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Yolk sac tumor: what are the particularities? A case report

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CASE REPORT

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Abstract

We report the case of a 25-year-old girl admitted for management of an abdomino-pelvic mass. Radiological and biological findings were in favour of a malignant ovarian secretory tumor. While insisting on fertility preservation, unilateral adnexectomy combined with chemotherapy was the appropriate treatment.

Key Words: MOGT (malignant ovarian germ cell tumors), OYST (ovarian yolk sac tumors)

Introduction

TGOM germ cell tumors originate in the egg-producing cells (known as germ cells) of the ovaries. These tumors account for 2-3% of all ovarian cancers. Most are found only in the ovary at the time of diagnosis. They usually appear in adolescent girls and women in their twenties[1].

Dysgerminoma is the most common cancerous germ cell tumor of the ovary. In a third of cases, dysgerminoma has spread into the abdomen and pelvis (extra-ovarian spread) by the time of diagnosis. Approximately 20% of dysgerminomas affect both ovaries.

Yolk tumors OYST (tumors of the endodermal sinus) account for around 20% of cancerous germ cell tumors of the ovary. In half of all cases, extra-ovarian spread is observed at the time of diagnosis. Stage 1 vitelline tumours rarely involve both ovaries.

Immature teratomas are composed of cancer cells that resemble the cells of a developing embryo. They usually occur in a single ovary, but can sometimes spread to the other. Immature teratomas are most often diagnosed in girls and young women under the age of 20.

We present an observation of a yolk sac tumor. The clinical, biological, radiological, therapeutic and prognostic features are described.

Observation

A 25-year-old unmarried girl with no notable pathological history was admitted for management of increased abdominal volume associated with abdominopelvic pain with dysuria. The pain was not radiating or associated with digestive symptoms. The patient's general condition was deteriorating, with weight loss not quantified. General examination revealed a conscious patient, normotensive to 120/70 mm/hg, heart rate 86 beats/min, apyretic, BMI 20. Physical examination showed a firm, tender abdominopelvic mass protruding 4 fingerbreadths beyond the umbilicus, with negative giardano on both sides. A gynaecological examination was not performed as the patient was a virgin. The lymph nodes were free.

Pelvic ultrasound revealed a voluminous 20 cm abdominopelvic tissue mass extending into the epigastric region, the origin of which was difficult to determine, with left ureteropelocal dilatation due to compression or invasion (figure1).

Biological workup showed microcytic hypochromic anemia at 9.4g/dl, C-reactive protein at 30 mg/L and negative urine cytobacteriological examination. Tumor marker assays revealed elevated alpha fetoproteins (37,000) and CA125 (157.4), with normal HCG (0.56).

An abdomino-pelvic MRI was therefore ordered, showing a voluminous 19*12*25 cm abdominopelvic mass of left ovarian solid cystic origin, predominantly tissue, and responsible for bilateral ureteropelocal dilatation (figure 2).

The patient underwent exploratory and therapeutic laparotomy. Intraoperatively, a huge 25 cm mass was found over the friable left ovary, with an irregular, nippled surface and a mixed tissue-liquid consistency with no adhesions and moderate ascites. The contralateral ovary was healthy. There was no evidence of peritoneal carcinosis (figure 3). The patient underwent a left adnexectomy, referred for anatomopathological study with peritoneal cytology. The definitive pathology showed a histological and immunohistochemical appearance of a malignant germ cell tumor of the yolk sac tumor type (figure 4). The peritoneal fluid was inflammatory and free of tumour cells.

The extension work-up did not reveal any secondary localization. The patient will also receive 3 sessions of neo-adjuvant polychemotherapy.

Discussion

TGOMs account for less than 3% of ovarian cancers[2]. They are therefore much less frequent than adenocarcinomas. Their incidence is estimated at 0.34 per 100,000 women, which is much lower than for testicular germ cell tumours[3]. However, in young women, they are the most common malignant ovarian tumours[4]. Their incidence increases progressively from birth, peaking between the ages of 15 and 19 and then decreasing[5]. The median age at diagnosis is 18[6].

