CASE REPORT

Poorly differentiated ovarian teratoma: case report with a 17-year survival after undergoing cytoreductive surgery

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SUMMARY

Immature ovarian teratomas are rare malignant tumors originating in ovarian germ cells, representing about 1% of ovarian cancer cases. Main treatment is surgery and chemotherapy. The degree of differentiation is an important prognostic factor. A well-differentiated teratoma (G1/G2) is usually associated with a higher survival rate than poorly differentiated tumors (G3). The current report shows a case of poorly differentiated immature ovarian teratoma diagnosed in a 42-year-old patient with a disease-free survival above the mean rate described in the literature (17 years), highlighting the importance of cytoreductive surgery in advanced cases conducted by specialized surgical oncologists.

INTRODUCTION

Immature teratomas are malignant tumors that derive from cells that have the potential to develop into different types of tissues in the body, including the skin, internal organs and muscles. Immature ovarian teratoma is a rare cancer that represents about 1% of ovarian cancers. Main treatment is surgery and chemotherapy.[1] The degree of differentiation of teratomas plays a primordial role in determining patient prognosis. Patients with more differentiated tumors (G1/G2) have a five-year survival of 91.4%, in comparison to those with poorly differentiated (G3) tumors, where the survival rate is 56%.[2] Therefore, early identification and accurate pathological classification are essential for appropriate treatment.

The aim of this study is to report a case of immature ovarian teratoma that underwent surgical and chemotherapy interventions and has had a long-term survival (17 years), compared to the mean survival rates reported in the literature. The importance of complete cytoreduction by surgeons specialized in gynecologic oncology is highlighted.

CASE PRESENTATION

A 42-year-old woman, with important history of past illness (arterial hypertension, diabetes mellitus, smoking and alcohol consumption), was referred to our service in February/2008 with a history of ovarian cancer previously treated with surgery and chemotherapy in another healthcare service. She reported vague abdominal symptoms and had been diagnosed with left ovarian tumor, measuring 17 cm, with bowel loop involvement. In both previous surgeries in April 2007, a bilateral salpingoophorectomy, omental biopsy and peritoneal fluid collection were performed. Peritoneal carcinomatosis was considered unresectable. Chemotherapy was indicated in the hope that surgical rescue could be attempted later. Alpha fetoprotein (AFP) and β-HCG levels were normal.

Histopathology study revealed a poorly differentiated (G3) immature ovarian teratoma, measuring 16 cm in the right ovary, and with disease in the left ovary. The fallopian tubes showed no disease. There was disease in the epiploon, and oncotic cytology was negative.

The patient underwent chemotherapy with four cycles of BEP (bleomycin, etoposide and cisplatin). In August/2007, a new surgery was attempted and again the disease was considered unresectable. Only peritoneal biopsies were done, confirming the diagnosis of poorly differentiated teratoma, and the patient was referred to palliative chemotherapy. She was also referred to another surgical team where she underwent total hysterectomy, total pelvic peritonectomy (Figure 1), epiploectomy and resection of a tumor of the epiploon measuring 15 cm, along with removal of the spleen and tail of the pancreas (Figure 2).

Furthermore, resection of multiple peritoneal implants was performed with residual tumor implants of less than 3 mm, considered a suboptimal cytoreduction. There were no suspicious pelvic and retroperitoneal lymph nodes, and resection of retroperitoneal lymph nodes was not performed. After surgery, the patient received three cycles of chemotherapy with BEP, demonstrating a favorable clinical response and improvement of the symptoms reported. Total abdominal tomography and laboratory tests performed after treatment did not reveal any evidence of peritoneal cancer.

Regular follow-up of the patient was performed with imaging tests and physical examination. However, local and distant

