

## Case Report

# Ovarian angioleiomyoma: a case report

Sun-Jae Lee<sup>1</sup>, Youn Seok Choi<sup>2</sup>, Kwan-Kyu Park<sup>1</sup>

<sup>1</sup>Department of Pathology, Catholic University of Daegu College of Medicine, Daegu, Republic of Korea;

<sup>2</sup>Department of Obstetrics and Gynecology, Catholic University of Daegu College of Medicine, Daegu, Republic of Korea

Received September 15, 2014; Accepted November 1, 2014; Epub October 15, 2014; Published November 1, 2014

**Abstract:** Angioleiomyoma (ALM) is a rare and painful, benign neoplasm that is referred to an uncommon type of leiomyoma originating from smooth muscle cells of arterial and venous walls. ALM is very rarely found in the female genital organs such as uterus or ovary. Herein, we present the case of a huge primary ovarian ALM in a 35 year-old woman, clinically and radiologically mimicking an ovarian fibroma, which histologically has diffuse myxoid degeneration. This is the third case of ovarian ALM and the first case of ovarian ALM with diffuse degeneration reported in English literatures, to the best of our knowledge.

**Keywords:** Angioleiomyoma, ovary, degeneration

### Introduction

Angioleiomyoma (ALM) is a rare and painful, benign neoplasm that is referred to an uncommon type of leiomyoma originating from smooth muscle cells of arterial and venous walls [1, 2]. ALM commonly occurs between the fourth and sixth decades of life [1]. It usually occurs in the superficial soft tissue of the lower extremities [3]. However, ALM is very rarely found in the female genital organs such as uterus or ovary [1, 2, 4-6]. There are only a very few cases of ALM in the ovary reported in the literature [1, 4].

Herein, we present the case of a huge primary ovarian ALM in a 35 year-old woman, clinically and radiologically mimicking an ovarian fibroma, which histologically has diffuse myxoid degeneration.

### Patient information

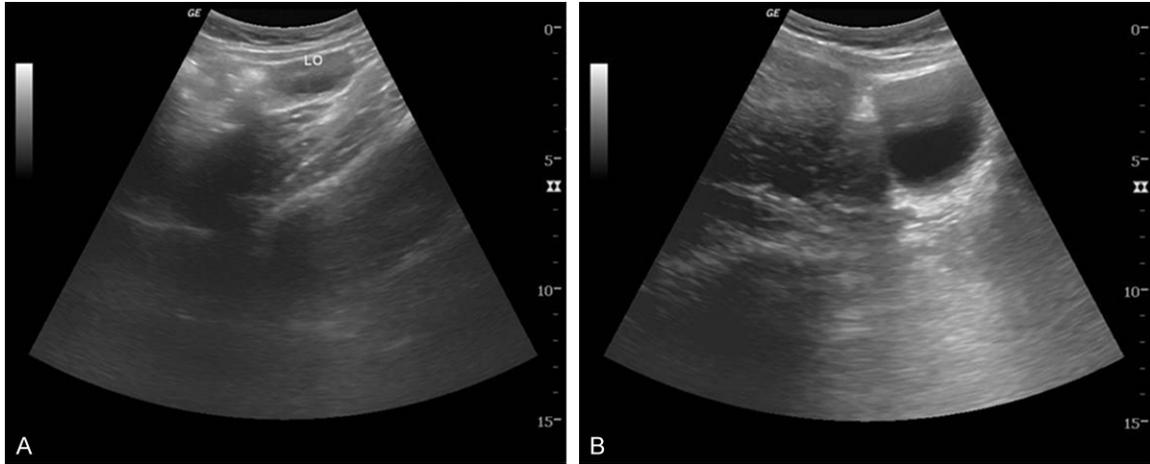
A 35-year-old female patient with gravida 0 consulted for an evaluation of a palpable pelvic mass which she had noticed for 6 months. She did not have any other complaints, except irregular menstrual cycle and mild dysmenorrhea. The patient did not take any hormonal medication.

