


Adenocarcinoma of intestinal type arising in mature cystic teratoma of ovary: A diagnostic dilemma

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Abstract

Adenocarcinomas of intestinal type arising in mature cystic teratoma of ovary (MCT) are extremely rare and remain a diagnostic dilemma because of its similarities with MCT. Serum tumor markers CEA and SCC and also MRI may help in the preoperative diagnosis. Pathologist experience helps in intraoperative diagnosis.

KEYWORDS

adenocarcinoma, dermoid cyst, diagnosis

1 | INTRODUCTION

Adenocarcinomas of intestinal type arising in mature cystic teratoma (MCT) of ovary are extremely rare. We present one case of a gastrointestinal transformation of a mature teratoma occurring in a 48-year-old woman. Diagnosis was made on the definitive pathological examination of the resected ovary.

The most common germ cell tumor of ovary is the mature cystic teratoma (MCT) which makes up 62% of all ovarian neoplasms in women younger than 40 years.¹ More than 80% of malignant transformations of teratomas are to squamous cell carcinomas. Malignant transformation to adenocarcinomas is very rare. To the best of our knowledge, only eleven cases of adenocarcinomas of intestinal type arising in mature cystic teratoma were reported in the literature. Malignant transformation occurring in MCT of the ovary is

rarely diagnosed preoperatively, and accurate intraoperative diagnosis may be difficult because of the rarity of this tumor.

2 | CASE PRESENTATION

A 48-year-old woman presented to the department of gynecology with complaints of a pelvic painful mass that evolved since 5 months. She was a premenopausal female, gravida 5, para 2 with a regular menstrual cycle.

Physical examination revealed a palpable solid pelvic mass extending to the umbilicus of approximately 20 cm in its greatest dimension.

Ultrasound scans revealed the presence of an approximately 18 cm multiloculated cystic mass of the right ovary. There were no abnormal findings of the uterus and left adnexae.

Serum electrolytes, liver function tests, and complete blood counts were normal. Serum tumor markers carbohydrate antigen 19-9 (CA199), carcinoembryonic antigen (CEA), CA 125, and α -fetoprotein (AFP) were not performed.

Exploratory laparoscopy revealed a cystic mass of the right ovary of approximately 20 cm. The uterus, other pelvic cavity, and the upper abdominal organs were all macroscopically normal. The cyst was accidentally ruptured when inserting the trocar. The patient underwent conversion to laparotomy. A right annexectomy was performed.

Intraoperative frozen section analysis confirmed the diagnosis of a dermoid cyst. Only a peritoneal washing was performed.

Following fixation and serial sectioning, the definitive histology was that of an adenocarcinoma of intestinal type arising in a mature cystic teratoma of the ovary.

A second operation was decided and a staging procedure was performed including a contralateral oophorectomy, hysterectomy, bilateral pelvic lymphadenectomy, omentectomy, appendectomy, and peritoneal washings.

All of these specimens were benign resulting in a completely resected FIGO stage IA, intestinal-type adenocarcinoma of the right ovary arising in mature cystic teratoma.

Postoperatively, the patient's CT scan and serum tumor markers were normal. The patient also had a colonoscopy and upper endoscopy. Both of them were within normal limits. Six cycles of adjuvant chemotherapy with paclitaxel and carboplatin were indicated.

3 | PATHOLOGIC FINDINGS

In gross examination, the surgical specimen consisted of a 17 cm multiloculated mucoid cystic mass containing a few foci of hair.

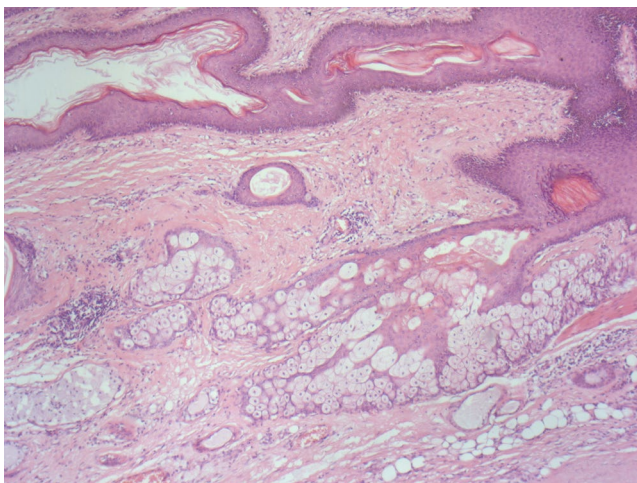


FIGURE 1 The dermoid cyst containing few foci of sebaceous glands, smooth muscle, and mature adipose tissue

Microscopically, the cystic surface was covered by a squamous or a cuboid to columnar epithelium devoid of atypia. The cystic wall included sebaceous glands, smooth muscle, and mature adipose tissue (Figure 1).

In some areas, the cyst showed carcinomatous cells arranged in glands of different shape and size. These glands were lined by cylindrical cells having enlarged nuclei containing a coarse chromatin and large nucleoli with some mitosis (Figures 2 and 3). They were surrounded by a fibrous stroma. The pathological diagnosis was an adenocarcinoma arising in a dermoid cyst.

Tumor cells showed immunohistochemistry (IHC) positivity of CK-20, CK-19, and ACE. Moreover, the IHC analysis showed negativity of CK-7, thyroglobulin, TTF1, and Pax 8. The mutation analysis of molecular alteration did not reveal a RAS gene mutation in the intestinal adenocarcinoma part.

