CASE REPORT

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Giant pure ovarian dysgerminoma in a young 19-year-old patient

Disgerminoma puro de ovario gigante en paciente joven de 19 años

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ABSTRACT

Ovarian dysgerminoma is a very rare neoplasm. It occurs mostly in young women with nonspecific clinical manifestations, although they may express abdominal pain, sensation of mass and menstrual alterations. We report the case of a young multiparous 19-year-old female patient with sensation of intra-abdominal mass, of progressive growth and associated with oppressive pain. On tomographic examination an ovarian-dependent adnexal mass was found, so she underwent a right adnexectomy and freezing biopsy plus staging. Macroscopic evaluation revealed a tumor measuring 25 x 20 x 13 cm, weighing 5,760 grams. By microscopic evaluation and immunohistochemistry studies it was diagnosed as pure dysgerminoma. Based on the average size and history, it was classified as a giant dysgerminoma.
Key words: Ovary, Ovarian neoplasms, Dysgerminoma

RESUMEN

El disgerminoma de ovario es una neoplasia muy infrecuente. Se presenta mayormente en mujeres jóvenes con manifestaciones clínicas inespecíficas, aunque pueden señalar dolor abdominal, sensación de masa y alteraciones menstruales. Se comunica el caso de una paciente joven de 19 años multípara con sensación de masa intraabdominal, de crecimiento progresivo y asociado a dolor de tipo opresivo. En el examen tomográfico se encontró una masa anexial dependiente de ovario, por lo que se le practicó una anexectomía derecha y biopsia por congelación más estadiaje. La evaluación macroscópica evidenció una tumoración de 25 x 20 x 13 cm, que pesaba 5,760 gramos. Mediante evaluación microscópica y estudios de inmunohistoquímica se la diagnosticó como disgerminoma puro. En base al tamaño promedio y antecedentes, fue catalogada como disgerminoma gigante.
Palabras clave: Ovario, Neoplasias ováricas, Disgerminoma

Introduction

Ovarian cancer (OC) is classified by the origin of one of its three main components: epithelium, stroma and germ cells[1]. Malignant germ cell tumors (MGCT) include ovarian dysgerminomas (OD), immature teratomas, yolk sac tumors and mixed germ cell tumors[2]. They constitute approximately 0.9% to 2% of all ovarian malignancies[3]. They usually occur in young patients and are diagnosed before the age of 30 years in up to 85% of cases[4]. The clinical presentation includes late symptoms such as a feeling of heaviness in the pelvis, abdominal distension, lower abdominal pain and menstrual disorders[2].

Its simile in the male is seminoma, which occurs most frequently between the ages of 30-45 years. Clinically, it is a testicular mass that evolves from an *in-situ* germ cell neoplasm, progresses to intratubular seminoma in the post-pubertal stage and then to invasive seminoma in middle age[5].

In dysgerminoma, imaging studies are essential to locate and define the characteristics of the tumor[6]. Macroscopically it is solid and well encapsulated. Microscopically it is composed of round cells with lymphoid infiltrate separated by fibrous strands[7]. Laboratory tests are not specific, but there may be elevation of alkaline phosphatase (ALP), human beta chorionic gonadotropin (βhCG), alpha fetoprotein (AFP), lactate dehydrogenase (LDH) and, in some cases, calcium[8]. OD and seminoma have the

same immunohistochemical markers to confirm the diagnosis: SALL4, OCT4, CD117[2], placental alkaline phosphatase (PLAP)[2] and D2-40 with a mutation in c-KIT[4]. Current treatment consists of fertility-sparing surgery, adjuvant chemotherapy and postoperative radiation. Five-year survival is between 75% and 90% in early stages[9].

Case report

19-year-old patient from Huancavelica-Peru, with obstetric formula G2P2002 and 1 year of illness characterized by sensation of a progressively growing intra-abdominal mass associated with oppressive pain of moderate intensity, of intermittent course. On physical examination, vital signs were stable. The abdomen was globular, soft, depressible, non-painful to palpation and there was a non-mobile mass of approximately 20 x 25 cm.

A Papanicolaou was performed with cervical biopsy, and a squamous carcinoma in situ with glandular extension was found. The following day laboratory tests were performed and highlighted the elevation of CA-125, LDH and βhCG (Table 1). Additionally, a computerized tomography (CT) scan was performed, which revealed an abdominopelvic mass (Figure 1A).