Ultrasonographic examination revealed a huge solid mass without cystic contents, which was attached to the right adnexa and uterine serosa (**Figure 1**). The lesion was suspected to be a subserosal leiomyoma or a solid ovarian tumor. The tumor marker levels were all within normal limits ( $\alpha$ FP 2.02 ng/ml, CA19-9 17.5 U/ml, CA-125 25.03 U/ml, and  $\beta$ -hCG < 1.20 mIU/ml) in serologic examination.

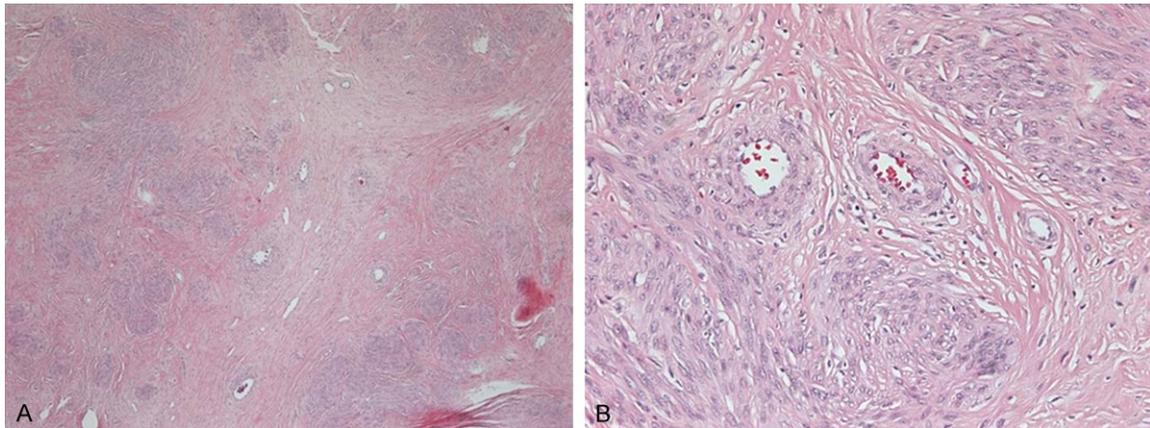
Based on the physical and radiologic findings, surgical intervention was recommended. The patient underwent a complete resection of the mass. At the time of exploratory laparotomy, there was no ascites, but a large, well-circumscribed, solid mass, arose from the rete ovary of the right adnexa, with multiple protuberates over the surface, to beneath the umbilicus. There were no adhesions present among the omentum, the sigmoid colon, and the tumor itself. The uterus and the left adnexa were grossly unremarkable. The tumor was resected, with the right salpingo-oophorectomy. A frozen section suggested a benign ovarian solid tumor such as a leiomyoma or fibrothecoma.

Macroscopic examination showed a fragmented solid mass, measuring 15 × 12 × 9 cm in aggregates. The cut surface showed multinodu-

## A case report of an ovarian angioleiomyoma



**Figure 1.** Ultrasonographic findings of the tumor. A huge solid mass without cystic contents is shown in pelvic cavity, which is attached to the right adnexa and uterine serosa (A). The observed left adnexa are unremarkable (B).



