A case of primary ovarian adenomyoma mimicking ovarian malignancy

J.O. Kim[1], J.M. Baek[2], I.C. Jeung[3], E.K. Park[3], H.N. Lee[3], Y.S. Lee[3]

[1]Department of Pathology, [2]Department of General Surgery,
[3]Department of Obstetrics and Gynecology, The Catholic University of Korea (Korea)

Summary

Adenomyoma is a benign tumor composed of smooth muscle and benign endometrium. These tumors typically originate within the uterus. An extrauterine adenomyoma is an extremely rare entity. After an extensive literature search, only four cases of primary ovarian adenomyoma appear to have thus far been reported. Here, we report a case of ovarian adenomyoma in a 39-year-old woman mimicking malignant neoplasma of the ovary, along with a brief literature review.

Key words: Ovarian tumor; Adenomyoma.

Introduction

Smooth muscle tumors of the ovary are rare and adenomyomas presenting outside the uterus are extremely uncommon. Adenomyomas, benign tumors composed of smooth muscle and non-neoplastic endometrium, typically originate within the uterus. Thus far, only four cases of primary ovarian adenomyoma have been reported [1-4]. Owing to the extremely low prevalence of this tumor type, we have only very limited data regarding its clinical features and presentation. Recently, we encountered a case of a 39-year-old woman with unilateral ovarian adenomyoma mimicking an ovarian malignant neoplasm.

We report this case and also include a brief literature review.

Case Report

In April 2009, a 39-year-old woman with 0-0-0-0 parity visited our hospital with a chief complaint of diffuse low abdominal pain that had persisted for two weeks. At presentation, her general condition was relatively good, and no special findings were reported on the physical examination, with the exception of some abdominal tenderness. The patient's medical and family history were unremarkable. No pelvic examination was possible because the patient had not yet experienced sexual intercourse. A rectal examination was conducted and a huge palpable mass was detected in the left adnexal area. Upon abdominal computed tomography (CT), a huge left ovarian tumor measuring over 10 cm was detected; it was heterogeneous, harbored solid and cystic regions, and was interpreted as malignant. Additionally, an 8 × 7 cm sized subserosal myoma originated from the right fundal area of the uterus and a moderate quantity of ascites were noted (Figure 1). Pelvic magnetic resonance imaging (MRI) detected a 13 × 10 × 8 cm sized multiseptated large ovarian tumor of the left ovary, which was suspected to be malignant or borderline malignant (Figures 2A, 2B). No other specific findings, such as metastasis or invasion of adjacent tissue, were detected. Routine blood testing, biochemical testing, urine analysis, and electrocardiography were normal, with the exception of mild leukocytosis. The results of tumor marker analysis were as follows: CA 125 587.6 U/ml, CA 19-9 44.32 U/ml, CEA 0.739 ng/ml, CA 15-3 8.16 U/ml and β-hCG 0.554 mIU/ml. We performed gastroendoscopy and colonoscopy, and both were also negative. All of these findings were suggestive of a primary ovarian malignant neoplasm. Based on the diagnosis of ovarian cancer, the patient underwent an explo-laparotomy. Laparotomy revealed a huge ovarian tumor harboring a cystic portion with hemorrhage and a hard solid portion. The tumor mass originated from the left ovary and adhered to the left tube, colon, omentum, and adjacent tissue. The surface of the tumor was in a partially ruptured state and approximately 400 cc of a brownish turbid intrabdominal fluid was noted. Other internal

