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Case Report

A typical case of ovarian fibrothecoma in a paucisymptomatic postmenopausal woman[☆]

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ABSTRACT

Ovarian fibrothecomas are a rare type of gonadal stromal cell tumor. They are mostly benign and unilateral in 90% of cases and mainly affect postmenopausal women. The clinical presentation and radiological features are vague. As a result, they may be misdiagnosed or confused with other entities. The definitive diagnosis is made by histology. We present a case of ovarian fibrothecoma in a 65-year-old woman with a history of thyroid neoplasia who presented only with frustrating abdominal pain with no other associated signs.

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Introduction

Ovarian fibrothecomas are ovarian tumors belonging to the group of sex cord and stromal tumors and represent the most common primary solid ovarian tumors in asymptomatic women of all ages. They account for 3% to 4% of all ovarian neoplasia [1,2]. Thus, fibroids are composed almost entirely of fibroblasts without any thecal cells, whereas fibrothecoma and thecoma are composed of different proportions of thecal cells, rich in fat and potentially estrogen-secreting [3,4].

Fibrothecomas are tumors with nonspecific clinical and imaging features. The challenge for the radiologist is to be able

to make the diagnosis quickly when faced with atypical features, with a view to rapid management, and to be able to reassure the patient, especially in the rare cases where they may be associated with ascites and/or an elevated CA125 level.

Case report

The patient was a 68-year-old woman undergoing treatment for thyroid neoplasia, who had noticed an increase in her abdominal circumference for several months without any notion of pain or abdominal heaviness; the clinical

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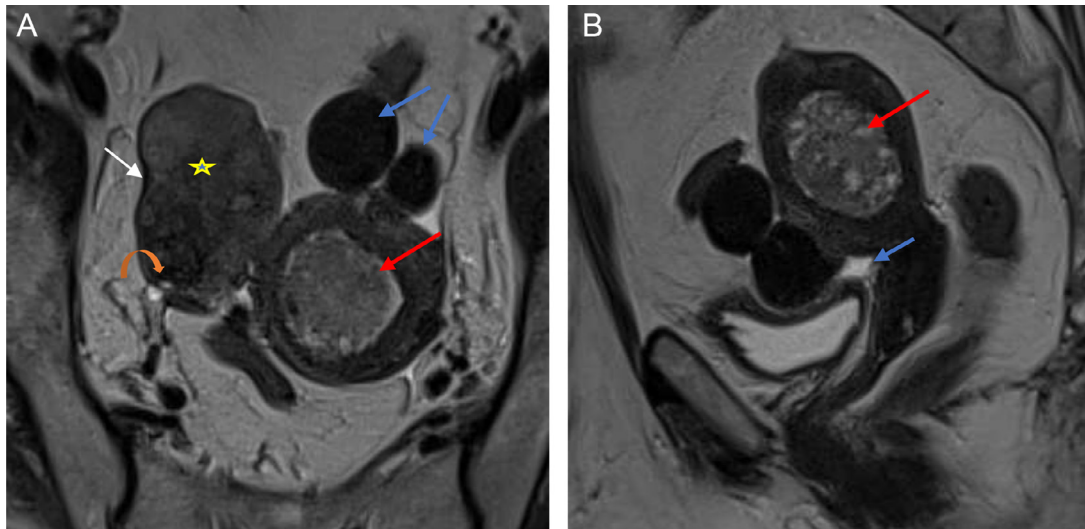


Fig. 1 – Fibrothecoma of the right ovary: Axial (A) and sagittal (B) T2-weighted MRI: lobulated solid mass with heterogeneous signal, T2 intermediate (yellow star) with a hypointense T2 pseudocapsule (white arrows) and small subcapsular cystic zone (curved arrow) associated with glandular-cystic endometrial hypertrophy (red arrow). Note: pedunculated uterine myomas and thin layer of pelvic effusion (blue arrow).