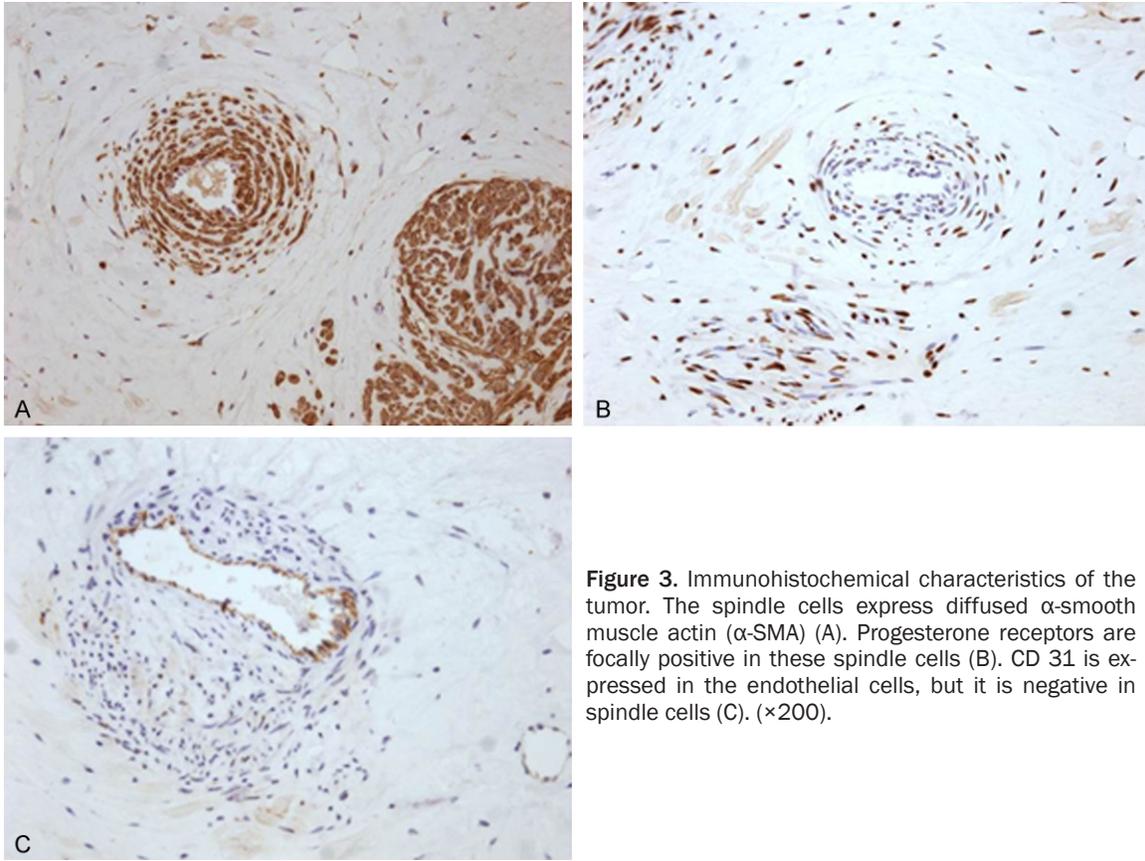
**Figure 2.** Microscopic features of the tumor with hematoxylin-eosin staining. The tumor consists of intersecting fascicles of spindle cells with eosinophilic cytoplasm and regular bland blunt nuclei, merged into numerous vascular structures with thick and proliferated walls and several dilated lumina. Diffuse myxoid and hyaline degeneration is observed in the background of fascicles, which has multinodular appearance. (A:  $\times 100$ , B:  $\times 200$ ).

lar whorling and intersecting fascicles intermitted with glistening and myxoid background.

Microscopic examination with hematoxylin-eosin staining revealed that the tumor consisted of intersecting fascicles of spindle cells with eosinophilic cytoplasm and regular bland blunt nuclei, merged into numerous vascular structures with thick proliferated walls and several dilated lumina (**Figure 2**). Scattered spindle cells with low cellularity without mitotic figures were observed to be infiltrating the vessels. Diffuse myxoid and hyaline degeneration were observed in the background of fascicles with multinodular appearance (**Figure 2A**). There was no necrosis observed. In the immunohisto-

chemical examination, the spindle cells expressed diffused  $\alpha$ -smooth muscle actin ( $\alpha$ -SMA, 1:500, Dako-patts, Copenhagen, Denmark) with negativity for CD31 (1:500, Novo, Newcastle, UK) and CD34 (RTU, Leica Microsystems, Wetzlar, Germany), which demonstrate the nature of smooth muscle in these cells (**Figure 3A** and **3C**). Estrogen (1:200, Dako-patts, Copenhagen, Denmark) and progesterone receptors (1:500, Novo, Newcastle, UK, **Figure 3B**) were focally positive in these spindle cells. Furthermore, CD 31 and CD34 marked the endothelial cells proving that the enlarged structures are truly vessels (**Figure 3C**). The histological and immunohistochemical results were consistent with the diagnosis of an