Ovarian yolk sac tumors (OYSTs) are rare tumors occurring most often in adolescent girls or young women. After ovarian dysgerminomas, they are the second most common histological subtype of malignant ovarian germ cell tumors (OMGCs)[6-8]. They can occur either in isolation (pure OGTT), or in association with another germinal tumor contingent. In this case, they are classified as mixed germ cell tumors, whose prognosis seems to be linked above all to the yolk component, which is the contingent that is potentially the most resistant to treatment. No specific risk factors have been identified for OGTTs. Nor has a familial predisposition syndrome been described. However, given the rarity of these tumors, demonstrating a familial predisposition is difficult[7].

TGOM and, by extension, OYST occur mainly in adolescent girls and young women. They are very fast-growing tumors, prompting early consultation: most patients are seen within 15 days of the onset of clinical signs, which are non-specific but suggestive of a tumoral process[9]. Abdominal pain and the appearance of an abdominopelvic mass are the main diagnostic symptoms[10,11]. Ascites is associated in 50% of cases[10]. Fever occurs in almost a quarter of patients[11]. Less frequently, but not exceptionally, the presentation may be suggestive of an abdominal surgical emergency. The International Federation of Gynecologists and Obstetricians (FIGO) classification for epithelial ovarian tumors is also applied to germ cell ovarian tumors[12]. The majority (55%) of OYSTs are discovered at a localized stage (stage I). The diagnosis is made at stage II (5%) in exceptional cases, at stage III in 25-30% of cases and at stage IV in 10% of cases[9-13].

No imaging test can be used to confirm the diagnosis. However, radiological examinations are useful for orienting the etiology and assessing the extent of the disease. OVSTs have several non-specific imaging features: they are frequently solid tumors that may be partly cystic, with signs of hyper vascularization and hemorrhagic territories[14,15]. Magnetic resonance imaging has not been extensively studied for OYST, but appears to be useful in showing the hyper vascular and hemorrhagic nature of the tumor in T1-weighted sequence[15].

Alpha-fetoprotein, a highly specific marker for vitelline tumours[16,17], aids diagnosis and patient monitoring[18]. It is also a necessary element in determining response to chemotherapy, with normalization of AFP levels if treatment is effective[18]. Finally, AFP is a more sensitive marker than CT for early detection of relapse[16].

OYSTs are quite heterogeneous in morphology, with a variety of histological aspects. Moreover, in the WHO classification, pure Ovarian Dysgerminomas are distinguished from mixed forms associating Ovarian Dysgerminomas with one or more other germ cell tumour subtypes (WHO, 2003). In the vast majority of cases, the tumour is unilateral, unlike ovarian dysgerminoma[10,17.] The tumours are often encapsulated, bulky (mean diameter 15 cm)[10], typically solid and cystic, forming a honeycomb appearance with a smooth outer surface, grey/yellow in color with necrotic and haemorrhagic territories[19]. Progression is usually locoregional in the abdominopelvic cavity. At a very advanced stage, the incidence of liver metastases is high[9]. Lymph node and lung metastases are rarely detected clinically, although the incidence is 62% and 41% respectively in autopsy series[10].

Historically, treatment was based on surgery to remove as much of the tumour as possible, but the advent of cisplatin-based chemotherapy has considerably improved prognosis, with cure rates of around 90%[9,20]. The surgical approach has since evolved towards a conservative approach to preserve fertility in these young women[6,21,22,23]. Currently, the treatment recommended by the French group for the study of rare ovarian tumours is two or three cycles of BEP (Bleomycin- Etopiside- Platinium) in the adjuvant setting. In the case of tumour residue after surgery, or if the tumour is immediately metastatic, four cycles of BEP are proposed[24].

The identification of prognostic factors seems essential in order to adapt treatment, with the aim of limiting toxicity while maintaining the best efficacy. These factors can be classified as clinical, histological and treatment-related prognostic factors[9,11,13,20,25,26].