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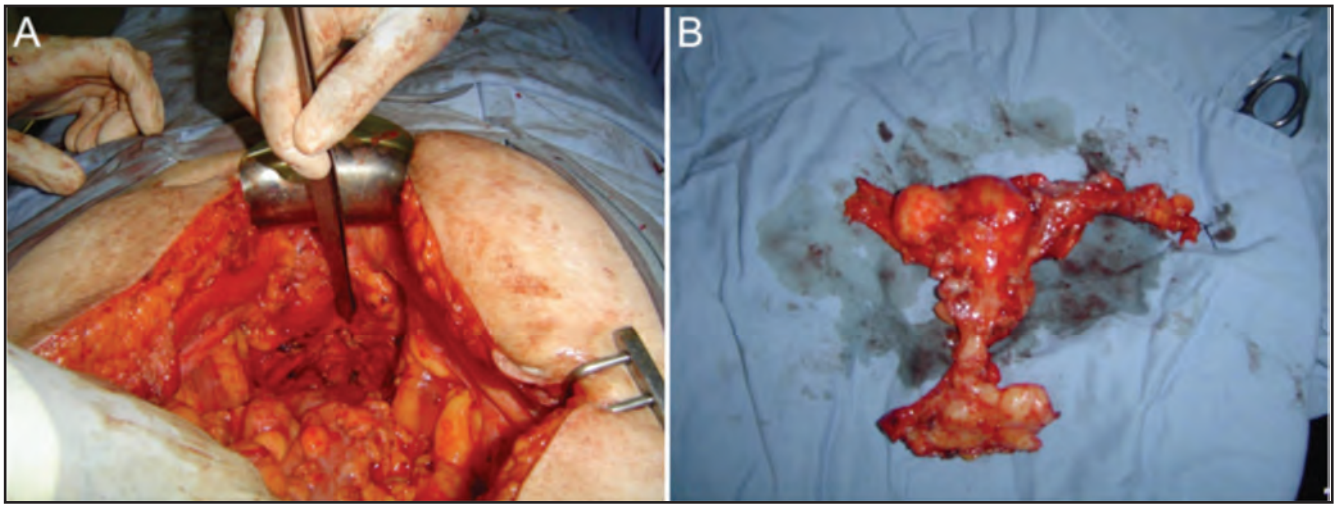

Fig. 1: A: Total pelvic peritonectomy. B: Surgical specimen of total hysterectomy with pelvic peritoneum

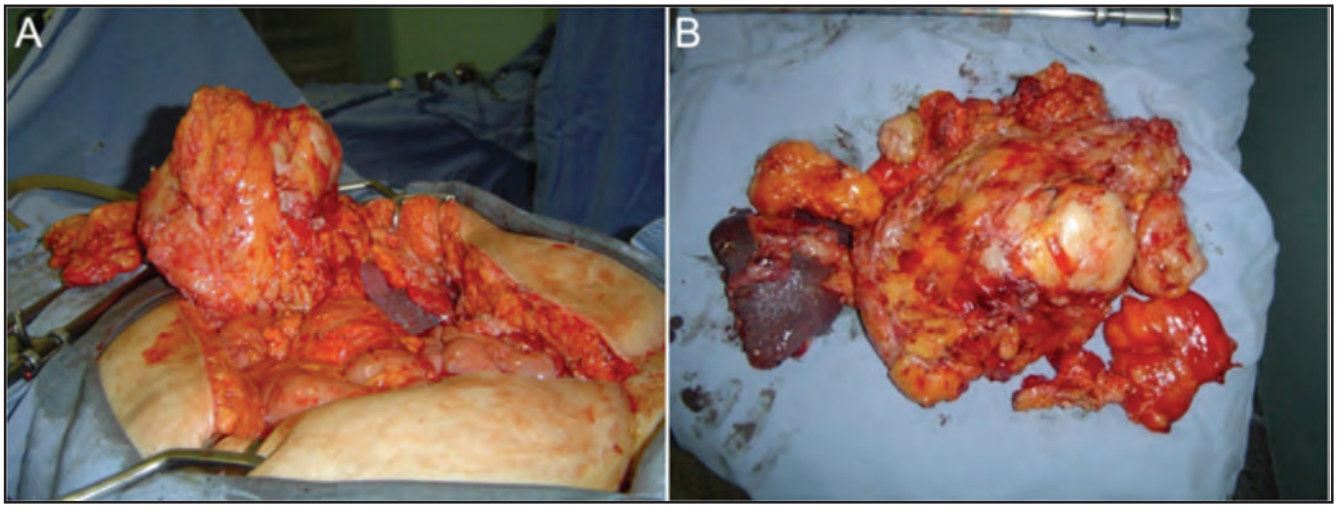

Fig. 2: A: Tumor in the epiploon and visualization of spleen adhesion to tumoral mass. B: Surgical specimen of resection of the epiploon, spleen and tail of the pancreas

disease recurrences were not observed during follow-up care. In June/2013, on the abdominal tomography for control, a solid lesion suspicious of tumor recurrence, measuring around 5 cm in its largest diameter was identified in the vaginal vault. Imaging tests showed no other abdominal or thoracic lesion, and the asymptomatic patient was in good general health. During the clinical exam of the vagina, the vaginal vault lesion was not palpated. Complete resection of the vaginal vault lesion was done by the abdominal and vaginal route. No other lesions were identified in the abdominal cavity during surgery. The patient had a good postoperative recovery without any complications.