examination was unremarkable apart from the pelvic mass palpated; the pelvic ultrasound performed was in favor of endometrial hyperplasia with a latero-uterine mass. An MRI scan was performed for better characterization and revealed an endometrial thickening 32 mm thick at most, regular, heterogeneous, with a predominantly dual tissue component (T1 iso signal, intermediate T2 signal) and cystic component (clear T2 hypersignal, T1 iso signal), containing a few hemorrhagic areas in T1 hypersignal, with diffusion restriction and slightly enhanced after injection of Gadolinium (Figs. 1 and 2B). This thickening respected the junctional zone. There

was a well-limited right latero-uterine mass with lobulated contours, heterogeneous tissue signal (T1 iso signal, intermediate T2 signal), moderate diffusion restriction, moderate and heterogeneous enhancement after injection of gadolinium, measuring 6 cm in diameter (Figs. 1A and 2). The endometrial thickening was quite significant and with the restriction of endometrial diffusion, the diagnosis (benign or malignant) was not certain. We suggested granulosa tumor or ovarian fibrothecoma as possible diagnoses. Tumor markers (serum CA-125 levels) were normal. The recommended histology was only carried out after 22 months and came back in favor of

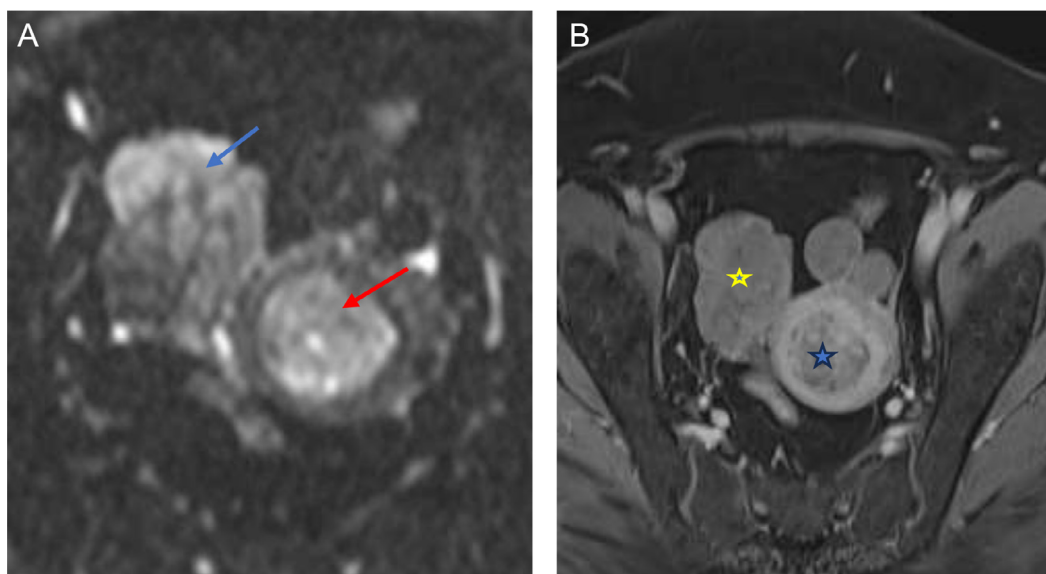


Fig. 2 – Axial diffusion sequence B1000 (A) and post gadolinium (B): diffusion hypersignal of the endometrial thickening (red arrow) and the mass (blue arrow) raising suspicion of a malignant origin but with moderate enhancement of the fibrothecoma (yellow star) and the endometrium (blue star) in relation to the myometrium.

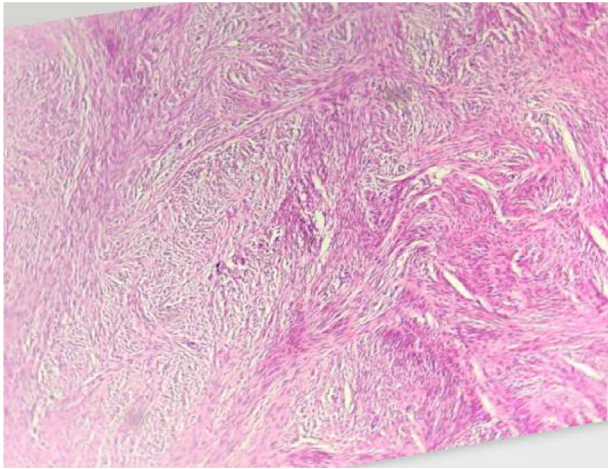


Fig. 3 – Ovarian fibrothecoma: benign tumor proliferation of the ovary, made up of a population of cells with ovoid to round nuclei and pale grey cytoplasm and a component of fusiform nuclei, with an overlap between the fibroma and the HE stroma, without cytonuclear atypia and without mitoses, 40X.

ovarian fibrothecoma. It revealed a benign tumor proliferation consisting of a population of cells with ovoid to round nuclei, pale grey cytoplasm and a component of fusiform nuclei, with overlap between the fibroma and the stroma, no cytonuclear atypia and no mitosis (Fig. 3). During this waiting period, the patient did not present any gynecological symptoms.

