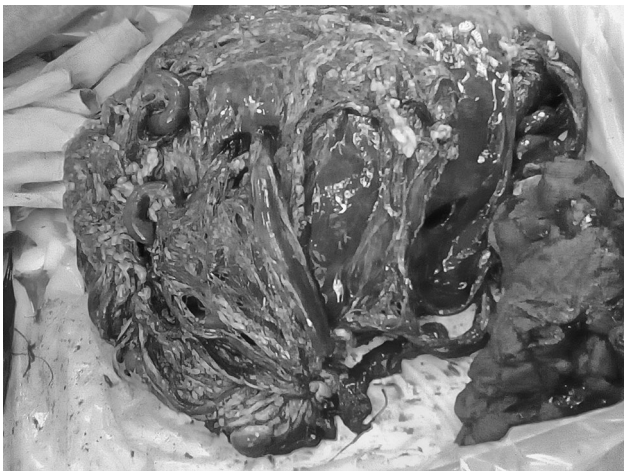

Figure 2. — En bloc resection of the uterine tumor and the greater omentum. The uterus is indistinguishably separated.

revealed leiomyoma partim angiomatosum of the uterine tumor, serosal cystadenoma was found in the tumor of the left ovary, and inflamed ilio-obturator lymph nodes as well.

Discussion

Fibroids are common tumors of the uterine, affecting 70% of women [1]. The clinical manifestation of myomas varies from asymptomatic presentation to heavy uterine bleeding leading to severe anemia [7]. The most common histopathological type is leiomyoma. Other rarer types are: epithelial, myxoid, atypical. So far only 24 cases of AL of the female reproductive system were documented: 19 in the uterine, two in the retroperitoneal cavity, two in the broad ligament, and one in the ovary.

Most ALs are large, solid tumors, with irregular shape, reaching tens of centimeters [8-11]. They can affect any part of the uterine, but mainly they derive from the fundus and corpus of the uterine. ALs are benign tumors. Clinical manifestation of AL is similar to other fibroids. Small tumors can be asymptomatic, while larger lesions can cause abdominal pain, menorrhagia, and anemia. Pedunculated ALs can mimic tumor of the ovary [12-14]. Tumors accompanied by ascites can present as pseudo-Meigs syndrome [11]. Retroperitoneal and broad ligament tumors are difficult to distinguish from ovarian tumors. Even computed tomography or magnetic resonance imaging may be insufficient [15]. In such cases only laparotomy or laparoscopy can establish localization of the tumor.

Similarly as in the present patient, Thomas *et al.* reported a case of AL accompanied by ascites and elevated levels of Ca125 - 477.1 IU/ml [11]. In other cases markers were normal [13, 14, 16, 17]. Possible reason of elevated Ca125 level in these patients was peritoneal effusion rather than AL per se.

The characteristic histological feature of AL is the presence of thick-walled vessels (capillaries predominate in leiomyomas). In most cases of AL no malignancy features in microscopic examination were found. However, case of marked cellular atypia with bizarre hyperchromatic nuclei has been described [11]. In case of doubt, extra sampling from the tumor should be performed to exclude sarcoma. Differential diagnosis should include leiomyoma, endometrial stromal nodule, angiomyofibroblastoma, and perivascular epithelioid cell tumor. Immunohistochemical assay can be helpful in establishing the diagnosis. AL presents positive expression for SMA, desmin, h-caldeson and vimentin, while expression for HMB-45 and MART-1 were negative [18].

AL is a benign tumor. None of the cases required adjuvant therapy. Large tumors were treated with hysterectomy with/without bilateral salpingo-oophorectomy, while in small lesions local excision was performed. So far, no local recurrence, intraperitoneal spread or distant metastases of AL have been documented.

Fibroids are mainly supplied with blood by the uterine artery. Ovarian artery is the second most common vessel delivering blood to myomas and occurs in about 3.8% [6, 19]. Chang *et al.* evaluated the prevalence of extra-gonadal vascularization of the uterus. They found that the most commonly collaterals originated from the inferior mesenteric artery and occurred in 1.3% of myomas. Rarely fibroids were nourished by the branches of the round ligament artery (0.2%) and interior pudendal artery (0.2%) [6]. Ripps *et al.* presented a case of fibroid supplied by branches from the transverse colon and greater omentum [20]. In the literature cases of myoma vascularization directly from the aorta, superior mesenteric and the internal iliac arteries have been described [21-23].

AL is very rare histological subtype of the uterine fibroid. Extra-gonadal vascularization of the uterine is uncommon.

To the present authors' knowledge, this is the first case described in the literature of AL with collateral blood supply from the splenic artery.

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References

[1] Baird D.D., Dunson D.B., Hill M.C., Cousins D., Schectman J.M.: "High cumulative incidence of uterine leiomyoma in black and white women: ultrasound evidence". *Am. J. Obstet. Gynecol.*, 2003, *188*, 100.

[2] Mäkinen N., Mehine M., Tolvanen J., Kaasinen E., Li Y., Lehtonen H.J., *et al.*: "MED12, the mediator complex subunit 12 gene, is mutated at high frequency in uterine leiomyomas". *Science*, 2011, *334*, 252.

