

## PRIMITIVE MALIGNANT GERM CELL TUMORS OF THE OVARY: ABOUT 11 CASES AND LITERATURE REVIEW.

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### Abstract

*Malignant germ cell tumors of the ovary are rare entities. They mainly affect young patients and can threaten the functional prognosis.*

*We carried out a retrospective study of 11 cases of primary malignant germ cell tumors of the ovary, diagnosed at the paraclinical training and research unit in Pathological Anatomy and Cytology of the CHU/JRA, within a period of 10 years, from January 2010 to December 2019. The ages of these patients varied from 2 to 63 years old with an average of 30,5 years old. In the majority of cases (7 cases), radical treatment was achieved. The guiding clinical information was in all cases an abdominopelvic mass. The right side was the most affected (6/11) and one case was bilateral. The most common histological type was dysgerminoma (4/11).*

**Keyword:** *germ cell, ovary, tumor, histology.*

## INTRODUCTION

Primitive malignant germ cell tumors of the ovary are rare. They represent 2 to 3% of ovarian tumors and affect especially young patients through the second and third decades of life [1]. Our studies were aiming to settle the epidemiological and anatomy-pathological characteristics of these rare tumors.

## MATERIALS AND METHODS

We carried out retrospective and descriptive studies of the cases of primary malignant germ cell tumors of the ovary diagnosed at the paraclinical training and research unit in Pathological Anatomy and Cytology of the CHU/JRA, within a period of 10 years, from January 2010 to December 2019. We included all malignant germ cell tumors cases of the ovary at any age. Our data collection concerned the age, the affected side, the sample type, the clinical orientation signs, and the histological types.

## RESULTS

Throughout the study period, we identified 68 patients diagnosed with malignant ovarian tumor. Among these cases, 11 cases were primary malignant ovarian germ cell tumors, accounting for 16.17%. The ages of these patients varied from 2 to 63 years old with an average of 30.5 years old, whereas within 63.63% of cases, the patients were less than 25 years old. In the majority of cases (7 cases), an oophorectomy was performed. The right side was the most affected (6/11) and one case was bilateral.

The guiding clinical information was in all cases an isolated abdominopelvic mass or associated with other signs (abdominal pain, weight loss). (Table I).

**Table I.** Epidemiological, clinical and anatopathological characteristics of our patients

N°	Age (years)	Sample type	Laterality	Macroscopy aspect	Size (cm)	Histological types
1	2	Oophorectomy	Right	Cerebroid and cystic mass	19	Immature teratoma
2	21	Oophorectomy	Unspecified	Cystic cavity with cerebroid mass	32	Immature teratoma
3	11	Oophorectomy	Right	Cystic and solide mass	22	Yolk sac tumor
4	24	Oophorectomy	Right	Solide mass	7	Yolk sac tumor
5	19	Oophorectomy	Left	Cyst with vegetation	13	Mixte germ cell tumor
6	18	Cystectomy	Right	Solide mass	3	Dysgerminoma
7	21	Oophorectomy	Right	Solide mass with hemorrhage	20	Dysgerminoma
8	56	Hysterectomy and adnexitectomy	Bilateral	Solid mass with necrosis	18	Dysgerminoma
9	63	Oophorectomy	Right	Solide mass with hemorrhage	10	Dysgerminoma
10	46	Biopsy	Unspecified	Cyst without vegetation	2	Choriocarcinoma
11	54	Hysterectomy and adnexitectomy	Left	Mass	1,5	Choriocarcinoma

## DISCUSSION

Germ cell tumors of the ovary constituting a heterogeneous group of tumors developed from primordial cells derived from the embryonic gonad. They are considered to be lesions related to errors in the differentiation and/or migration of primordial cells [2]. The degree and the differentiation pathway determine an histological type: the proliferation of undifferentiated germ cells induces a dysgerminoma, that of totipotent germ cells leads to an embryonic carcinoma, that of cells progressing in the extra-embryonic differentiation pathway leads to a tumor of the yolk sac or choriocarcinoma and that of cells with embryonic differentiation to mature and/or immature teratomas.

Malignant germ cell tumors of the ovary (MGCTO) are rare tumours. They represent 6% of ovarian tumours, with an annual incidence of around 0.5% per 100,000 women in France, around a hundred new cases annually [3]. During the study period, we recruited 68 cases of primary ovarian malignancy with 11 cases of germ cell tumor, representing 16.17% of cases. Over a 20-year period, Tewari et al found 72 cases of MGCTO [4]. Mamouni N et al found 6 cases of MGCTO in 4 years [5].

