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Case Report

Juvenile Granulosa Tumor of the Ovary: Case Report

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Abstract: Granulosa tumors are rare ovarian tumors with a better prognosis compared to other ovarian tumors. Their progression is usually slow, and the tumor remains confined to the ovary for a long time. The adult form characterized by microfollicular proliferation, a coffee bean appearance and Call Exner bodies is the most common. The solidocystic appearance of these tumors is generally nonspecific and diagnosis can be difficult preoperatively. However, the hormonal secretion that accompanies it helps guide the diagnosis and guide treatment. Surgery remains the basis of treatment and must be extensive in elderly patients. It carries a risk of recurrence which can occur several years after the excision surgery. We report the observation of a 38-year-old patient with a history of a mother suffering from breast cancer, who presented with a granulosa tumor of the left ovary diagnosed postoperatively. The epidemiological, clinical and therapeutic aspects are discussed.

Keywords: Granulosa tumors, microfollicular proliferation, prognosis.

INTRODUCTION

Granulosa tumors represent 5% of malignant ovarian tumors and are the most common sex cord and stromal tumors with an incidence of 0.58–1.6/100,000 women per year [1, 2]. The adult form (TGA) is the most common (95%) [1, 3]. Stimulated by follicle-stimulating hormone (FSH), Granulosa cells enable follicular growth and are responsible for hyperestrogenism through the production of estrogens, anti-Müllerian hormone (AMH) and inhibin B. Endometrial hyperplasia, endometrial carcinoma and breast cancer are observed in 20%, 8% and 3% of TGA cases, respectively [1]. Localized stage I forms, according to the classification of the International Federation of Obstetricians and Gynecologists (FIGO), are common and have a good prognosis [1]. But relapses, sometimes late, require prolonged monitoring [3]. The objective of the study was to report a series of 17 cases of TGA.

CLINICAL CASE

Ms. TO aged 38, originally from and residing in Bejaïa, has had four procedures, four living children and has no personal pathological history: a mother suffering from breast cancer and an aunt suffering from breast cancer. the ovary. She was admitted for heaviness-type abdominopelvic pain, dull intermittent pain predominating on the left, associated with amenorrhea of three months without other anomaly. On admission, she was in good general condition (ASA1), with well-colored conjunctival mucous membranes. The abdomen was supple with palpation combined with vaginal examination, an abdominopelvic mass extending two centimeters above the pubis, firm, regular, painless, and mobile. Abdominopelvic ultrasound described a lateral uterine mass measuring 69 x 68 cm, with mixed content and a solid predominance. In the blood, the white blood cell level was 4800/µl, the hemoglobin was 11.4g/dl and the CA125 level was 18. Laparotomy found a solid tumor developed at the expense of the left ovary, regular,

encapsulated, without capsular breakage. It was free of adhesions and measured approximately 10 cm long axis. There were also two corporeal-fundal uterine myomas of four centimeters. Chu Khellil AMRANE's gynecology team performed a left adnexectomy without capsular rupture associated with a myomectomy. The anatomo-pathological examination described the appearance of a juvenile granulosa tumor with diffuse proliferation creating small lacunae or Call-Exner bodies (figure 1). At x400 magnification, we noted monomorphic cubic cells with incised nuclei giving a "coffee bean appearance" with finely nucleolated vesicular chromatin and a lacuna or Call-Exner body in the center. The tumor was confined to the ovary without capsular rupture and was classified pT1Nx. The immediate evolution was favorable. A reoperation with additional wide excision was decided.

