American Journal of Surgery Case Reports

Short Communication

Juvenile Granulosa Cell Ovarian Tumors: Report of a Case

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Abstract

Granulosa tumors were described for the first time in 1855 by Rokitansky, due to their appearance similar to that of granulosa cells in ovarian follicles.

The natural history is only reported in the literature through short series, in the 19th century, it was Vonkahladen, in 1895, which individualized granulosa cell tumors, sarcomas, and carcinomas.

In 1914 Vonwerdt published his first series of six cases and the name granulosa tumor was adopted. In 1977, Scully et al. subdivided juvenile and adult granulosa tumor.

Juvenile granulosa tumors of the ovary are rare tumors which represent 5% of ovarian tumors of children and adolescents, the majority of these tumors are diagnosed at FIGO stage 1 with a favorable prognosis.

The purpose of this work is to present the anatomy clinical and radiological characteristics of this tumor in order to allow an early diagnosis and to improve its prognosis, to establish a therapeutic chronology for this tumor which is found in 5% of cases.

We report the observation of juvenile granulosa cell tumor affecting a 33-year-old woman operated on for pain.

Pelvic, the tumor was located in the left ovary with moderate ascites. Different prognostic factors for ovarian granulosa cell tumor are being studied; the treatment is surgical.

Keywords: Ovarian tumor; Juvenile granulosa cells; Surgery; Chemotherapy

Introduction

Granulosa tumors are rare ovarian tumors; arise from the parenchyma and the sexual cord of the ovary, they represent 0.6 to 3% of all ovarian tumors, and 5% of malignant ovarian tumors, they are the most common tumors stromal [1], we distinguish two types: adult granulosa tumor (TGA) between 40 and 70 years, the most common, and juvenile granulosa tumor (TGJ) younger age.

The latter represent characteristics different from other tumors such as the morphological appearance which is different with more pronounced histological signs, a higher risk of recurrence [2,3].

Clinically the tumor manifests itself as a tumor syndrome with painful abdominal distension, an endocrine syndrome linked to hyper estrogen giving pseudo-precocious puberty in juvenile forms [4]. Their diagnosis is essentially based on histological data.

Juvenile granulosa tumors generally have a very good prognosis: 92% 5-year survival [5]. Poor prognostic factors are: large size, ascites and capsular rupture.

Observation

K.F woman aged 33 years, mother of two children, originally from Tiaret and residing in that, with no particular medical or surgical history, who presented to the department for paroxysmal pelvic pain accentuated on the left, clinical examination revealed good general condition, blood pressure 12/06, abdominal palpation revealed accentuated sensitivity on the left.

Ultrasound (Figure 1) revealed a heterogeneous left lateral uterine mass measuring 10 cm in long axis, with a dual tissue and fluid component of suspicious appearance with moderate ascites.

The scan of abdominopelvic (Figure 2) revealed a large, well-circumscribed median pelvic mass measuring 80/110 mm for its longest axes, extending in height to approximately 85 mm, enhancing after injection of the contrast product and heterogeneously delimiting fluid areas in places, calcification coming into contact with the uterine font with respect for the fatty interface of separation.

Homogeneous free ascites of average peri-lesional abundance with pelvic filling, absence of adenomegaly. The biological assessment is unremarkable. Tumor markers: CA 125: 62.84 U/ml.

A midline subumbilical laparotomy was carried out, revealing a large tumor located on the left measuring approximately 08/12 cm, solid cystic, grayish white with necrotic and hemorrhagic changes, Ascites of medium abundance of citrine yellow color.

The patient initially underwent an enucleation cystectomy with sampling of ascitic fluid. The pathological study of the surgical specimen (Figure 3) suggested the diagnosis of ovarian granulosa cell tumor. Cytobacteriological examination of the sample fluid: absence of malignant cells.

Discussion

Granulosa tumors are rare ovarian neoplasia, 70% of malignant

Citation: Sihem D, Yacine BM. Juvenile Granulosa Cell Ovarian Tumors: Report of a Case. Am J Surg Case Rep. 2024;5(1):1114.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Jan 31st, 2024

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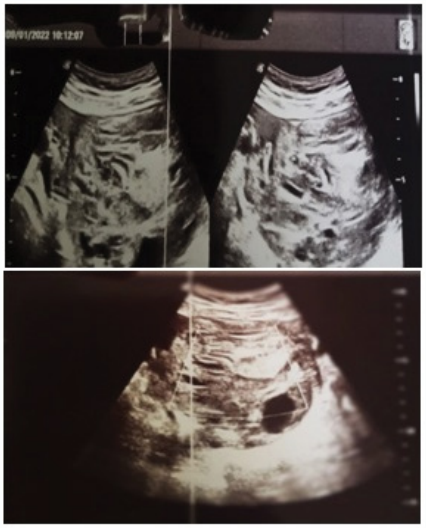

Figure 1: Ultrasound appearance of the tumor.

