Adenosarcoma ovarii in a 51-year-old woman: case report

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Summary

This case report presents a 51-year-old woman with an adenosarcoma of ovarian origin which is a very rare tumor. She came for consultation due to abnormal vaginal bleeding. The case also illustrates the difficulty of its correct diagnosis and discusses the possible reasons of wrong preoperative and intraoperative diagnosis.

Key words: Adenosarcoma; Ovary; Diagnosis.

Introduction

Adenosarcomas are tumors characterized by a benign neoplastic epithelial component and a malignant mesenchymal component. Malignant mixed tumors of Müllerian origin are combined mesenchymal and epithelial neoplasms which, in order of increasing malignancy, are labeled: mesodermal (Müllerian) adenosarcoma; carcinosarcoma; malignant mixed mesodermal (Müllerian) tumor [1]. Müllerian adenosarcoma is a rare neoplasm that can arise in both uterine and extrauterine locations. Extraendometrial variants (originating in the ovary, adnexa, or myometrium) are much less common, and they tend to present a more advanced stage due to their location. The sarcomatous portion of Müllerian adenosarcoma can vary from low grade to very high grade and the clinical behavior of the tumors can be indolent or aggressive [2]. Müllerian adenosarcomas of ovarian origin are a rare type of tumors. According to the description by Clement and Scully in 1974, 51 cases of ovarian origin have been described [3-5].

Case Report

The patient was a 51-year-old woman and came for consultation in March 2011 due to abnormal vaginal bleeding over a five month period. Upon physical examination and upon abdominal palpation a painless abdominal pelvic tumor of ~ 20 cm with undefined borders was identified. On vaginal palpation, a tumor was found at the fundus of the sac, which did not allow delimitation of the uterus or ovaries. Among well-recognized risk factors for adenosarcoma of the ovary, she did not use a combination of oral contraceptive pills, but on the other hand, had a first pregnancy at 23 years of age, which decreases the risk of the disease. Pelvic ultrasound examination revealed large uterine myoma, but without pathological findings of the ovaries. Laparotomy was performed and a soft, whitish-gray left ovarian tumor was identified, with 22 cm at its greatest diameter. The right ovary appeared normal, and the uterus demonstrated multiple subserous and intramural myomas and deformed shape. Peritoneal lavage was performed along with total Aldrige hysterectomy and bilateral salpingo-oophorectomy.

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Macroscopic description

The specimen obtained through surgery was an enlarged and deformed uterus with recognizable right adnexa, elongated left uterine tuba, and a large tumor on left ovary. The uterus mass was 298 grams and its dimensions 120 by x 95 by x 65 mm. The interval orifice was closed and the endocervix presented cyts. The portio and vaginal mucosa was smooth, shiny and whitish deformed by numerous intramural and subserous myoma, from isthmus to fundus with dimensions 20-55 mm. Cavum uteri was compressed, and eccentric. The endometrium was approximately three mm thick. All myomas were firm, with vortical structure, without necrosis. Right ovary was 20 x 16 x 8 mm, gyrus-like, smooth, with small rupture on the surface; uterine tube was 50 x 5 mm without pathology. Left uterine tube was elongated, with free fimbriae and were 80 x 4 mm. Tumor on the left ovary was encapsulated and had a mass of 1.5 kg and had dimensions of a 220 x 170 x 159 mm. The external surface demonstrated to be smooth and pearly-white. On cutting, subcapsular cystic spaces were appreciated and its walls were solid. The nodular zone of the tumor tissue was very firm, whitish, shiny, and without necrosis.

The processing method included the hematoxylin and eosin (H&E) staining protocol and immunohistochemistry.

Microscopic description

On the cervix, ectropion with metaplastic squamous epithelium were seen without atypia. In the endocervical wall there were numerous retention cysts. The endometrium showed signs of proliferation of stroma and epithelium, without atypia. The myomas were all moderately cellular with mitosis under 2 / 10HPF and no necrosis or hemorrhage could be seen. The right ovary was with scars, and the cellular cortex had rare inclusion cysts. There was a hemorrhagic zone, without cysts.