In the majority of cases (>80%), all stages of OYST are cured by treatment[9,27]. It is therefore important to detect and monitor the occurrence of long-term treatment toxicities in these patients. Post-treatment fertility is also a major concern in these often nulliparous patients. Post-therapeutic monitoring must therefore be closely monitored for the first few years after treatment. For the first two years, it should include a clinical examination every three to six months, AFP assays every month for three months and then every three months, and abdominopelvic ultrasound imaging every three to six months.

Conclusion

Mixed ovarian malignant germ cell tumours are rare. The discovery of an abdomino-pelvic mass in a young girl whose imaging findings are in favor of a malignant ovarian tumor should lead us to suspect the diagnosis. Tumor marker assays (LDH, HCG and alpha fetoprotein) confirm the diagnosis of a germ cell tumor. Management is based on surgery combined with chemotherapy (platinum-based), while preserving fertility.

References

1. Canadian Cancer Society Ovarian cancer tumors.
2. WHO. Pathology and genetics of tumours of the breast and female genital tract. IARC. 2003.
3. Huyghe E, Plante P, Thonneau PF. Testicular cancer variations in time and space in Europe. Eur Urol. 2007;51(3):6218. DOI : https://doi.org/10.1016/J.EURURO.2006.08.024
4. Young Jr JL, Cheng Wu X, Roffers SD, et al. Ovarian cancer in children and young adults in the United States, 1992–1997. International J Ame Cancer Soc. 2003;97(S10):2694-700. DOI : https://doi.org/10.1002/cncr.11351
5. You W, Dainty LA, Rose GS, et al. Gynecologic malignancies in women aged less than 25 years. Obstet Gynecol. 2005;105(6):1405-9. DOI : https://doi.org/10.1097/01.AOG.0000163254.98217.91
6. Tewari K, Cappuccini F, Disaia PJ, et al. Malignant germ cell tumors of the ovary. Obstet Gynecol. 2000;95(1):128-33. DOI :https://doi.org/10.1016/S0029-7844%2899%2900470-6
7. Even C, Lhommé C, Duvillard P, et al. Ovarian yolk sac tumour: general review. B Cancer. 2011;98(8):963-75. DOI: 10.1684/bdc.2011.1387
8. Lai CH, Chang TC, Hsueh S, et al. Outcome and prognostic factors in ovarian germ cell malignancies. Gynecol Oncol. 2005;96(3):784-91. DOI : https://doi.org/10.1016/J.YGYNO.2004.11.027
9. Nawa A, Obata N, Kikkawa F, et al. Prognostic factors of patients with yolk sac tumors of the ovary. Am J Obstet Gynecol. 2001;184(6):1182-8. DOI :https://doi.org/10.1067/MOB.2001.113323
10. Kurman RJ, Norris HJ. Endodermal sinus tumor of the ovary. A clinical and pathologic analysis of 71 cases. Cancer. 1976;38(6):2404-19. DOI : https://doi.org/10.1002/1097-0142%28197612%2938%3A6%3C2404%3A%3AAID-CNCR2820380629%3E3.0.CO%3B2-3
11. Kawai M, Kano T, Furuhashi Y, et al. Prognostic factors in yolk sac tumors of the ovary. A clinicopathologic analysis of 29 cases. Cancer. 1991;67(1):184-92. DOI : https://doi.org/10.1002/1097-0142%2819910101%2967%3A1%3C184%3A%3AAID-CNCR2820670131%3E3.0.CO%3B2-C
12. Jl B. FIGO staging classifications and clinical practice guidelines in the management of gynecologic cancers. Int J Gynecol Obstet. 2000;70:209-62.
13. Ayhan A, Taskiran C, Bozdag G, et al. Endodermal sinus tumor of the ovary: the Hacettepe University experience. Eur J Obstet Gynecol Reprod Biol. 2005;123(2):230-4. DOI : https://doi.org/10.1016/J.EJOGRB.2005.04.021
14. Levitin A, Haller KD, Cohen HL, et al. Endodermal sinus tumor of the ovary: imaging evaluation. AJR Am. J. Roentgenol. 1996;167(3):791-3. DOI : https://doi.org/10.1016/J.EJOGRB.2005.04.021
15. Yamaoka T, Togashi K, Koyama T, et al. Yolk sac tumor of the ovary: radiologic-pathologic correlation in four cases. J Comput Assist Tomogr. 2000;24(4):605-9. DOI : https://doi.org/10.1097/00004728-200007000-00017
16. Talerman A, Haije WG, Baggerman L. Serum alphafetoprotein (AFP) in diagnosis and management of endodermal sinus (yolk sac) tumor and mixed germ cell tumor of the ovary. Cancer. 1978;41(1):272-8. DOI : https://doi.org/10.1002/1097-0142%28197801%2941%3A1%3C272%3A%3AAID-CNCR2820410138%3E3.0.CO%3B2-Z
17. Gershenson DM, Del Junco G, Herson J, et al. Endodermal sinus tumor of the ovary: the MD Anderson experience. Obstet Gynecol. 1983;61(2):194-202.
18. Ishiguro T, Yoshida Y, Tenzaki T, et al. AFP in yolk sac tumor and solid teratoma of the ovary: significance of postoperative serum AFP. Cancer. 1981;48(11):2480-4. DOI : https://doi.org/10.1002/1097-0142%2819811201%2948%3A11%3C2480%3A%3AAID-CNCR2820481122%3E3.0.CO%3B2-8
19. Roth LM, Talerman A. Recent advances in the pathology and classification of ovarian germ cell tumors. Int J Gynecol Pathol. 2006;25(4):305-20. DOI : https://doi.org/10.1097/01.PGP.0000225844.59621.9D
20. Rouge TD, Pautier P, Rey A, et al. Prognostic factors in women treated for ovarian yolk sac tumour: a retrospective analysis of 84 cases. Eur J Cancer. 2011;47(2):175-82. DOI : https://doi.org/10.1016/j.ejca.2010.08.012
21. Mitchell PL, Al-Nasiri N, A'Hern R, et al. Treatment of nondysgerminomatous ovarian germ cell tumors: an analysis of 69 cases. Cancer. 1999;85(10):2232-44.
22. Tong X, You Q, Li L, et al. Prognostic factors of patients with ovarian yolk sac tumors: a study in Chinese patients. Oncol Res Treat. 2008;31(12):679-84. DOI : https://doi.org/10.1159/000173418
23. Cicin I, Saip P, Guney N, et al. Yolk sac tumours of the ovary: evaluation of clinicopathological features and