[3] Mathonnet M., Descottes B., Valleix D., Labrousse F., Truffinet V., Denizot Y.: "Quantitative analysis using ELISA of vascular endothelial growth factor and basic fibroblast growth factor in human colorectal cancer, liver metastasis of colorectal cancer and hepatocellular carcinoma". *World J. Gastroenterol.*, 2006, *12*, 3782.

[4] Flake G.P., Andersen J., Dixon D.: "Etiology and pathogenesis of uterine leiomyomas: a review". *Environ. Health Perspect.*, 2003, *111*, 1037.

[5] Hachisuga T., Hashimoto H., Enjoji M.: "Angioleiomyoma: a clinicopathologic reappraisal of 562 cases". *Cancer*, 1984, *54*, 126.

[6] Chang S., Lee M.S., Kim M.D., Yoon C.J., Jung D.C., Lee M., *et al.*: "Inferior mesenteric artery collaterals to the uterus during uterine artery embolization: prevalence, risk factors, and clinical outcomes". *J. Vasc. Interv. Radiol.*, 2013, *24*, 1353.

[7] Wise L.A., Laughlin-Tommaso S.K.: "Epidemiology of uterine fibroids – from menarche to menopause". *Clin. Obstet. Gynecol.*, 2016, *59*, 2.

[8] Grigoriadis C., Androutsopoulos G., Zygouris D., Arnogiannaki N., Terzakis E.: "Uterine angioleiomyoma causing severe abnormal uterine bleeding". *Clin. Exp. Obstet. Gynecol.*, 2014, *41*, 102.

[9] Handler M., Rezai F., Fless K.G., Litinski M., Yodice P.C.: "Uterine angioleiomyoma complicated by consumptive coagulopathy". Gynecol. *Oncol. Case Rep.*, 2012, *2*, 89-91.

[10] Thomas S., Radhakrishnan L., Abraham L., Matthai A.: "Uterine Angioleiomyoma with atypia, raised CA-125 levels, and pseudo-Meigs syndrome: An alarming presentation". *Case Rep. Pathol.*, 2012, Article ID 519473.

[11] Sharma C., Sharma M., Chander B., Soni A., Soni P.K.: "Angioleiomyoma uterus in an adolescent girl: a highly unusual presentation". *J. Pediatr. Adolesc. Gynecol.*, 2014, *27*, e69.

[12] Hakverdi S., Dolapçioğlu K, Güngören A, Yaldiz M., Hakverdi A.U.: "Multiple uterine angioleiomyomas mimicking an ovarian neoplasm: a case report". *Eur. J. Gynaecol. Oncol.*, 2009, *30*, 592.

[13] Jin C.H., Yi K.W., Kim Y.S., Shin J.H., Kim T., Hur J.Y., *et. al.*: "Uterine angioleiomyoma: unusual appearance at laparoscopy". *J. Minim. Invasive Gynecol.,* 2013, *20*, 149.

[14] Singh S., Naik M., Bag N.D., Patra S.: "Angioleiomyoma of uterus masquerading as malignant ovarian tumor". *J. Midlife Health*, 2017, *8*, 145.

[15] Wee-Stekly W.W., Mueller M.D.: "Retroperitoneal tumors in the pelvis: A diagnostic challenge in gynecology". *Front. Surg.*, 2014, *1*, 49.

[16] Lee S.J., Choi Y.S., Park K.K.: "Ovarian angioleiomyoma: a case report". *Int. J. Clin. Exp. Pathol.*, 2014, *7*, 8235.

[17] Agorastos T., Dinas K., Patsiaoura K.: "Cystic degenerated angioleiomyoma mimicking ovarian pathology''. *Acta Obstet. Gynecol. Scand.*, 2001, *80*, 863.

[18] Garg G., Mohanty S.K.: "Uterine angioleiomyoma: a rare variant of uterine leiomyoma". *Arch. Pathol. Lab. Med.*, 2014, *138*, 1115.

[19] White A.M., Banovac F., Yousefi S., Slack R.S., Spies J.B.: "Uterine fibroid embolization: the utility of aortography in detecting ovarian artery collateral supply". *Radiol.*, 2007, *244*, 291.

[20] Ripps B.A, Nason W.B., Tan T.B., Mietling S.W.: "Thrombosis, leiomyoma, and GnRH-a therapy: a case report". *J. Reprod. Med.*, 1997, *42*, 124.

[21] Song C.I., McDermott M., Sclafani T., Charles H.W.: "Aberrant arterial supply to uterine fibroids from branches of the superior mesenteric artery". *Cardiovasc. Intervent. Radiol.*, 2014, *37*, 1618.

[22] Dixon S., Tapping C.R., Chuah P.S., Bratby M., Uberoi R., Anthony S.: "Successful fibroid embolization of pelvic and inferior mesenteric artery collaterals after previous uterine artery embolization". *Acta Radiol.*, 2013, *53*, 292.

[23] McLucas B.: "Extra-gonadal collateral supply to uterine leiomyomata: A case report". *Minim. Invasive Ther. Allied Technol.*, 2009, *2*, 103

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