Gliomatosis peritonei in mature ovarian teratoma

Gliomatosis peritoneal asociada a teratoma maduro de ovario

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ABSTRACT

Ovarian teratomas are tumors derived from the three germinal layers. Gliomatosis peritonei is a rare condition characterized by glial tissue implants in peritoneum, commonly associated with immature ovarian teratoma, but not with mature teratomas. Definitive diagnosis is based on histopathological examination. Currently, there are no guidelines for its follow-up. Prognosis is favorable; however, implants may present fibrotic regression or exceptionally degenerate into glioblastoma. Treatment should be similar to that of metastatic ovarian carcinoma if immature glial tissue or other teratomatous components are found in peritoneum or omentum. Long-term follow-up is necessary in patients with residual peritoneal disease due to risk of recurrence and malignant transformation. We present a case of gliomatosis peritonei associated to mature ovarian teratoma.

Key words: Ovarian neoplasms, Teratoma, mature, Ovary.

RESUMEN

Los teratomas de ovario son tumores compuestos por tejidos derivados de las tres líneas germinales embrionarias. La gliomatosis peritoneal es una afeción poco frecuente caracterizada por implantes de tejido glial en el peritoneo comúnmente asociada con teratoma inmaduro de ovario, no así con teratomas maduros. El diagnóstico definitivo se realiza por histología. No existen pautas sobre cómo realizar el seguimiento de los pacientes con esta afeción. El pronóstico es favorable. Sin embargo, los implantes pueden experimentar regresión fibrotica o excepcionalmente degenerar a glioblastoma. Si están presentes tejido glial inmaduro u otros componentes teratomatosos en el peritoneo o epiplón, el tratamiento será similar al del carcinoma metastásico de ovario. En pacientes con enfermedad peritoneal residual, es necesario un seguimiento a largo plazo, debido al riesgo de recurrencia y transformación maligna. Se presenta un caso de gliomatosis peritoneal asociado a teratoma maduro de ovario.

Palabras clave: Neoplasias ováricas, Teratoma, maduro, Ovario.

INTRODUCTION

Gliomatosis peritonei is a rare entity characterized by the presence of benign glial implants in peritoneum, omentum and lymph nodes. It is generally associated with immature ovarian teratomas and rarely with mature teratomas(1). Because of its low frequency, there are no protocols for its follow-up, and prognosis has not been established. Some reports link it to pregnancy and ventriculoperitoneal shunts for hydrocephalus(2,3). We present a case of gliomatosis peritonei associated with a mature ovarian teratoma.

CASE REPORT

A 21-year-old patient, gravida 3, para 2, abortus 1, consulted for intermittent, nonradiating pain of moderate intensity in the right iliac fossa for 7 days, associated to abdominal bloating, increased body temperature and loss of appetite. She denied any urinary alterations, changes in bowel movements, weight loss, chronic or endocrine diseases, cancer, smoking, using prescription or illicit drugs, and had no contributing family history. She had smoked approximately 10 cigarettes a day for the last two years. Regarding her gynecological history, she experienced menarche at age 14, and had irregular periods every 2 to 3 months, lasting 6 to 7 days; her first sexual intercourse was at age 16. Her last period had happened 13 days before her symptoms started.
At physical examination, the patient was conscious and oriented. Heart rate, blood pressure and respiratory rate were normal. Abdomen was soft and depressible; bowel sounds were present. We palpated a soft tumor located 2 cm above the navel, slightly movable, painful on deep palpation. Pelvic examination revealed pain on movement of the cervix. Cervix was closed, without evidence of bleeding or vaginal discharge. We could not palpate the tumor with this method. Upon rectal exam, anal sphincter tone and rectal cavity were normal, and there was a palpable tumor occupying part of the pouch of Douglas.

Transvaginal ultrasound showed a homogenous uterus of smooth contours measuring 8 x 3 x 3 cm, with hyperechoic, thickened endometrium (11 mm) and intact subendometrial halo. It also revealed a hyperechoic, heterogeneous abdominopelvic tumor measuring 20 x 19 x 11 cm, probably originating in the right ovary, presenting multiple cysts of variable size and a solid area of 4 x 1 cm. Its walls were thick and measured 4 mm, and it had multiple septations 3 mm wide, with papillary outgrowth and projections in the internal portion; we assigned it a Sassone score of 12 (Figure 1). The left ovary measured 3 x 2 x 2 cm. There was a small amount of free intraperitoneal fluid, and minimal right pyelectasis. Ultrasound showed no alterations in the other organs. Blood tests, liver and renal function tests, electrolytes and coagulation profile were within normal values. Beta-hCG was negative.

Concentrations of tumor markers CA-125 (165 UI/mL, reference value (RV) under 35 UI/mL) and alpha-fetoprotein (40.5 ng/mL, RV under 10 ng/mL) were elevated. Given these findings, we considered the possibility of a malignant ovarian tumor.