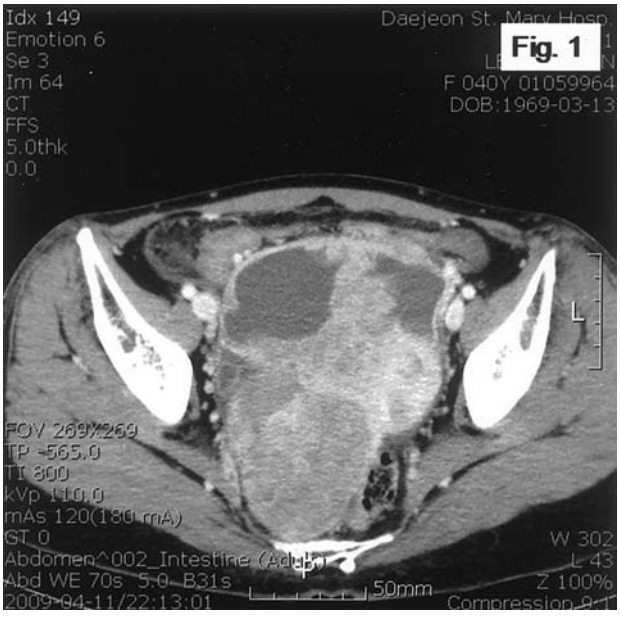

Figure 1. — CT scan imaging of the abdomen and pelvis shows a large multiseptated tumor originating from the left ovary, measuring 13 x 10 x 8 cm.

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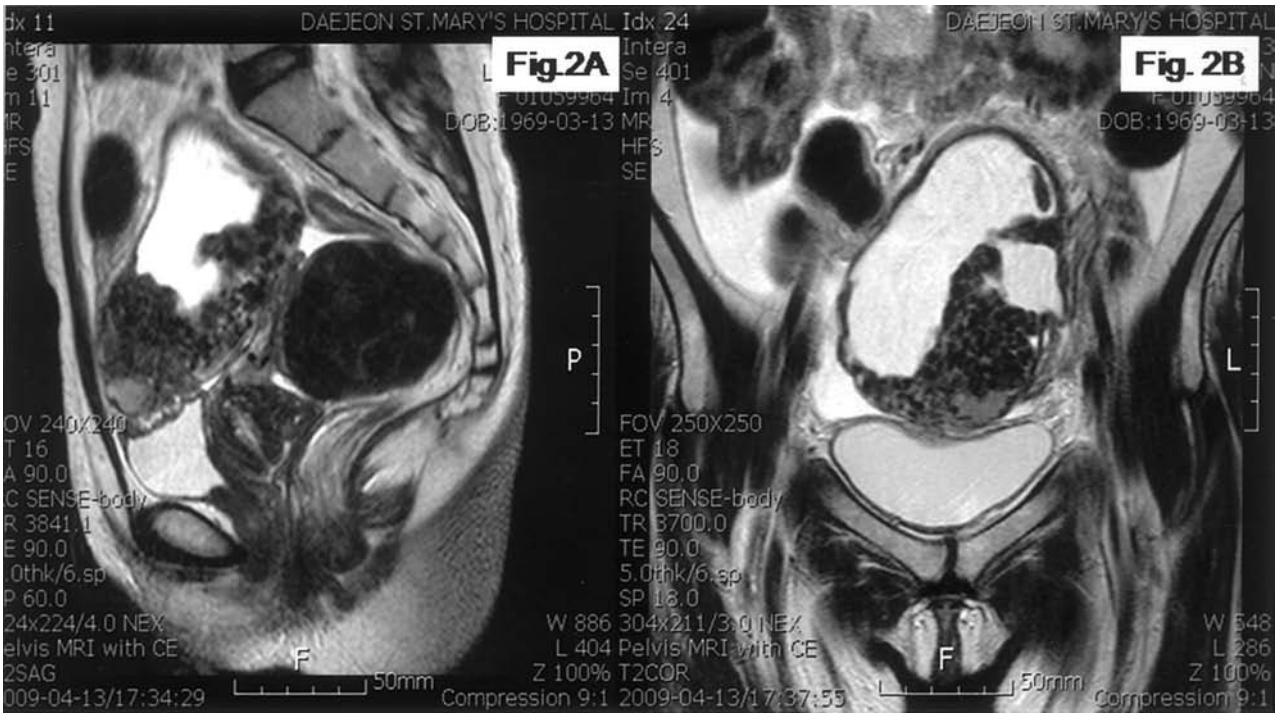

Figure 2. — MRI of the pelvis shows a show huge ovarian tumor composed of cystic and solid portions on sagittal T2WI (A) and coronal T2WI (B). The cystic portion shows high signal intensity and the solid portion shows heterogeneous low signal intensity.