The left ovary included a fibroma that was slightly to moderately cellular, with high collagenisation of interstitium; mitoses were very rare, less than 1 / 10HPF. In subcapsular location there were cystic and papillary spaces, with biphasic pattern characterized centrally by medium or small glands and covered by endometrial epithelium that was not atypical and that externally covered the papillary formations. Atypical spindle cell stroma, generally low grade, proliferated around the glands, focally of intermediate grade with anisonucleosis and multinucleated cells of histiocytic aspect.

The histopathological diagnosis was adenosarcoma of the ovary, intermediate grade (FIGO Stage IA).

Fig. 1

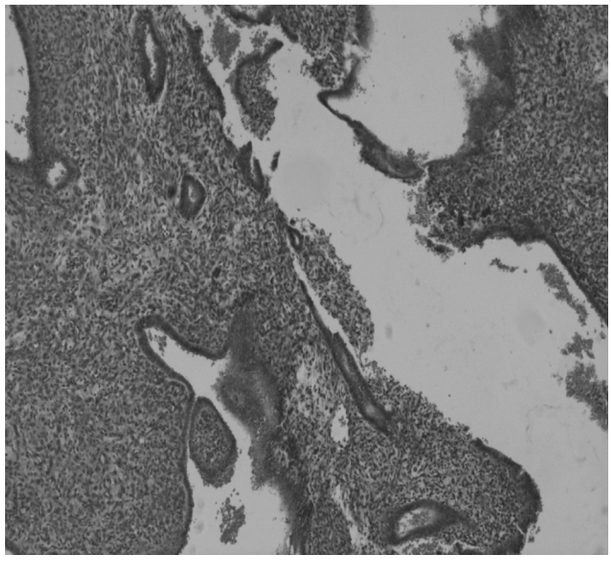

Fig. 2

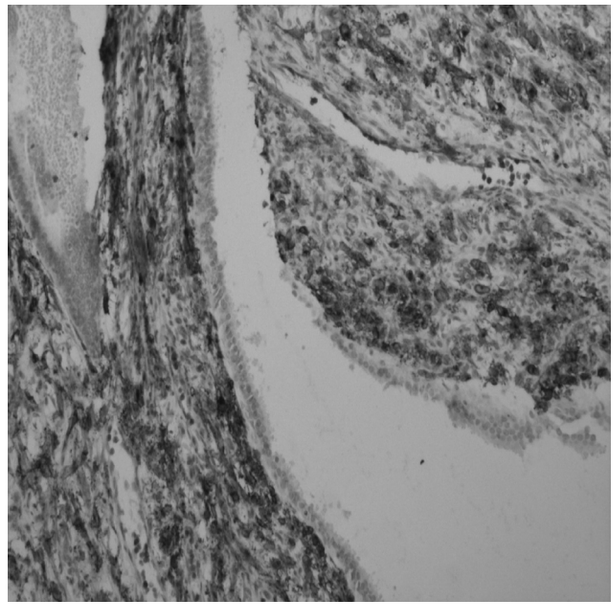

Fig. 3

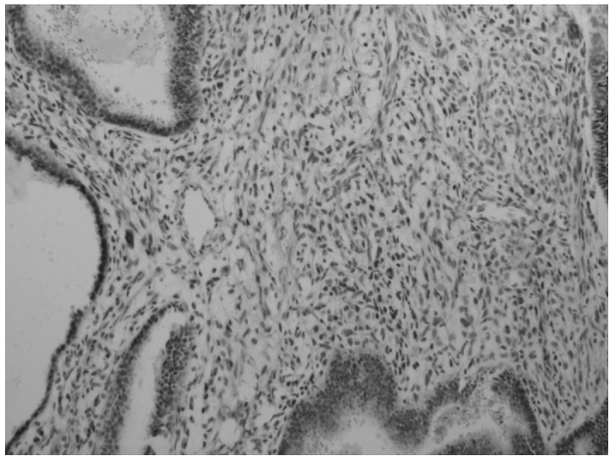

Figure 1. — Ovarian adenosarcoma with benign epithelial and malignant stromal component. H&E, x20.