## A case report of an ovarian angioleiomyoma



**Figure 3.** Immunohistochemical characteristics of the tumor. The spindle cells express diffused  $\alpha$ -smooth muscle actin ( $\alpha$ -SMA) (A). Progesterone receptors are focally positive in these spindle cells (B). CD 31 is expressed in the endothelial cells, but it is negative in spindle cells (C). ( $\times 200$ ).

ovarian angioleiomyoma. The postoperative course was uneventful.

### Discussion

ALM is a rare benign neoplasm which arises from the vascular smooth muscle (tunica media). It has benign features and a good prognosis. It predominantly occurs in the subcutis and less often in the deep dermis of the lower extremities, presenting a painful mass in approximately 60% of the cases [1]. A comprehensive literature review showed that ALM is rarely located in the female genital tract. Only a few case reports of ALM arising from the ovary have been reported [1, 4].

Although there have been several case reports of similar tumors in uterus in the English-language literature [2, 5-7], angioleiomyoma is not recognized as a leiomyoma variant in the WHO classification and is not specifically mentioned in most of the literatures of gynecological pathology [7].

A diagnosis of ALM should be considered for a patient presenting with a hypervascular tumor

[1]. The characteristic features are bland, spindle-shaped smooth-muscle cells, numerous thick-walled arteriole-like vessels, and swirling of the smooth-muscle cells around the vessels [7]. In this case, we were able to find some characteristic features of ALM such as bland, spindle-shaped smooth-muscle cells and thick-walled arteriole-like vessels, with smooth-muscle cell proliferation around the vessels. However, the hypervascularity was not detectable in this case, because diffuse myxoid degeneration of this tumor concealed the vascular proliferation.

The main differential diagnosis is likely to be other benign uterine leiomyomas with prominent blood vessels, and the term angioleiomyoma should be reserved for those lesions with the typical features we previously described [7]. Leiomyomas that exhibit perinodular hydropic degeneration may contain prominent blood vessels, but the histological appearance of those lesions bears little similarity to the cases we described [8]. We had to rule out fibro-thecoma and cellular fibroma for differential diagnosis because of the degenerative background of the tumor.

## A case report of an ovarian angioleiomyoma

The tumor in the present case was suggestive of ovarian origin, because it replaced nearly the entire ovary and the normal ovarian tissue could not be discerned; the tube was stretched over the tumor. Primary ovarian leiomyoma must also be differentiated from a pedunculated subserosal (parasitic) uterine leiomyoma, which may have lost its original attachment and has instead become attached to the ovary, thus deriving its blood supply [9].

On histological examination, many vessels with thick and proliferated walls were found. Special stains for smooth muscle cells, such as actin and desmin, portrayed the smooth muscle bundles clearly. CD31 and CD34, on the other hand, highlighted the endothelial cells on the vessel walls. These special stains are helpful in differentiating ALM from other spindle cell neoplasms, such as fibro-thecomomas, cellular fibromas, sclerosing stromal tumors, angiofibroma, fibroma, and angiomyofibroblastoma. The lack of CD31-positive stromal cells within the tumor is also associated with the characteristic morphology [6]. These findings were similar to other previous reports of ovarian ALMs [1, 4].

The pathogenesis of ALM is not yet conclusive. Additional karyotypic changes have been reported [5]. Some studies suggest that ALM is hormone-dependent as it occurs more often in females, and the results show that ALM is immunoreactive with progesterone receptors but not the estrogen receptors [10]. These findings, however, are not consistent with those of the present case. Our case was positive for both progesterone and estrogen receptors in the immunohistochemical study.

