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

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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

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).

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 infiltration 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\(^{\text{(2)}}\), placental alkaline phosphatase (PLAP)\(^{\text{(2)}}\) and D2-40 with a mutation in c-KIT\(^{\text{(4)}}\). Current treatment consists of fertility-sparing surgery, adjuvant chemotherapy and postoperative radiation. Five-year survival is between 75% and 90% in early stages\(^{\text{(9)}}\).

**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).

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).

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**Table 1. Laboratory values.**

<table>
<thead>
<tr>
<th>Laboratory tests</th>
<th>Normal values</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>CA125 (U/ml)</td>
<td>0 - 35</td>
<td>125</td>
</tr>
<tr>
<td>AFP (IU/ml)</td>
<td>0 - 8</td>
<td>95.15</td>
</tr>
<tr>
<td>LDH (U/L)</td>
<td>140 - 280</td>
<td>794</td>
</tr>
<tr>
<td>βhCG (mUI/L)</td>
<td>0</td>
<td>134430</td>
</tr>
<tr>
<td>HCG (mIU/ml)</td>
<td>0 - 5</td>
<td>142420</td>
</tr>
</tbody>
</table>

*APF: alpha-fetoprotein; LDH: lactic dehydrogenase*
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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, 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.

ODs are usually diagnosed with a size larger than 15 cm. In a review of 140 cases of OD the average size was found to be 13 cm in diameter. A case of a pregnant woman with a giant OD of 25 x 19 x 24 cm has been published. 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. 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.

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.

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