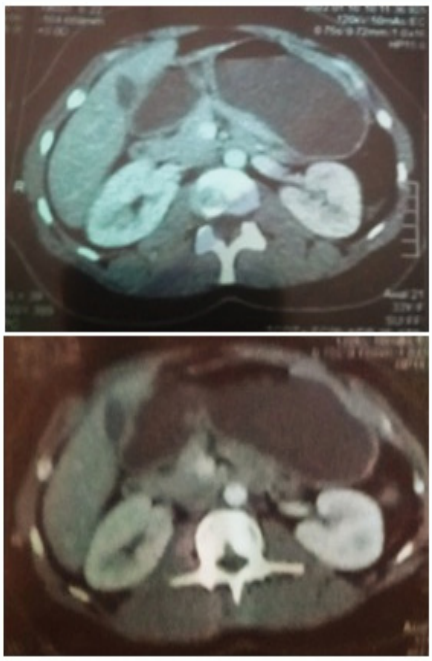

Figure 2: Scan aspect of the tumor.

tumors in the group of mesenchymal and sex cord tumors [6] and 5% among malignant ovarian tumors [1].

Menopausal status, gravidity and parity have no influence on the appearance of granulosa tumors. The juvenile form is far less common, it represents only 5% of granulosa tumors [2], juvenile granulosa cell tumor classically occurs in women young people aged 30 years or in the pre-pubertal period [7,8].

It represents 10% of the etiologies of precocious puberty [7], a picture of hyperestrogen with pseudo precocious puberty is revealing in 70% of cases, much more rarely signs of virilization can appear linked to hyperandrogenism [1,7-9].

The ultrasound and tomographic appearance of TCGJ is that of an extensive multi-cystic ovarian tumor often with hemorrhagic

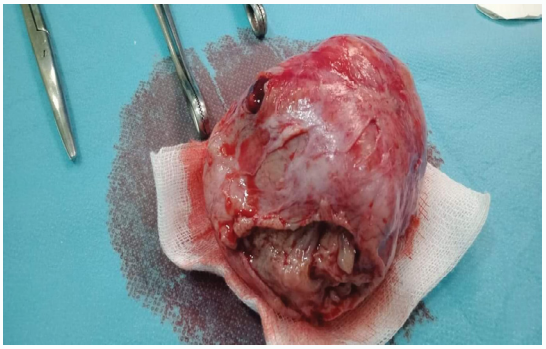

Figure 3: Piece operatoire.

changes, calcifications and peritoneal nodules are rarely reported [4,7], magnetic resonance imaging MRI visualizes a complex pelvic mass, the hemorrhagic character of cyst (hyper signal on weighted T1) is quite suggestive [3,4].

Biologically: serum and urinary levels of eostradiol generally increase, plasma levels of gonadotropins (FSH, LH) are reduced [1, 9,10] in virilizing forms, testosterone and delta 4 Androstenedione levels are increased [9].

TCGJ often presents as a large cystic, multilocular tumor with citrine yellow or gray tissue contingent, pure solid or unilocular cystic tumors are reported, extensive hemorrhagic and necrotic changes are common suggesting a high degree of malignancy [3,4,9], the diagnosis is histological revealing a diffuse or macrofollicular proliferation of luteinized medium cells, mitotic activity is generally absent in the adult form, and evident in the juvenile form the ovarian tumor was discovered in our patient during excruciating abdominal pain which motivated her to consult. Our patient was a young adult; she presented (pathology result): Absence of tumor remnant on the left appendix. The hysterectomy piece with the right appendix, the omentectomy piece and the appendix do not show any tumor infiltration.

The three lymph nodes found in the right dissection as well as the four others found in the left dissection are all reactive, without tumor infiltration, Stage Pt1apN0pMx. The differential diagnosis arises mainly with undifferentiated carcinomas, adenocarcinomas, carcinoid tumors and endometrioid carcinoma. The treatment of these tumors is surgical, conservative for young women wanting a pregnancy and presenting an early stage 1a [9,10], advanced stage tumors and early stage tumors with poor prognosis factors (large size, ascites, histological factor indicating a high risk of recurrence), and recurrent forms requiring polychemotherapy.

Our patient was class stage 1a, she benefited from radical surgical treatment given her bulky size with moderate ascites and capsular rupture [11,12].

Conclusion

Granulosa tumor is a rare tumor but you have to know how to think about an ovarian tumor. It can occur at all ages, the juvenile form is the rarest, it constitutes a particular anatomopathological entity, and it can be associated with other histological lesions such as glandular cystic hyperplasia of the uterus which can go up to degeneration. Currently the treatment is based on surgery, whether it is an initial intervention or for recurrence. The prognosis remains relatively favorable; it depends on the stage of the tumor and the histological type.

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