Table 1. Laboratory values.

Laboratory tests	Normal values	Results
CA125 (U/ml)	0 - 35	125
AFP (IU/ml)	0 - 8	95.15
LDH (U/L)	140 - 280	794
βhCG (mUI/L)	0	134.430
hCG (mIU/ml)	0 - 5	142.420

APF: alfa-fetoprotein; LDH: lactic dehydrogenase

Right adnexectomy with freeze biopsy plus conservative staging was planned. The pathology report revealed that the tumor dimensions were 25 x 20 x 13 cm, with a weight of 5,760 grams. In addition, the capsule was intact, smooth and shiny. Multiple sections showed firm tissue with a 'fleshy' appearance, with gelatinous foci. The right uterine tube had no alterations (Figure 1B).

Microscopic description of the slides identified nests of tumor cells with alveolar pattern (Figure 2A). The tumor cells characteristically presented abundant pale cytoplasm, single nucleus with prominent round nucleolus, mitotic range of 3 in 10/HPF (high power fields) (Figure 2B).

Regarding immunohistochemistry studies, neoplastic cells were positive for SALL4, CD117 and OCT ¾ (Figure 3).

Figure 1. A. Simple and contrasted computerized tomography (CT): in the pelvic excavation a solid, heterogeneous image with apparent dependence on the right adnexa was observed. Presence of free fluid in the peritoneal cavity. B. Macroscopic appearance: hemorrhagic wine-brown tissue.

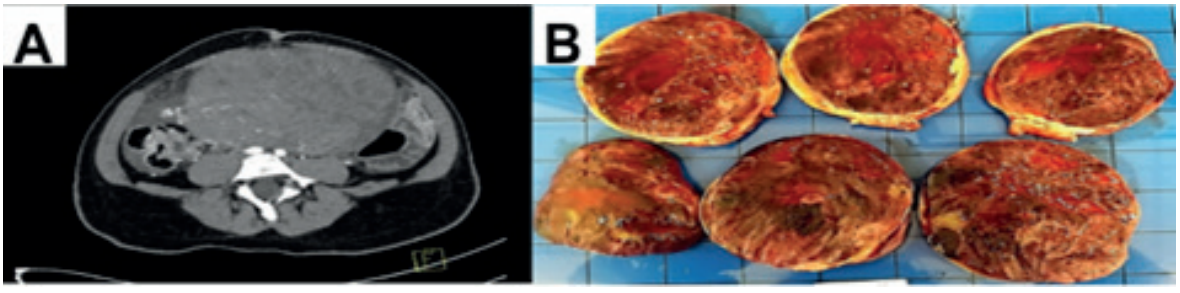


Figure 2. A. Microscopic view with HE staining. 10X: Alveolar pattern separated by fibrous connective tissue septa. B. 40X: Scarce lymphocytes are observed in the septa.