In the exploratory laparotomy, we found approximately 200 mL of free intraperitoneal fluid, which we sampled for cytology. The right ovary presented a multiloculated cystic tumor measuring 20 x 17 x 13 cm, with its capsule intact (Figure 2). The fallopian tube was elongated and strongly adhered to the tumor. The omentum had a spherical tumor of approximately 2 cm in diameter, with multiple implants in appendix, intestines and colon. There was no seeding in diaphragm, liver, gallbladder, spleen and kidneys. We resected the lesion in omentum, took a sample of the peritoneal implants and of several portions of the ovary and sent them for frozen section biopsy, which was informed as mature cystic ovarian teratoma and gliomatosis peritonei. We performed right oophorosalpingectomy, partial omentectomy, comprehensive surgical staging, appendectomy, biopsy of some of the peritoneal implants, bilateral pelvic lymphadenectomy, and biopsy of some paraaortic lymph nodes. The patient tolerated the procedure well and the surgery ended without complications.

Microscopic evaluation showed mature tissue derived from the three germinal layers with ectodermal predominance and an organized pattern, with skin next to skin appendages, co-

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**Figure 1. Abdominopelvic ultrasound image depicting cystic tumor with multiple septal.**

**Figure 2. Mature teratoma of the right ovary after resection.**
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Figure 3. a) ovarian tissue with squamous and columnar epithelium, pilous follicles, fat and cartilage. b) section from the ovarian teratoma, with skin and skin appendages. c) peritoneal nodes, with fibroconnective and adipose tissue, and mesothelial lining with mature glial tissue. d) glial tissue with nonspecific peripheral inflammatory response. Hematoxylin-eosin staining.

The patient had no post-operative complications and was discharged 4 days later. We reevaluated her one week after discharge; afterwards, she did not return for the follow-up consultation.

Discussion

Ovarian teratomas are tumors composed of tissue derived from the three embryonic germ layers (endoderm, mesoderm and ectoderm), present in varying quantities within the tumor. Gliomatosis peritonei is a condition where glial tissue implants appear in peritoneum; its peak incidence occurs during the second decade of life(4). The association of gliomatosis peritonei with immature ovarian teratomas is rather common, unlike its association with mature teratomas(5). Given its low frequency – there are only circa 100 reports –, we do not know its real incidence(2).

Several theories propose a pathogenesis for this condition and possible factors responsible for its development in association with ovarian teratomas. Some authors suggest that glial implants come directly from the teratoma, possibly by rupture. This mechanism is based on the presence of implants in omentum close to capsule defects. The theory of dissemination by angiolymphatic vessels builds on the findings of mature glial tissue in paraaortic and pelvic lymph nodes in patients with immature ovarian teratoma(6). It has also been proposed that the implants do not come from the teratoma, but from pluripotent stem cells in peritoneum, due to genetic analyses evidencing differences between glial implants and teratomas. This phenomenon may happen because of pluripotent Müller cells undergoing glial metaplasia in the peritoneal surface or subcelomic mesenchyma in response to factors secreted by the teratoma, which may induce specific differentiation pathways. The occurrence of gliomatosis peritonei in patients with ventriculoperitoneal shunt, which would allow cerebrospinal fluid to transport glial tissue, supports this theory. Nevertheless, the specific mechanisms by which peritoneal cells undergo glial transformation remain unknown(7).

Alpha-fetoprotein is a serum marker used for the diagnosis and follow-up of germ cell tumors. Its use in the follow-up of patients with ovarian teratoma may be hindered by its low sensitivity and specificity, although it could help rule out metastasis of immature ovarian teratomas(8). Concentrations may remain within normal values in cases of gliomatosis peritonei, even if they were elevated with the original teratoma. They may also appear normal in recurrent tumors containing immature elements(7).

Glial peritoneal implants are small and may be in parietal or visceral peritoneum. Pelvic structures can be evaluated by transvaginal ultrasound. However, implants are usually very small (smaller than 3 mm), so they may not be detected. They can also be hidden by intestinal gas, or outside the visualization field(9). CT SCAN and MRI may provide a better detection, although their etiology may be confused with carcinoma.
Gliomatosis peritonei in itself has low malignant potential. Its prognosis relates to the type of the primary ovarian teratoma. Survival rate at 5 years varies between 30 and 82%, depending on histologic findings and surgical staging. Good prognosis is also associated with implants composed exclusively of mature glial tissue, although recurrence is more frequent and disease-free survival is shorter. Implants may remain stable, undergo fibrous regression, or transform into glioblastoma\(^{(11)}\). Long-term follow-up is necessary to rule out malignant transformation, given that some cases progress to malignancy up to 7 years after surgery, especially when associated to immature ovarian teratoma or in cases without comprehensive surgical staging, histologic evaluation of implants and treatment with chemotherapy\(^{(5,13)}\). Until now, there have been no reports of malignant transformation of gliomatosis peritonei associated to mature cystic teratoma. Patients with immature ovarian teratoma and glial implants have a better prognosis if peritoneal surface, omentum and diaphragmatic surfaces have implants exclusively or almost exclusively composed of mature glial tissue\(^{(9,11)}\).

In conclusion, gliomatosis peritonei consists of mature glial tissue implants in peritoneal surface. It is often associated with immature ovarian teratomas, not with mature teratomas. Despite the inactivity of residual peritoneal disease, it requires long-term follow-up. Prognosis is generally favorable.

**References**


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