prognostic factors. Eur J Obstet Gynecol Reprod Biol. 2009;146(2):210-4.
DOI : https://doi.org/10.1016/j.ejogrb.2009.02.052

24. Rouge TD, Pautier P, Duvillard P, et al. Survival and reproductive function of 52 women treated with surgery and bleomycin, etoposide, cisplatin (BEP) chemotherapy for ovarian yolk sac tumor. Ann Oncol. 2008;19(8):1435-41.
DOI : https://doi.org/10.1093/annonc%2Fmdn162
25. Peccatori F, Bonazzi C, Chiari S, et al. Surgical management of malignant ovarian germ-cell tumors: 10 years' experience of 129 patients. Obstet Gynecol. 1995;86(3):367-72. DOI: https://doi.org/10.1016/0029-7844%2895%2900192-T
26. Culine S, Lhomme C, Kattan J, et al. Cisplatin-based chemotherapy in the management of germ cell tumors of the ovary: The Institut Gustave Roussy Experience. Gynecol Oncol. 1997;64(1):160-5.
DOI :https://doi.org/10.1006/GYNO.1996.4547
27. Dällenbach P, Bonnefoi H, Pelte MF, Vlastos G. Yolk sac tumours of the ovary: an update. Eur J Surg Oncol. 2006;32(10):1063-75.
DOI : https://doi.org/10.1016/J.EJSO.2006.07.010

Figures

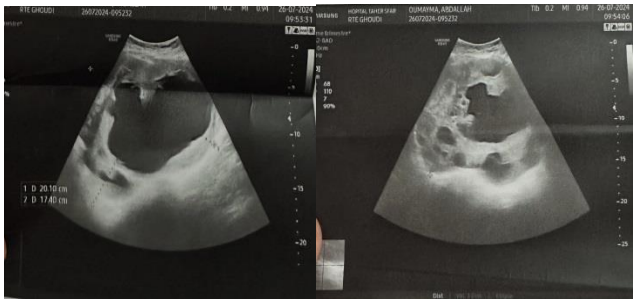

Figure 1: Pelvic ultrasound showing a voluminous 20 cm abdominopelvic tissue mass extending into the epigastric region.