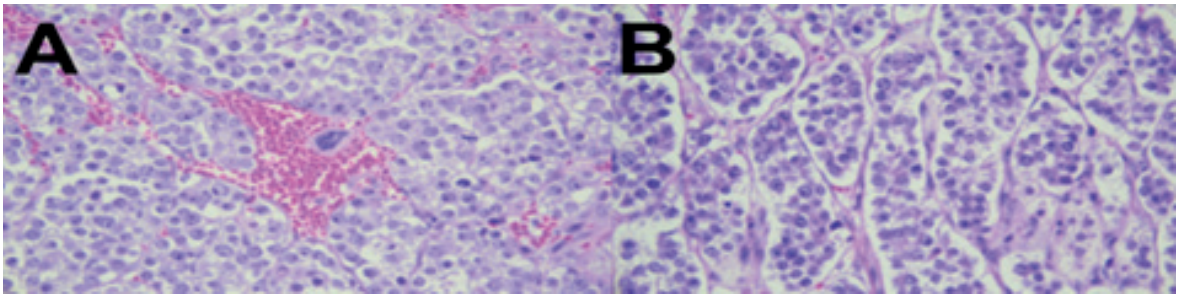
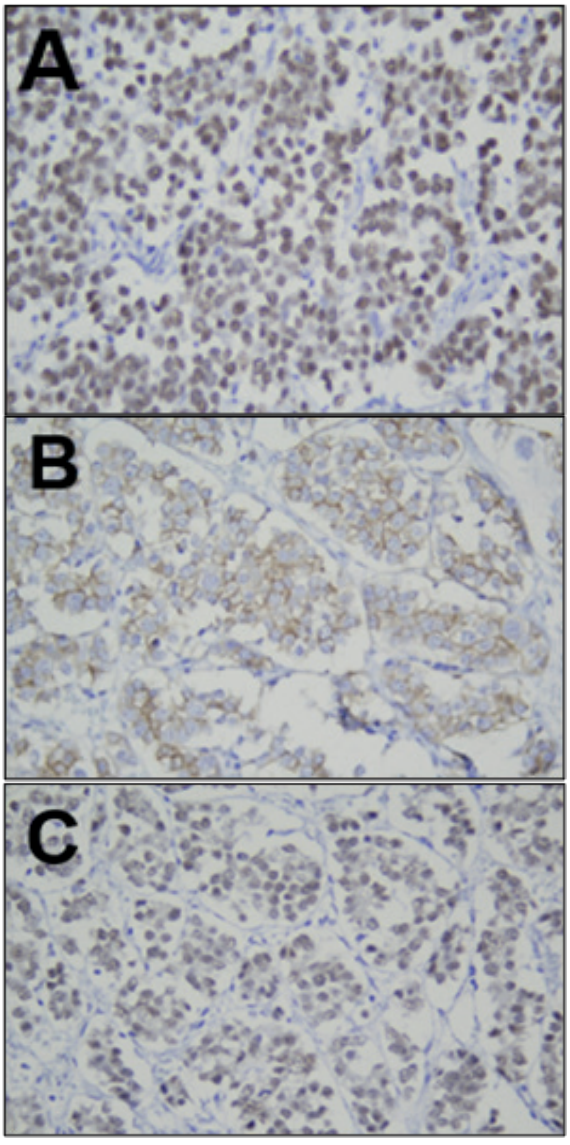


Figure 3. A. Immunohistochemistry, 40X. SALL4 positive, nuclear pattern in tumor cells. B. Immunohistochemistry, 40X. CD 117 positive, cell membrane staining is observed in tumor cells. C. Immunohistochemistry, 40X. OCT ¾ positive, nuclear pattern in tumor cells.



Upon characteristic microscopic evaluation and confirmation by immunohistochemistry studies, it was concluded that the case was pure OD. The staging of the patient, according to the International Federation of Gynecology and Obstetrics (FIGO) classification, was as stage IA, and according to TNM classification (T: tumor, N: nodule, M: metastasis) with pT1a (p: pathologic, T: tumor). The patient responded well to treatment and remains without signs of recurrence.

Discussion

OD is the most common subtype of MGCT. It usually presents during early reproductive ages as in our patient. Although the symptoms are non-specific, Rungoutok M. and Suprasert P. reported in their study that the three most common symptoms were pelvic mass sensation, lower abdominal pain and abdominal distension[10], as occurred in the presented case.

CT allows determining the location and extension of the mass. In this case, a well-defined right solid mass of neoformative character was evidenced. However, multilobulations with interposition of fibrovascular septa and a prominent vascular pedicle with tortuous vessels could also be found[11].

ODs are usually diagnosed with a size larger than 15 cm[12]. In a review of 140 cases of OD the average size was found to be 13 cm in diameter[11]. A case of a pregnant woman with a giant OD of 25 x 19 x 24 cm has been published[7]. In our case, the patient presented a tumor measuring 25 x 20 x 13 cm. According to the described history, it is classified as a giant OD.

Immunohistochemistry in our case, SALL4 helped to differentiate MGCT (sall4+) from sex cord tumors (sall4-). Oct 3/4 is expressed in germ cells and is found negative in yolk sac tumor. CD117 is only positive in OD or seminomas compared to the rest of MGCTs[5]. A review of 140 cases emphasizes that dysgerminoma has very particular findings in immunohistochemistry compared to its differential diagnoses, which is key to its precise diagnosis[11].

OD does not usually present hormonal alterations, but up to 5% of cases may show elevated βhCG and within serology, elevated LDH and to a lesser extent ALP. Our case presented elevation of these values[11].