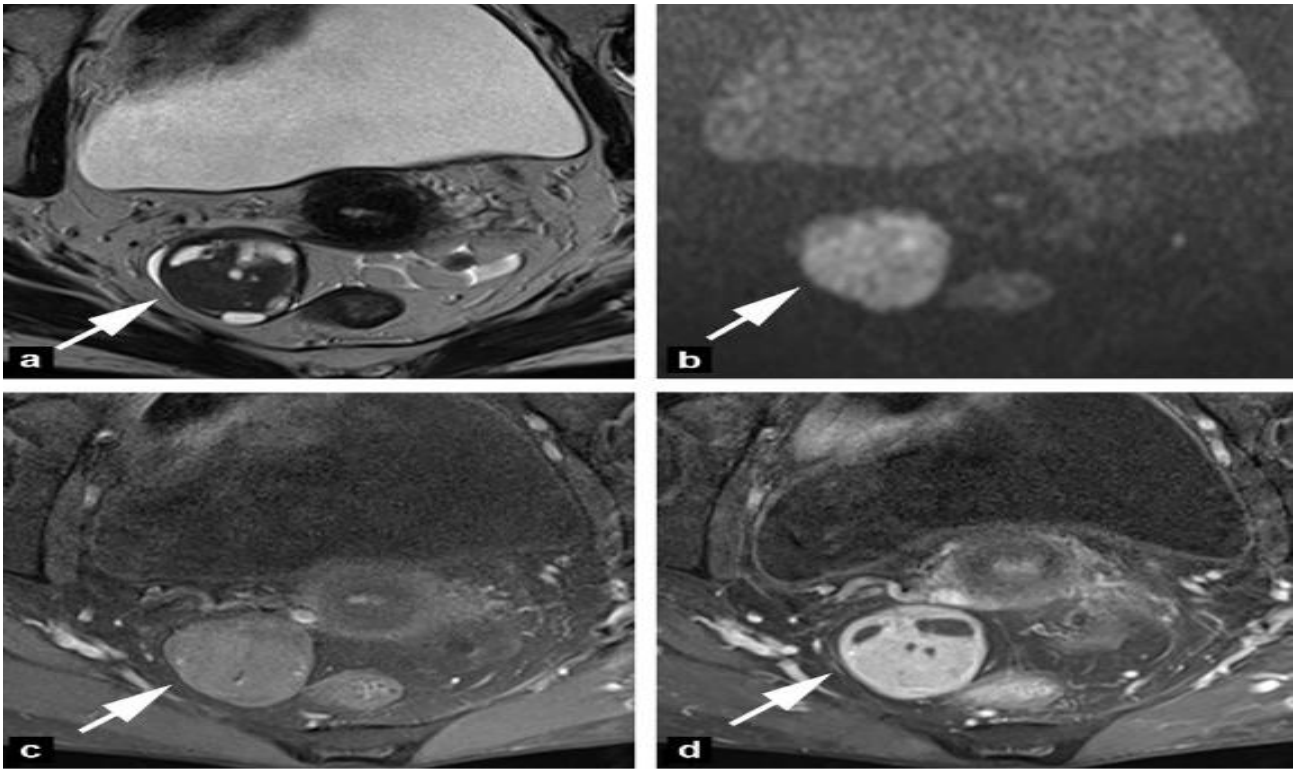

Figure 1: Granulosa tumor of the left ovary (arrow) in a 38-year-old patient (stage pT1a). The mass appears solidokystic with multiple pure fluid compartments and a tissue portion with hypersignal b1000 on the diffusion sequences, enhanced homogeneously. There is no hematic component: a: T2-weighted axial section; b: axial diffusion section b1000; c: axial T1-weighted image with fat suppression; d: axial T1-weighted image with fat suppression and gadolinium injection

DISCUSSION

GCTs are the most common stromal and sex cord tumors of variable malignancy, but they remain rare. They are classified into "adult" and "juvenile" types by their difference in clinical and histopathological presentation, but their macroscopic (and therefore radiological) appearance is generally similar [7]. Many of them (nearly 95%) are of the "adult" type, which can occur at any age with a peak incidence between 49 and 55 years [7, 8]. They are slow growing with a tumor often limited to a single ovary at diagnosis. Long-term monitoring is, however, necessary due to their potential for recurrence and late recurrences (up to 37 years after diagnosis [9]). Their pathogenesis is, to date, unknown. These tumors develop from granulosa cells which form the inner layer of the "wall" of the follicles and which, under the effect of FSH, will secrete both estrogens by aromatization of androgens and inhibin, which will exert a negative feedback on the production of FSH. This perfectly explains the symptoms of hyperestrogenism or, more rarely, virilization (2.5% of GCTs) [10] classically associated with these tumors. Our patient did not present clinical signs of hyperestrogenism. This absence is not enough to rule out the diagnosis of GCT in the presence of an adnexal mass because signs of hyperestrogenism are only encountered in less than two out of three cases [3]. Imaging has become an essential element for detection, characterization and extension of adnexal masses. Several studies [11, 12] have now proven the superiority of MRI compared to CT and Doppler ultrasound for characterizing these masses. In the literature, few studies report the MRI semiology of GCTs [4, 5] with populations not exceeding seven patients, accompanied by a few clinical cases [6, 9, 13, 14]. Preoperative diagnosis is difficult; it was only made prospectively in one case out of the three patients presented. Indeed, their appearance in imaging is not unambiguous, on the one hand, due to their excessively