Malignant germ cell tumors of the ovary are linked to the existence of a cytogenetic characteristic common to all ovarian, testicular or extra-gonadal germ cell tumors, the presence of an isochromosome of the short arm of chromosome 12 [i(12p)] which is not found in any other type of cancer [6]. These tumors occur at any age but are discovered most frequently between the 1<sup>st</sup> and 6<sup>th</sup> decades. In the series by Mamouni N et al [5], the average age of the patients was 22 years with extremes of 1 and 30 years. For Tewari et al [4], the average age at diagnosis was 19 years, with extremes of 9 and 37 years [7]. In our series, the average age was higher with 30.5 years with extremes of 2 and 63 years, but in 64% of cases, our patients were under 25 years old.

The revealing signs of an ovarian tumor are related to the consequences of the rapid increase in tumor volume leading to compression phenomena, pain which may be acute or chronic, an increase in abdominal volume with the perception of a mass, or more rarely torsion, hemorrhage or tumor rupture. A predominance of right-sided involvement is most often observed. In our series, it is above all the existence of an isolated abdominal mass or associated with abdominal pain or weight loss that has been reported. The right side was involved in 6 cases (54.54%), the left side in 2 cases. For 2 patients, the side was not specified and one was bilateral.

Macroscopically, the lesions differ according to histological type. Immature teratomas appear as a mass containing fat and solid portions with numerous microcystic structures of variable size [8]. Their diameter is between 9 and 28 cm with an average of 14 cm [9].

On the macroscopic level, for 2 of our cases, the masses were semi-solid, cerebroid in appearance and semi-cystic with fatty content, measuring respectively 19 and 32 cm in long axis. They were larger compared to those reported in the literature, which can be explained by the rapid evolution of the tumor and the delay in consultation because sometimes people prefer to consult a traditional practitioner before seeing a doctor and the tumor has time to grow. In one of our cases, the patient underwent massage sessions which delayed the diagnosis and led to the extension of the tumor into the peritoneal cavity.

As for dysgerminoma, it presents as a solid, bulky tumor (on average 15 cm) with a bumpy outer surface and the section shows a lobulated, beige appearance containing necrotic and hemorrhagic cystic foci [10]. For our cases, the tumor measured between 3 and 20 cm long axis, with an average of 12.7 cm, which is comparable to what is reported in the literature. The same is true for the macroscopic aspect.

For mixed germ cell tumors, the appearance depends on the type of lesion association present but most often they are solid in appearance [11]. In our case, the tumor was cystic with vegetations on its internal face.

Histologically, there are 2 major groups of MGCTO, dysgerminoma and non-germinomatous tumors. Dysgerminoma is the most frequent form with 30 to 40% of all MGCTO and 10 to 15% are bilateral [12]. In our series, it is also the most frequent form (36.36%) with 1 case of bilaterality (9.09%). For Tewari et al [4] and Mamouni and coll [5], it is in second place after immature teratomas. For non-germinomatous tumors, immature teratoma comes in 2nd position, with 20% of cases. It mainly affects young patients in the second decade [13]. In our series, we observed 2 cases of immature teratoma, 2 cases of Yolk sac tumor and 2 cases of non-gestational choriocarcinoma. In the series of Tewari et al [4], the immature teratoma holds the 1st place (29 cases), followed by the mixed germ cell tumor (15 cases) and the yolk sac tumor (8 cases). For Mamouni N et al [5], immature teratoma also takes first place (3 cases), followed by mixed germ cell tumor (yolk sac tumor with dysgerminoma) and non-gestational choriocarcinoma.

Regarding therapeutic management, when the tumor affects a young woman of childbearing age, the surgeon should consider the possibility of a non-epithelial tumor and do a tumor marker assay before surgery for diagnostic orientation, in particular the assay of HCG and alpha feto-protein.

The gesture therefore consists at least of a unilateral adnexitomy, a complete exploration of the pelvis and the entire abdominal cavity, peritoneal lavage and/or removal of any ascites present when the abdomen is opened, biopsies systematic peritoneal examinations (including at the level of the omentum) and a sample of any suspicious element. It is perfectly legitimate in young women in case of doubt, to perform the surgery in two stages to obtain the definitive results of the histology [14].

Rare malignant tumors of the ovary occur most often in young women (median age: 20 years) even before the first pregnancy, and it is essential to be conservative and to respect the genital tract as much as possible to preserve fertility. In addition, they are tumors with a very good prognosis unlike epithelial tumors.

For our cases, a radical treatment was carried out in 9 out of 11 cases because the tumors were discovered at an already advanced stage and required extensive excision.

## CONCLUSION

MGCTO are rare, we have only recruited 11 cases in 10 years. The most common histological type was dysgerminoma. MGCTO are tumors that preferentially affect young women. Their growth is rapid with a significant increase in their volume, which makes conservative treatment difficult or impossible, thus hampering the obstetrical future of the patient. Earlier diagnosis would have improved management.

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