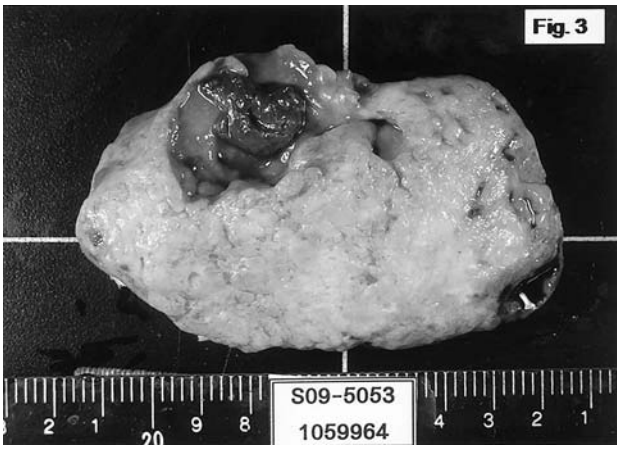

Figure 3. — The ovary evidences a whitish solid component with a whirling pattern to multifocally cystic spaces filled with serosangineous fluid and blood clots.

organs, including the right ovary and tube were grossly free, except for the known uterine myoma. Our presumptive diagnosis was ovarian malignancy, and we planned to conduct surgical staging. However, a histologic evaluation of the ovarian tumor during laparotomy resulted in its identification as a benign ovarian tumor composed of an endometrial cyst and ovarian fibroma. The surgical staging was cancelled and we closed the patient's abdomen after carrying out the myomectomy.

Grossly, the ovary measured 12.5 x 8.5 x 5.5 cm, weighed 280 g, and was firm to soft in consistency. The cut of the firm area was whitish, solid, and evidenced a whirling pattern. We noted multiple large to cystic spaces filled with serosanginous fluid and blood clots in the soft area, varying in diameter from 5 cm to 0.2 cm (Figure 3). Microscopically, the mass consisted of bundles of increased spindle cells possessing cigar-shaped nuclei with irregularly dispersed proliferative endometrial glands, stromal cells, and hemorsiderin-laden macrophages within the stroma and endometrial glandular lumen (Figures 4A, 4B). The increased smooth muscle bundles exhibited multiple hyalinization foci. The endometrial glands evidenced focal stratification, but no nuclear atypia and mitotic activity were noted. Necrosis was absent in the bundles of smooth muscle and endometrial glands. The results of immunohistochemistry for smooth muscle actin, desmin, and vimentin were positive on smooth muscle cells (Figure 6).

Based on the pathologic and immunohistochemical results, the intraoperative diagnosis of endometrial cyst and ovarian fibroma was denied, and the patient was diagnosed with primary ovarian adenomyoma. The postoperative course was uneventful. The patient was discharged from hospital on postoperative day 5 with no problems. No adjuvant treatment was administered. Tumor marker levels were checked on postoperative day 1, and were as follows: CA125 193.8 U/ml and CA19-9 16.28 U/ml. The tumor marker levels were normalized, and no abnormal findings have been noted after six weeks of follow-up.

Discussion

Adenomyomas, benign tumors composed of smooth muscle and non-neoplastic endometrium, typically originate within the uterus. An adenomyoma presenting outside the uterus is a relatively uncommon occurrence. To the best of our knowledge, only four cases of ovarian adenomyoma have been reported thus far, and each of these cases exhibited somewhat different features. The first reported primary ovarian adenomyoma was detected in an endometriotic cyst on the right ovary of a 36-year-old