Although it is very rare, a diagnosis of ALM should be considered for a patient presenting with a hypervascular tumor. Especially, the pre-operative diagnosis of an angioleiomyoma is rarely possible, and it usually cannot be differentiated from malignant tumors [2]. Differential diagnosis on the basis of radiological features is difficult and histopathological examination is essential [2]. In one case of Hsu et al., ultrasound had probably been the most convenient and non-invasive tool for diagnosis [1]. A strong blood flow observed in color Doppler indicated the presence of abundant vessels among the tumor, thus malignancy was our presumptive diagnosis [1]. Magnetic resonance imaging and computed tomographic scan might also be helpful for differential diagnoses [1].

Surgical excision is the treatment of choice, and it is usually the curative treatment. However, it should be performed with caution because the ALM contains many thick-walled vessels which enhance the risk of intraoperative hemorrhage.

In conclusion, while there are a few cases of ALM described in the female genital tract [2, 5-7], we underlined the importance of histological and immunological exams in the diagnosis of ALM originating from the ovary. This is the 3<sup>rd</sup> case of ovarian ALM and the 1<sup>st</sup> case of ovarian ALM with diffuse degeneration reported in English literatures, to the best of our knowledge. Pathologists should be aware of this possible diagnosis when a histological exam shows a proliferation of regular spindle cells expressing smooth muscle actin intermingled with abundant thick-walled blood vessels [4].

We believe angioleiomyoma should be recognized and categorized as a benign leiomyoma variant. Although we do not have follow-up data, the behavior of angioleiomyomas almost certainly presented to be benign, because the morphology clearly had indications of a benign lesion.

### Acknowledgements

This work was supported by the National Research Foundation of Korea Grant funded by the Korean Government [2012R1A1A4010-15639].

### Disclosure of conflict of interest

None.

**Address correspondence to:** Dr. Kwan-Kyu Park, Department of Pathology, Catholic University of Daegu College of Medicine, 3056-6 Daemyung 4-Dong, Nam-Gu, Daegu 705-718, Republic of Korea. Tel: +82-53-650-4149; Fax: +82-53-650-4834; E-mail: kkpark@cu.ac.kr

### References

- [1] Hsu TL, Changchien CC, Huang CC and Lin H. Angioleiomyoma originating from the ovary of an eleven-year-old premenarchal girl. *Gynecol Obstet Invest* 2008; 65: 262-265.
- [2] Sahu L, Tempe A and Agrawal A. Angioleiomyoma of uterus. *J Obstet Gynaecol* 2012; 32: 713-714.

## A case report of an ovarian angioleiomyoma

- [3] Ramesh P, Annapureddy SR, Khan F and Sutaria PD. Angioleiomyoma: a clinical, pathological and radiological review. *Int J Clin Pract* 2004; 58: 587-591.
- [4] Bouraoui S, El Amine El Hadj O, Rekik W, Goutallier-Ben Fadhel C, Kebir FZ, Lahmar A, Gara F and Mzabi-Regaya S. First case of angioleiomyoma originating from the ovary of an adult woman. *Gynecol Obstet Invest* 2010; 70: 8-10.
- [5] Hennig Y, Caselitz J, Stern C, Bartnitzke S and Bullerdiek J. Karyotype evolution in a case of uterine angioleiomyoma. *Cancer Genet Cytogenet* 1999; 108: 79-80.
- [6] Culhaci N, Ozkara E, Yuksel H, Ozsunar Y and Unal E. Spontaneously ruptured uterine angioleiomyoma. *Pathol Oncol Res* 2006; 12: 50-51.
- [7] McCluggage WG and Boyde A. Uterine angioleiomyomas: a report of 3 cases of a distinctive benign leiomyoma variant. *Int J Surg Pathol* 2007; 15: 262-265.
- [8] Clement PB, Young RH and 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.
- [9] Agrawal R, Kumar M, Agrawal L and Agrawal KK. A huge primary ovarian leiomyoma with degenerative changes-an unusual. *J Clin Diagn Res* 2013; 7: 1152-1154.
- [10] Marioni G, Marchese-Ragona R, Fernandez S, Bruzon J, Marino F and Staffieri A. Progesterone receptor expression in angioleiomyoma of the nasal cavity. *Acta Otolaryngol* 2002; 122: 408-412.