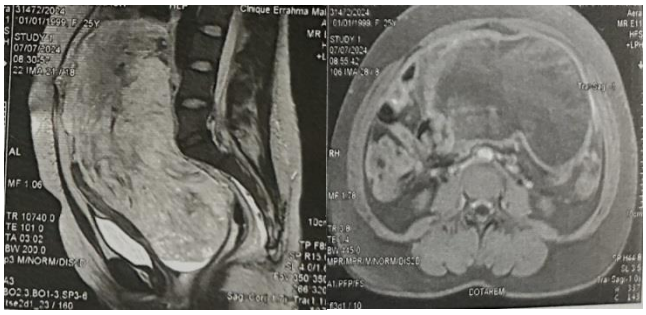

Figure 2: Abdominal and pelvic MRI showing a large 19*12*25 cm abdominopelvic mass of left ovarian solid cystic origin, predominantly tissue.

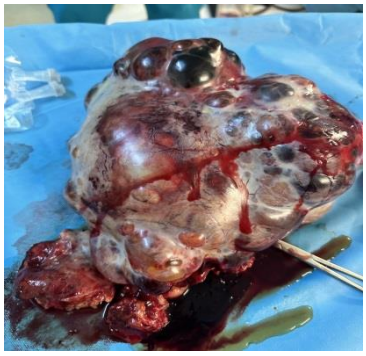

Figure 3: 25 cm mass over the left ovary with irregular, mameloid surface and mixed tissue and fluid consistency.

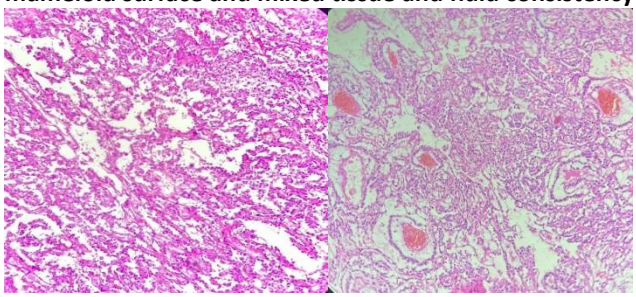

Figure 4: Proliferation of microcystic reticulated architecture/Schiller Duval body.

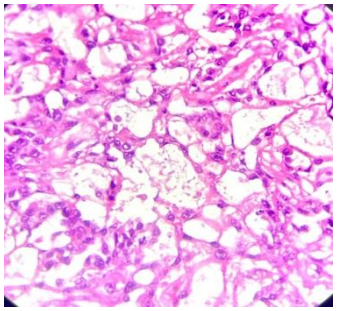

Figure 5: Cells with vesicular nuclei / extracellular hyaline globules.

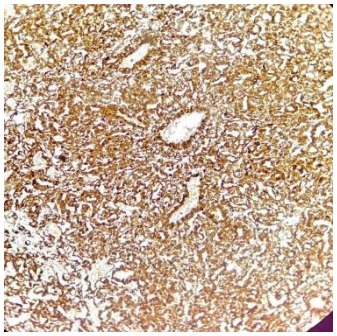

Figure 6: CK: intense and diffuse positivity.

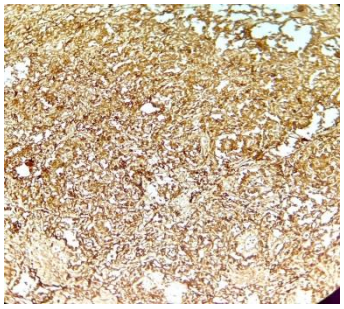

Figure 7: Alpha: intense and diffuse positivity.

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