Figure 2. — CD10 intracellular positivity in sarcomatous component of tumor. Immunohistochemistry, x40.

Figure 3. — Estrogen receptor (ER) positivity in benign epithelial and malignant stromal compartment of tumor. Immunohistochemistry, x400.

The described large benign tumor of the ovary, contained only peripheral / subcapsular mixed epithelial stromal tumor with malignant stromal component and the capsula was intact (Figure 1).

Through immunohistochemistry we confirmed homolog-type adenosarcoma with the presence of stromal cells of endometriod type that showed CD10 positivity (Figure 2), estrogen and progesteron receptor positivity (steroid receptors were positive in stromal and epithelial cells) but also, sarcoma cells showed smooth muscle differentiation and were desmine, smooth muscle actin and panactin positive. Ki 67 proliferative index was between 5 and 8%, with irregular pattern of postivity, with typical periglandular location, where an atypical mitosis was found.

The patient was discharged from the hospital with the diagnosis of ovarian fibroma, but the pathohistological tumor diagnosis was homologous adenocarcinoma of the left ovary. The tumor was classified as clinical Stage IA with sarcomatous overgrowth. It was decided not to treat the patient with any other additional or adjuvant therapy, by oncology advisory consilium. The patient is presently disease free six months after follow-up.

Discussion

The World Health Organization (WHO) classification of ovarian tumors includes also those classified as endometriod tumors. In these tumors, epithelial or stromal elements or both are demonstrated, and they resemble those neoplasias found more frequently in the endometrium. As with other neoplasias of the superficial epithelium of the ovary, they can be benign, borderline or malignant [6]. Included among the malignant neoplasias are adenosarcoma, mixed mesodermic tumor (Müllerian) or carcinosarcoma and stromal endometroid sarcoma. The first two could be homologous or heterogeneous. With regards to homologous tumors, the mesenchymatous structures are normal for the organ, whereas heterogeneous refers to the presence of mesodermal structures not usual for the organ (cartilage, bone, fat, striated muscle, etc.). There are few possible explanations of preoperative and intraoperative misdiagnosis in this case.

From the description by Clement and Scully of the histological pattern of uterine adenosarcoma in 1974 and the first extrauterine description by Clement and Scully in 1978 [7], multiple sites of origin for these tumors have been described [8-10], with the ovary being second in frequency, up to 18% of the cases [4]. However, to the authors' knowledge, this only represents 52 cases reported, including the present one. This is the reason why the authors did not assume this kind of outcome.

Another study referred to diagnostic imaging problems associated with the diagnosis adenosarcoma of the ovary. Adenosarcoma should be considered in patients with a predominantly solid pelvic mass on ultrasound imaging [11] like in this case, but differential diagnosis includes uterine myoma, which can resemble the adenosarcoma of the ovary; therefore, more meticulous ultrasound exam is stipulated in all cases of pelvic tumor masses.

Even macroscopic finding on laparotomy did not raise suspicion of adenosarcoma. Ovarian tumor was considered as ovarian fibroma. Macroscopically adenosarcomas of the ovary measure approximately 10 cm on average in diameter, some with smooth external surface. The present case, in contrast, was much bigger, with 22 cm in the biggest diameter, which can also be the reason of misdiagnosis. On cutting, ovarian adenosarcomas are medium brown in color with zones of necrosis and hemorrhage and with the presence of small cysts. The cut surface is spongy and multicystic with clear or yellowish-colored fluid. The largest tumors could present hemorrhagic zones. The authors did not cut the specimen during laparotomy because it could have raised suspicion of adenosarcoma, as fibromas have a different appearance. Fibromas are usually large at presentation with a mean diameter of over 10 cm and an average weight of 1 kg. They are solely or predominantly solid masses with well defined lobulated borders. Cystic degeneration, oedema and even hemorrhage occur, especially in large tumors. Calcification is present in less than 10% of cases [12].

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