Histopathological and immunohistochemical tests of the vaginal vault lesion demonstrated a benign myxoid fusocellular tumor. Immunohistochemical panel test showed AE1/AE3 negative, AML positive, Q-BEND10 focal positive, desmin negative, EMA negative, S100 negative and estrogen and progestrone receptor negative. Subsequently, benign bilateral breast masses were detected, in addition to simple kidney cysts and multiple thyroid nodules that were benign on cytology analysis by fine-needle aspiration (FNA). Nowadays, 17 years after the diagnosis of immature teratoma, the patient is in good overall health. She performs her normal work activities, has a good quality of life and has no evidence of active disease. She is taking transdermal and vaginal estradiol.

This study is part of a cancer patient project approved by the Research Ethics Committee of the State University of Piauí, with Technical Report No 4.311.835. The patient signed the Free Informed Consent Term (FICT).

DISCUSSION

Immature ovarian teratoma occurs predominantly in children and women in reproductive age. Around 80% of these cases are unilateral tumors. Histological analysis shows three immature germ layers containing embryonic elements that may include the neuroepithelium.[3]

Immature teratoma may manifest itself as a calcified pelvic mass, abnormal uterine bleeding or pelvic discomfort. The most common sites of dissemination are the peritoneum and retroperitoneal lymph nodes. Hematogenous spread to the lungs, liver or brain is rare. In approximately half of these cases, there is an increase in tumor markers, such as alpha fetoprotein (AFP) and β-HCG levels.[4] However, in the patient mentioned here, the results of these markers were within the normal range, which may also be related to the better prognosis presented.

Among the tumor markers, AFP is highlighted. It is the major fetal serum protein whose main function is plasma transport and regulation of oncotic pressure. Furthermore, AFP serum level provides an estimate of tumor growth period. In addition, β-HCG (human chorionic gonadotropin), an essential marker for diagnosis and monitoring of patients with germ cell tumors, was ordered to conduct a more specific evaluation of the clinical situation of the patient.[4]

In the case reported, the patient was a female in her forties. She is alive and without disease 17 years after diagnosis, although her case was considered inoperable in the first two surgeries. The presence of poorly differentiated immature cells is a poor prognostic factor, increasing the risk of recurrence. Nevertheless, when cytoreduction (even suboptimal) is achieved, it may contribute to determine a long survival period, as shown in the current case.[2]

Surgical intervention is the first treatment option for women with immature teratomas. This approach not only allows adequate tumor staging, but also enables the performance of optimal cytoreduction, which is the surgical goal to be pursued, improving the results of adjuvant treatment. In early-stage cases, fertility-sparing surgery should be offered to patients who wish to conceive in the future.[5] The patient in the case presented already had offspring and there was no longer any reproductive desire.

Qualification in gynecologic oncology surgery is fundamental in the prognosis of ovarian cancer patients. Staging and prognosis of these early-stage and advanced tumors improve when these patients undergo treatment with surgical teams specialized in complex surgeries, including multivisceral resection, as performed in the patient described in this report.[6]

Ultrasound diagnosis of immature ovarian teratomas is difficult, since these tumors often present with heterogeneous characteristics, including solid areas and diffuse calcifications.[5] Abdominal pelvic ultrasonography is essential, since it enables the confirmation of tumor origin and evaluation of tumor characteristics, e.g. size, extension, contents and the presence of septum, among other findings. Furthermore, based on these parameters it is possible to infer whether the tumor is benign or malignant. CAT scans and MRIs have specific indications in dubious cases and for the evaluation of the presence of recurrence and metastases.[7]

The introduction of chemotherapy, usually reserved for G3 or even G2 tumors, has improved the survival rates in women with malignant germ cell ovarian tumors. The most widely used protocol is the BEP.[5] In the follow-up period, the patient had a suspicious lesion in the vaginal vault. However, resection of the lesion demonstrated a rare benign lesion---myxoid fusocellular tumor.

The mean survival of patients with immature ovarian teratoma is five years. Nevertheless, patients whose tumors were completely removed have a 5-year survival rate of about 94%, while the expected survival rate in those with partial resection (as the patient described here) is lower than 50%.[2]

CONCLUSION

Experience with the case report that described a survival rate above the mean survival time reported in the literature (17 years) after cytoreductive surgery, shows the benefit in patients with advanced-stage ovarian tumors, mainly in those where cytoreductive surgery is a determining factor for survival, under specialized care of gynecologic oncology surgical teams. In this case, the importance of other possible less measurable factors in the presented outcome is also highlighted, such as biological characteristics of the tumor (e.g. normal levels of AFP and β-HCG) and tumor response to chemotherapy carried out after suboptimal cytoreduction.

CONFLICTS OF INTEREST

None.

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