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Enormous cellular ovarian fibroma with minor sex cord elements mimicking ovarian cancer: a rare case

C. Sofoudis[1], S. Balamoti[1], X. Grammatoglou[2], D. Gkolias[1], E. Papamargaritis[1], A. Gerolymatos

[1] Department of Obstetrics and Gynecology, Konstandopoulio General Hospital, Athens
[2] Department of Pathology, Konstandopoulio General Hospital, Athens (Greece)

Summary

Ovarian fibroma represents the commonest benign tumor of the ovarian stroma. The cellular fibroma is a tumor of uncertain malignant potential that may recur or be associated with peritoneal implants. Usually these are solid tumors, sometimes with small areas of cystic degeneration. The authors present a case of a 70-year-old female patient with an enormous cellular ovarian fibroma located at the region of the right adnexa. The aim of this study consists of the presentation, dissection, and proper management of this rare histologic subtype of ovarian tumor.

Key words: Ovarian fibroma; Exploratory laparotomy; Ascites.

Introduction

Cellular ovarian fibromas filled with minor sex cord elements represent a rare entity depicting a heterogenous mixed tumor of the ovary. These neoplasms are mainly solid filled with cystic components and scattered elements of sex cord origin. They occur mostly in perimenopausal and menopausal women. Rarely, they are hormonally active and may compromise the state of the endometrium. Besides this hormonal effect, they are usually asymptomatic and capable of reaching a great volume with the ultimate goal of creating gastrointestinal effects [1, 2]. The differential diagnosis remains difficult not only preoperatively – often mimicking ovarian cancer – but also, postoperatively where the distinction between different types of fibromas is yet to be defined. Another interesting aspect is their clinical longterm prognosis given that the cellular fibroma remains a tumor of uncertain malignant potential and risk of recurrence [3, 4]. Among the current literature there are few cases reported. Many studies must be conducted in order to establish proper classification and optimal therapeutic mapping.

Case Report

A 70-year-old female patient (para 2, gravida 2) without family history of gynecologic malignancy or hormonal therapy presented to our Department complaining of severe abdominal pain. The physical examination revealed presence of a large abdominal mass, mobile, palpable up to umbilicus without signs of ascites. The uterus margins were difficult to be examined. The parametrial spaces were free of malignancy. The Pap smear was normal.

Ultrasonography examination revealed a voluminous uterus of heterogenous consistency, multiple uterine adenofibromas of 30 mm diameter, an endometrial thickness of 17 mm, and a cystic tumor measuring 12×11cm localized at the right parametrial space/fossa.

Serum levels of tumor markers (CEA, Ca 125, Ca 19-9, AFP) were free of malignancy. An MRI was performed revealing a large, well-circumscribed mass mainly solid but also including a cystic component that displaced the uterus to the right, causing pressure effects to the bladder. No signs of ascites or lymphadenopathy were reported.

The patient underwent an exploratory laparotomy which revealed a large tumor of the right ovary arising from the minor pelvis, reaching the middle abdomen and causing adhesions to the mesenterium of the small bowel. The tumor was carefully detached from the mesenterium, followed by a right salpingo-oophorectomy. The frozen section report diagnosed a stromal tumor of ovarian origin. The left ovary also revealed a small cystic lesion, followed by a total hysterectomy with left salpingo-oophorectomy (Figure 1).

Assiduous exploration throughout the abdominal cavity (intestines, liver, omentum, and fallopian tubes) revealed no signs of extended lesions or malignancy. The uterus cavity was atrophic.

The surgical specimen was fixed in formalin. Twenty-seven paraffin blocks were prepared and the slides of the tumor revealed an ovarian stromal tumor fibroid origin with minor sex cord elements (less than 10% of the area of the tumor).The histologic examination revealed spindle cells resembling fibroblasts and producing collagen in a storiform pattern.The cellularity was not marked. There were no signs of atypia and mitotic activity.

Immunohistochemically, the tumor cells were positive for vimentin and negative for inhibin, calretinin, ker AE1/AE3, EMA, and CK7 (Figure 2). On the left adnexa, a serous cystadenoma of the ovary was recognized. The rest of the specimen revealed no

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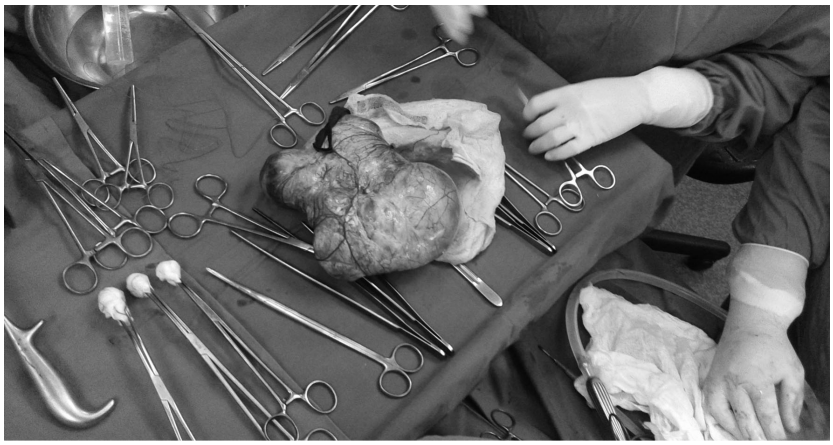

Figure 1. — Enormous cellular ovarian fibroma with minor sex cord elements.