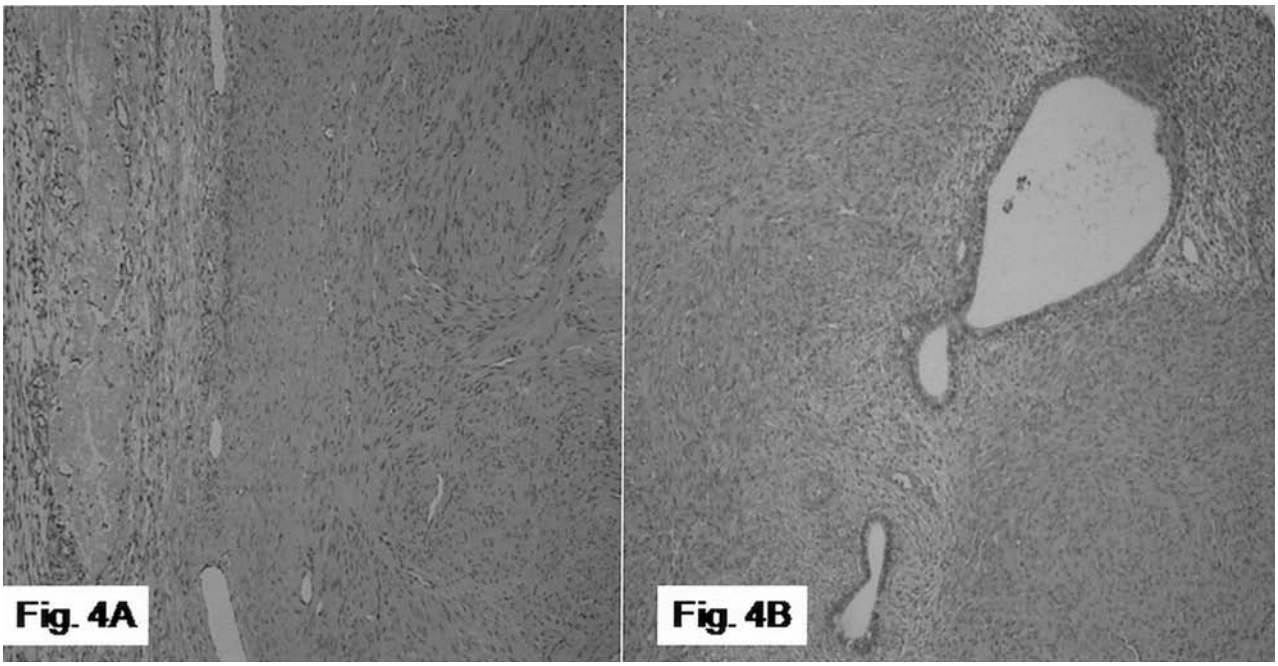

Figure 4. — The ovary evidences an increased number of smooth muscle bundles (A, H&E, 200x). Bundles of smooth muscle were increased and contain interspersed endometrial glands and stroma (B, H&E, 200x).