Discussion

Fibrothecomas are more common in peri-menopausal and menopausal women but can also occur in young people. Rare cases may be observed in patients before menarche [5]. Depending on the number of follicular membrane cells and fibroblasts, these tumors are divided histopathologically into 3 subtypes: fibromas, thecomas and fibrothecomas. The histological features of these tumors overlap, making accurate classification difficult [6]. The World Health Organisation's classification of sex cord and stromal tumors has recently been revised and these tumors have now been grouped into the following clinico-pathological entities: pure stromal tumors, pure sex cord tumors and mixed sex cord and stromal tumors [7]. Fibrothecomas differ slightly from fibroids by having sheets and nests of fleshy spindle cells with lipid-rich cytoplasm that are thecal-like cells in a background of neutral fibroid-like spindle cells [2]. Fibrothecomas differ slightly from fibroids by having sheets and nests of fleshy spindle cells with lipid-rich cytoplasm that are thecal-like cells in a background of neutral fibroid-like spindle cells [2]. The symptoms presented are not specific and the circumstances in which fibrothecal tumors of the ovary are discovered are highly variable and dominated by pelvic pain and metrorrhagia. This metrorrhagia may be related to an endocrine syndrome when the tumor is hormone-secreting. The secretion of estrogens

or androgens by the tumor leads either directly, or via peripheral conversion of androgens to estrone, to endometrial hyperplasia. In postmenopausal women, and in the absence of endocrine syndrome, metrorrhagia is most often related to endometrial atrophy [1,3]. These tumors are generally functional since most are frequently oestrogenic, although up to 10% may have androgenic activity, and present a wide spectrum of clinical features ranging from hyperandrogenic virilizing states to hyperoestrogenic manifestations [7]. They may be mistaken for a malignant mass when associated with ascites and pleural effusion in the context of Meigs syndrome with elevated serum levels of CA-125, which disappears after removal of the tumor. In bilateral forms, they may be associated with Gorlin syndrome or basal cell neuromatosis, which is much rarer [1,8]. Fibrothecomas larger than 6 cm may have a capsule and degenerative changes (peripheral subcapsular cystic areas, heterogeneous T2 signal and heterogeneous enhancement) [2] as in our case. Capsulated fibroids and fibrothecomas are larger than noncapsulated ones and can mimic malignant tumors of the ovary because they present as solid adnexal masses, sometimes associated with ascites and pleural effusions.

Clinically, the tumor appears as a solid, mobile mass with a regular surface and a wide range of sizes.

On ultrasound it is a hypoechoic solid mass with posterior attenuation; the images most often encountered are echogenic or mixed images, but anechogenic cases have also been reported [9].

MRI allows better characterization thanks to their signal and enhancement kinetics, enabling minimal surgery. It detects endometrial thickening in secretory forms, justifying the search for an adenocarcinoma, thereby reinforcing vigilance. It generally shows a hypo or iso signal on T1 and T2 weighted images in relation to the myometrium, with variable and often heterogeneous enhancement. MRI provides an excellent qualitative diagnosis as some adjacent tumors may demonstrate cystic and hemorrhagic degeneration [8]. But the final diagnosis is made by histology.

Several differential diagnoses of ovarian fibrothecoma are considered. Examples include uterine leiomyomas and various types of ovarian lesions such as dysgerminomas and granulosa cell tumors. The earlier the diagnosis is made; the sooner therapeutic intervention is performed to save patients from the adverse complications of ovarian fibrothecoma [10].

The gold standard of treatment for younger women is complete resection of the mass, while for older postmenopausal women it takes the form of bilateral salpingo-oophorectomy or radical hysterectomy in the case of tumors with multiple sites [1,3].

Conclusion

Ovarian fibrothecoma is a rare gynecological disorder. The clinical features are not specific and are sometimes poor. Often misdiagnosed, it is mistaken for other uterine pathologies such as leiomyomas due to their solid nature but can also mimic malignant pathologies. Although rare, it should be considered in patients with significant pelvic mass and post-

menopausal bleeding. Their prognosis is difficult to establish, but their behavior is almost always benign.

An MRI scan, backed up by histological analysis, is used to correct the diagnosis.

Patient consent

Informed consent was obtained from the patient included in this study to be published in this article.

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