variable macroscopic presentation, and on the other hand, their rarity. However, the signs considered suggestive are either a mainly multi-compartmental cystic mass with tissue portions of variable size, or a mixed solidocyst mass [4]. These aspects are those encountered in our three patients. Two out of three patients presented a solidocystic form in which the solid portion was predominant, in intermediate T2 signal and with very variable enhancement since case no. 1 presented an enhancement similar to that of the myometrium while case no. 3 did not. enhanced only very slightly. Kim also noted that the enhancement of the solid portion was variable but more frequently identical to or greater than that of the myometrium [4]. Low enhancement poses the problem of differential diagnosis with tumors of the fibrothecal group (fibroma, fibrothecoma) but these generally have a much lower T2 signal (at least equal to that of the adjacent pelvic muscles [15]). than in TCGs. Furthermore, the cystic component in GCT is formed of small fluid pockets scattered in the lesion, whereas in remodeled fibrothecomas (cystic, edematous or myxoid degeneration), the T2 hypersignal component is often focal, central or eccentric [16], "blurred" contours or with fluid components of variable size. The multilocular cystic form encountered in our patient has small compartments, some with hematic content, with septa of variable thickness and a focal tissue component. In the literature, this shape is described as a "sponge-like" appearance linked to the large number of small fluid pockets and rarely encountered in other ovarian tumors [5]. Concerning the hemorrhagic component, this was found in five of the seven tumors studied by MRI in Kim's study [4], roughly corresponding to the data of Morika *et al.*, [5], where a hemorrhagic component was found in three of the five patients. This hemorrhagic component in MRI is classic but not specific for GCT and was only found in one of our patients. It can also be found in other adnexal masses such as endometriomas, functional hemorrhagic cysts, but also certain malignant epithelial tumors such as endometrioid carcinomas [4]. Unlike multilocular GCTs, endometriomas and functional hemorrhagic cysts generally do not have a focal solid portion. Malignant epithelial tumors contain vegetations that GCTs never have and are more readily associated at the time of diagnosis with ascites and/or peritoneal implants. Other rarer aspects of GCT are reported, such as the pure unilocular cystic form with a more or less thick and enhanced wall [8]. We have seen that virilization syndrome is rather rare in GCTs but when it is present, then these tumors tend to be cystic [7]. Finally, totally solid GCTs would be exceptional [4, 8], then indistinguishable on imaging from other solid tumors. Treatment is essentially based on surgical excision. The attitude is similar to epithelial tumors and the excision must be wide including a total hysterectomy and a bilateral adnexectomy in elderly or peri-menopausal patients. However, conservative surgery can be considered in young patients. A uterine biopsy and curettage must be associated with it. Chemotherapy based on platinum salts is recommended in advanced forms. The prognosis of granulosa tumors is uncertain and strongly depends on the tumor stage. The tumor remains confined to the ovary for a long time and 78 to 91% of granulosa tumors are stage I [4, 7, 14]. In 1981, Bjorkholm already estimated 5-year survival at 95% in stages I, 55% in stages II and 25% in advanced forms [7]. The same prognostic results are noted in most publications. The 10-year survival of stages I, II, III/IV is respectively 84 to 95%, 50 to 65%, 17 to 33% [1]. Late recurrences can be observed after a free interval of more than 20 years. Hines reported a recurrence 37 years after the initial surgery [15]. The frequency of recurrences essentially depends on the stage of the tumor and the surgical excision. It is 9% in stages I and 30% in advanced stages [16]. These are often locoregional recurrences which remain indications for surgical excision in the event of extricability.

CONCLUSION

Granulosa tumors are rare ovarian tumors with a better prognosis compared to other ovarian tumors. Their progression is usually slow, and the tumor remains confined to the ovary for a long time. The adult form characterized by microfollicular proliferation, a coffee bean appearance and Call Exner bodies is the most common. The solidocystic appearance of these tumors is generally nonspecific and diagnosis can be difficult preoperatively. However, the hormonal secretion that accompanies it helps guide the diagnosis and guide treatment. Surgery remains the basis of treatment and must be extensive in elderly patients. It carries a risk of recurrence which can occur several years after the excision surgery. We report the observation of a 38-year-old patient without known risk factors, who presented with a granulosa tumor of the left ovary diagnosed postoperatively. The epidemiological, clinical and therapeutic aspects are discussed.

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