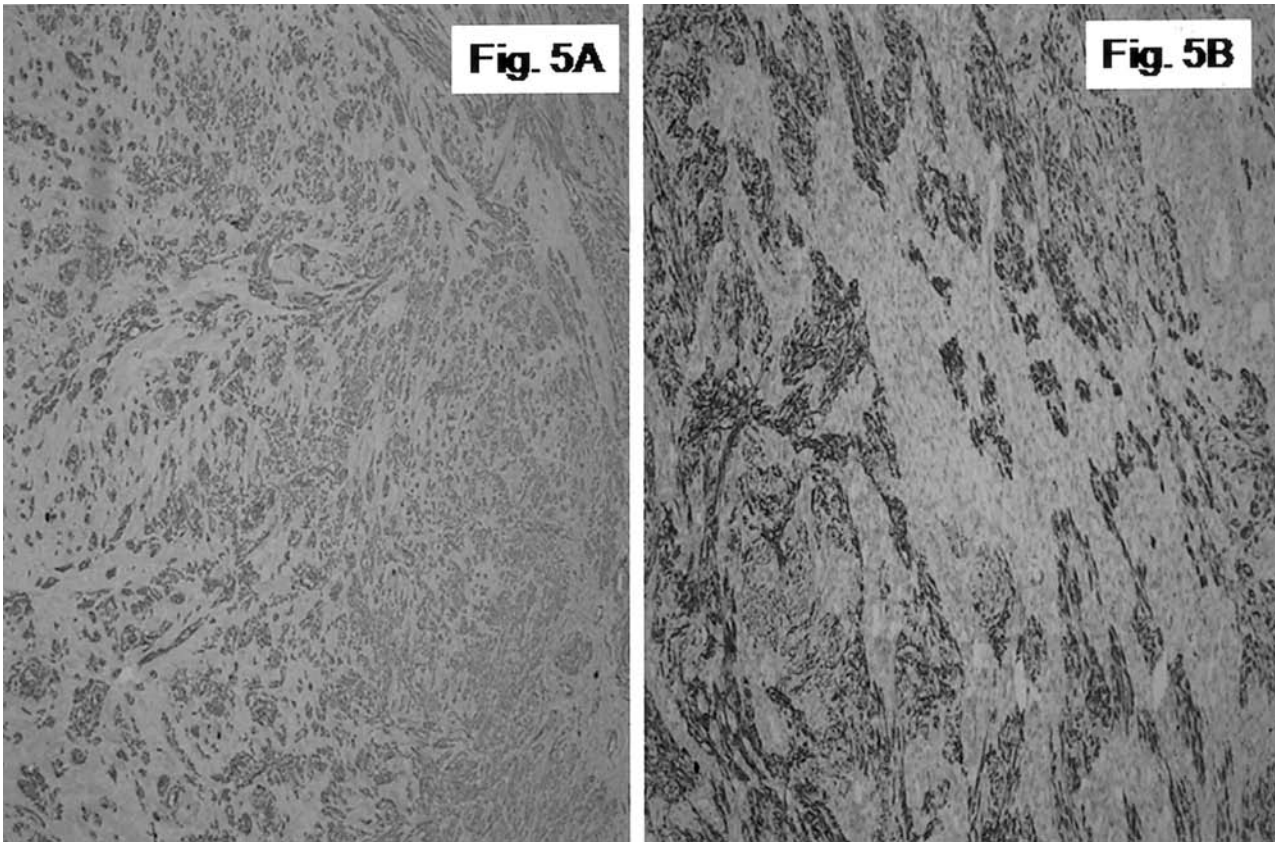

Figure 5. — Smooth muscle bundles immunohistochemically positive for smooth muscle actin (A, 200X) and desmin (B, 200x).

woman [1]. The second case was a 38-year-old infertile patient who had been treated with exogenous gonadotrophins [2]. The third case was a huge ovarian adenomyoma that mimicked an ovarian malignancy. This case was somewhat similar to our case, but the patient was a postmenopausal patient at 60 years of age [3]. The most recent case reported was that of a 45-year old woman with an endometrial polyp [4]. The nature of the reported adenomyomas varied from solid, to solid and cystic, and then to entirely cystic tumors. Microscopically, the tumors were composed of benign glands and smooth muscle cells, as well as a variety of pseudoneoplastic glandular lesions. The pathogenesis of the ovarian adenomyomas remains to be elucidated. Two theories have been advanced to explain the etiology of primary ovarian adenomyoma: the uterine or mullerian duct fusion defect theory, and the subcoelomic mesenchyme transformation theory. According to the mullerian duct fusion defect theory, the anomaly is developmental in origin, and is related to the formation of the female genital tract [5]. The alternative theory is based on the concept that the cells of the secondary mullerian system are multipotent, and may proliferate in response to estrogenic stimulation [6].

Because primary ovarian adenomyoma is such a rare condition, we currently have very little knowledge about this type of tumor. The main tumors in the differential diagnosis are fibroma and/or thecoma of the ovary and in some cases, ovarian malignancies must be excluded, as in our case [3, 7]. Upon imaging studies including transvaginal ultrasonography and pelvic MRI, the differential diagnosis between ovarian adenomyomas and the other ovarian solid tumors appears difficult [8]. Therefore, differential diagnosis still requires histopathologic examination, and immunohistochemistry can also prove useful in the establishment of a diagnosis [4, 7, 9]. The differential diagnosis of ovarian adenomyoma includes mixed tumor of ovarian fibroma and endometriosis, endometriosis, leiomyoma, and thecoma. In mixed tumors of fibroma and endometriosis of the ovary, the fibroma component consists of the spindle fibroblast-like cells arranged in a storiform pattern. By immunohistochemical testing, these cells are negative for smooth muscle actin and desmin. The endometriosis component cannot be differentiated, and harbors large to small multicystic spaces lined by endometrial glands and stroma, but can be differentiated by the presence of a prominent smooth muscle region. Leiomyoma can be excluded due to the presence of endometrial glands and stroma. Thecoma can be ruled out because it is grossly yellowish and harbors no fat-containing cells, endometrial glands, or stroma. Additionally, thecoma is negative for smooth muscle actin.

In this study, we have described the case of a 39-year-old woman with primary ovarian adenomyoma mimicking ovarian malignancy. In such cases, there is an even possibility of overtreatment if an accurate intraoperative histological test is conducted. The case described in this study demonstrates that further study will be required to identify clearly the characteristics of this rare tumor.

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Address reprint requests to:
Y.S. LEE, M.D.
Department of Obstetrics and Gynecology
The Catholic University of Korea (Korea)
e-mail: gom@catholic.ac.kr