In young patients, the standard treatment for stage IA consists of fertility-conserving surgery (considering the desire for parity) with adnexectomy with unilateral freezing biopsy plus staging. Chemotherapy is not usually indicated

at this stage unless there is recurrence[7]. In advanced stages, complete resection plus 4 cycles of chemotherapy is usually performed[13]. In the case of our patient, being a stage IA, a unilateral adnexectomy / oophorectomy plus freezing biopsy and conservative staging was performed. Although it is true that there is an incidental finding of carcinoma in situ, there is still no evidence that it is related to OD.

In conclusion, OD is a rare malignant tumor of unknown etiology and is common in young women of reproductive age. The most common symptoms are pelvic mass sensation, lower abdominal pain and abdominal distension. Immunohistochemical and anatomopathological studies are essential to confirm the diagnosis. The prognosis is favorable especially in the early stages, so these cases should be referred promptly to a specialized center and continue with individualized management.

References

1. Gaona-Luviano P, Medina-Gaona LA, Magaña-Pérez K. Epidemiology of ovarian cancer. Chinese Clin Oncol. 2020;9(4):47. doi: 10.21037/cco-20-34
2. Uccello M, Boussios S, Samartzis EP, Moschetta M. Systemic anti-cancer treatment in malignant ovarian germ cell tumours (MOGCTs): current management and promising approaches. Ann Transl Med. 2020;8(24):1713–1713. doi: 10.21037/atm.2020.04.15
3. Tîrnovanu MC, Florea ID, Tănase A, Toma BF, Cojocaru E, Ungureanu C, et al. Uncommon metastasis of ovarian dysgerminoma: A case report and review of the literature. Med. 2021;57(6):1–12.doi.org/10.3390/medicina57060534
4. Cheung A, Shah S, Parker J, Soor P, Limbu A, Sheriff M, et al. Non-Epithelial Ovarian Cancers: How Much Do We Really Know? Int J Environ Res Public Health. 2022;19(3): 1106. doi: 10.3390/ijerph19031106
5. Song S. Dysgerminoma PathologyOutlines.com website [Internet]. Michigan [cited 2023 May 6]. Available from: https://www.pathologyoutlines.com/topic/ovarytumordysgerminoma.html.
6. Cacioppa LM, Crusco F, Marchetti F, Duranti M, Renzulli M, Golfieri R. Magnetic resonance imaging of pure ovarian dysgerminoma: a series of eight cases. Cancer Imaging. 2021;21(1):1–7. doi: 10.1186/s40644-021-00427-1
7. Zhang XW, Zhai LR, Huang DW, Jiang ZD, Yu T, Liu SY, et al. Pregnancy with giant ovarian dysgerminoma: A case report and literature review. Medicine (Baltimore). 2020;99(41):1-7. doi: 10.1097/MD.0000000000021214
8. Hara N, Suwanai H, Abe H, Yakou F, Ishikawa T, Urayama M, et al. Hypercalcemia associated with dysgerminoma and elevation of calcitriol: A case report and review of the literature. SAGE Open Med Case Reports. 2022;10:1-6. doi: 10.1177/2050313X211068562
9. Adhikari S, Joti S, Chhetri PK. Paediatric Ovarian Dysgerminoma: A Case Report. J Nepal Med Assoc. 2022;60(255):985–988. doi: 10.31729/jnma.7894
10. Rungoutok M, Suprasert P. Oncology and reproductive outcomes over 16 years of malignant ovarian germ cell tumors treated by fertility sparing surgery. World J Clin Oncol 2022; 13(10): 802-812. doi: 10.5306/wjco.v13.i10.802
11. Warnnissorn M, Watkins JC, Young RH. Dysgerminoma of the ovary: An analysis of 140 cases emphasizing unusual microscopic findings and resultant diagnostic problems. Am J Surg Pathol . 2021;45(8):1009-1027. doi:0000000000001687
12. Rogers D, Menias C, Shaaban A. Malignant Germ Cell Tumors of the Ovary: Clinical and Imaging Features. Radiol Clin North Am. 2023 Jul;61(4):579-594. doi: 10.1016/j.rcl.2023.02.004. doi: 10.1016/j.rcl.2023.02.004
13. Sas I, Şerban DM, Tomescu LC, Nicolae N. Ovarian dysgerminoma in pregnancy: A case report. Medicine (Baltimore). 2021;100(14):e25364. doi: 10.1097/MD.0000000000025364