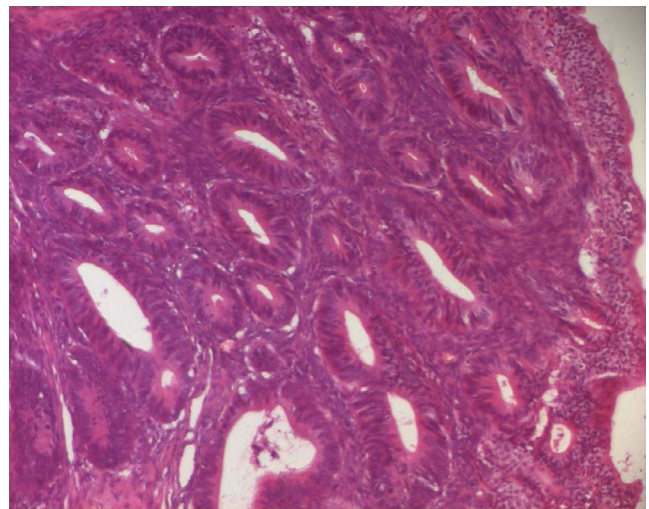


FIGURE 2 HE stain $\times 100$: carcinomatous cells arranged in glands of different size and shape

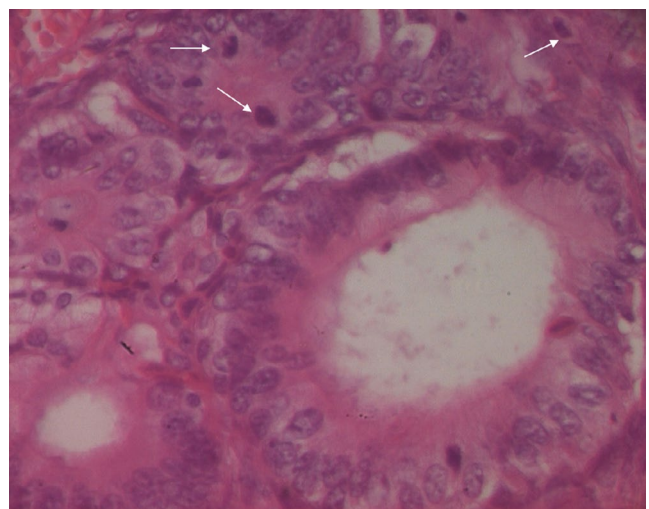


FIGURE 3 HE stain $\times 400$: atypical nuclei with mitosis (arrows)

4 | DISCUSSION

The median age at diagnosis of the published cases ranged from 13 to 77 years, with a mean age of 45 years. There were no specific symptoms of malignant transformation of an MCT, regardless of the type. The most common complaints were those of MCT which were abdominal pain, mass, or distention.²

Malignant transformation developing from a mature cystic teratoma is rarely diagnosed or even suspected preoperatively. Diagnosis of malignant transformation of MCT is generally made on the definitive pathological examination of the resected ovary. Symptoms and ultrasound scans are not specific. However, a fat-suppressed MRI may be helpful in the diagnosis of mature cystic teratomas even in tumors of <5 cm. Malignant transformation tends to have more solid components.³ Serum tumor markers carcinoembryonic antigen (CEA) and squamous cell carcinoma antigen (SCC)³ may also be predictive of a malignant transformation.

Carcinoembryonic antigen level was high in only one reported case,⁴ and CA 19-9 was elevated in six cases.⁵⁻⁹

According to Yamanaka Y et al,¹⁰ patients over 60 years old who have unilateral MCT with SCC value of at most 2.0 ng/mL and tumors at least 10 cm in size may be at high risk for malignant transformation.

Due to the rarity of the tumor, intraoperative histology may not identify the malignant transformation of MCT. In our case, there was discordance between intraoperative frozen section analysis and definitive histology. Careful observation and knowledge of limitations can be helpful in the intraoperative histology of frozen sections.¹¹

Intraoperative rupture of tumors has become more prevalent because of the increasing use of laparoscopic surgery. Two cases of intraoperative rupture of MCT with malignant transformation led to a peritoneal carcinomatosis associated with a locoregional relapse in one case.¹²

Adenocarcinomas of intestinal type arising in MCT of ovary are usually confined to one ovary (stage FIGO Ia). Other reported FIGO stages were Ic¹³ and IIc.⁴ Surgery is the mainstay of treatment. Even if there is no proof of the benefit of adjuvant chemotherapy for these tumors, it was administered for three reported cases.^{4,8,14} We decided to give adjuvant chemotherapy to our patient because of the risk of disease dissemination after the intraoperative rupture.

5 | CONCLUSION

Adenocarcinomas of intestinal type arising in MCT of ovary are extremely rare and remain a diagnostic dilemma because of its clinical and ultrasound scan similarities with MCT. Serum tumor markers CEA and SCC and also MRI may help in the preoperative diagnosis. Pathologist experience and knowledge of limitations help in improving the

intraoperative diagnosis. Surgery is the mainstay of treatment. These tumors are usually confined to one ovary (stage FIGO Ia). Adjuvant chemotherapy can be indicated in order to prolong survival.

CONFLICTS OF INTEREST

The authors have no conflicts of interest relevant to this article.

AUTHOR CONTRIBUTIONS

IB and WK: collected the data and wrote the paper. ABA, AB, RB, MH, FE, TT, IC, LBF, MM, SBA: collected the data and contributed to writing the manuscript.

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How to cite this article: Belaid I, Khechine W, Ben Abdelkader A, et al. Adenocarcinoma of intestinal type arising in mature cystic teratoma of ovary: A diagnostic dilemma. *Clin Case Rep.* 2020;8:644–647. <https://doi.org/10.1002/ccr3.2718>