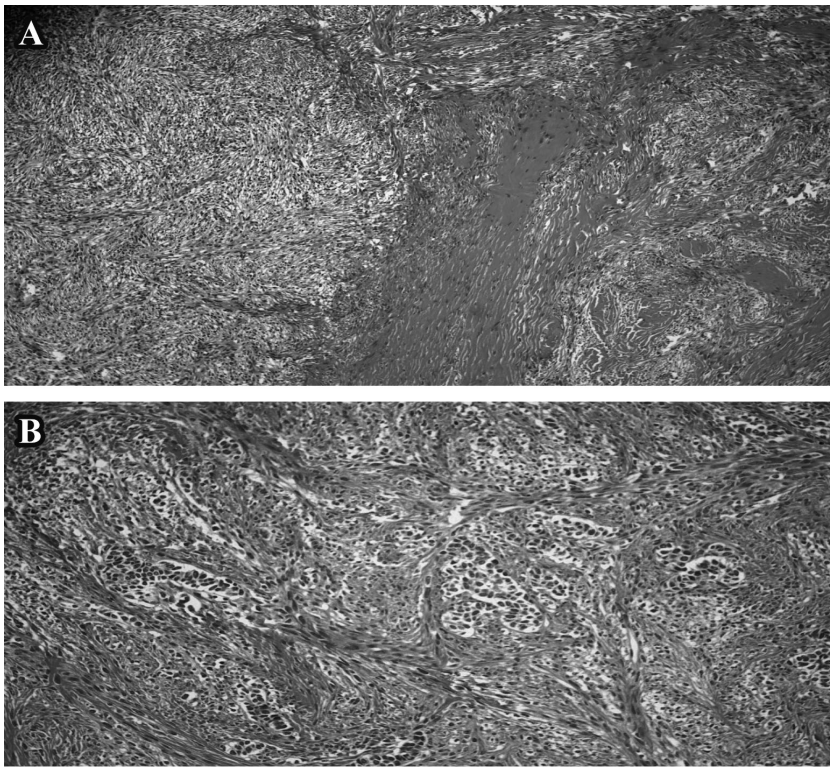

Figure 2. — (A) (H-Ex100). Spindle cells in a storiform pattern. (B) (H-Ex200). Aggregates of sex cord elements.

pathological findings.

The tumor size measured 24 ×20×9 cm depicting a smooth surface. Cut sections throughout showed a solid surface of the tumor adjusted with a 10-cm peripheral cystic part filled with a transparent serous fluid. Postoperatively, the patient remained in a good clinical condition and in the 4th pod was discharged from the hospital.

Discussion

Sex cord stromal tumors represent a heterogenous group of rare neoplasms that originate from the ovary and account for 5% to 8% of ovarian malignancies [5]. The ovarian fibroma with minor sex cord elements is part of this group with only a few case reports and small case series reported among the current literature. These tumors are usually hormonally inert and rarely easily diagnosed. By the time they reach a satisfactory amount of volume, they can create compressive effects on adjacent organs or produce amounts of free abdominal fluid mimicking ascites or even pleural effusion reproducing a Meigs' syndrome [6-8].

They are predominantly solid tumors with a co-exist-

ing cystic component. Many questions have been raised regarding their histological classification in the need of determining their malignant potential [9-11]. Recently, a new entity of the fibroma group refered to as mitotically active cellular fibroma (MACF), introduced by Irving *et al.*, includes all cellular fibrous tumors with mitotic figure more/equal to 4 MFS/10 HPFs that do not display severe nuclear atypia [12]. This diagnosis is a key to less extensive therapies, ensuring their low malignant potential [13, 14]. Based on the fact that the criteria for classification remain imperfect and there was no need for fertility sparing surgery, the authors performed in this case a total hysterectomy followed by bilateral salpingo-oophorectomy. Yang *et al*. reported a case of mucinous cystadenoma coexisting with stromal tumour with minor sex cord elements of the ovary [15].

It is well known that ovarian lesions are often misdiagnosed due to their confusing, similar characteristics. In some cases, even the frozen section fails to provide accurate information. Many studies have been conducted in order to establish the correct criteria [16]. The differential diagnosis includes ovarian epithelial cancer, germ-cell tumors, ovarian fibromatosis, Brenner tumors, pendiculated uterine fibromas, and metastatic ovarian tumors [17, 18].

Regarding the management options, surgical excision remains the gold standard. More excessive surgeries are performed in patients with signs of peritoneal dissemination/implants or a synchronous endometrial cancer. Follow up depends on the type of the surgical mapping. Regular gynecological follow up include a physical and pelvic examination, laboratory values, tumor markers, and Pap smear performance in case of preserved cervix [5, 19]. Adjuvant treatment is not recommended [9]. Further studies must be conducted in order to establish the biological behavior of these tumors.

Conclusion

Ovarian cellular fibroma with minor sex cord elements depicts a rare ovarian tumor usually misdiagnosed as ovarian epithelial cancer due to its mixed solid and cystic nature. It is originated from the sex cord stromal tumors (SCSTs) of the ovary, described as a very heterogenous group. Due to a few cases among the current literature, further evaluation is needed in order to describe its clinical course.

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Corresponding Author:
C. SOFOUDIS, M.D., Ph.D.
Ippokratous Str. 209
11472 Athens (Greece)
e-mail: chrisostomos.sofoudis@gmail.com