CLINICAL CASE

Primary abdominal wall clear-cell carcinoma in postsurgical scar

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Received on July 17, 2017; accepted on August 8, 2017
Available online: February 1, 2018

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doi:10.24875/j.gamo.M18000121

Abstract: Introduction: Clear cell carcinoma on the abdominal wall is a rare pathology. Clinical case: It is presented the performance, diagnosis and treatment of a 64-year-old patient treated due to the presence of left ovarian adenocarcinoma with endometriotic deposits in the contralateral ovary that develops clear cell carcinoma on the abdominal wall. Discussion: The malignant transformation of the endometriotic tissue is rare and is generally associated with an ovaric disease. The diagnosis still remains a challenge. It is believed that the treatment is based on surgery. Conclusions: Owing to the low incidence of the malignant transformation of the endometrosis, it is relevant to notify the different cases arising. (creativecommons.org/licenses/by-nc-nd/4.0/).
INTRODUCTION

Abdominal wall clear-cell carcinoma is an infrequent diagnosis. Cases have been described caused by malignant transformation from ovarian endometriotic foci. We present a case of abdominal wall clear-cell carcinoma in a patient who had been intervened 3 years prior for a borderline ovarian seromucinous adenocarcinoma with endometriosis foci in the contralateral ovary.

CLINICAL CASE

This is the case of a 64-year old hypertensive and asthmatic woman presenting with postmenopausal metrorrhagia and who was studied by the Gynecology Department.

Vaginal examination revealed a 7-8 cm, barely mobile mass at the level of the pelvis minor.

Ultrasonography revealed an echogenic, irregular mass occupying the bottom of the recto-uterine pouch and compressing structures such as the bladder and rectum, at 12-cm distance from the anal margin, and that appeared to depend on the ovary, with a dimension of 13 x 8.7 cm, solid-cystic consistency, thick septum and papillae within, all this suspicious of ovarian adenocarcinoma.

A computed tomography (CT) scan identified an anteriorly located atrophic uterus, compressed between the bladder and a large, highly irregular cystic mass of approximately 12 x 9.5 x 13 cm, extending towards both parametria. This structure showed multiple images of solid papillae with increased uptake and thick septa within. All this was consistent with clinical suspicion of ovarian cystadenoma. The left superior margin of this lesion touched the sigmoid colon without a fat plane of separation, which drove to think of possible focal infiltration of the intestinal wall. No pathological-size adenopathies were appreciated in the retroperitoneal, mesenteric or pelvic lymph nodes, although small (0.6-0.7 mm) nodular images with increased uptake and that might be reactive were identified, although malignancy could not be ruled out.

Tumor markers, evaluated prior to surgery, were the following: carbohydrate antigen 19.9 (CA 19.9) with values higher than 700 (0-37 IU/mL), cancer antigen 125 (CA125) was 153.1 (0-35 IU/mL) and carcinoembryonic antigen (CEA) was 2.2 (0-3 IU/mL).

Endometrial biopsy was negative for carcinoma, and colonoscopy did not show rectal mucosa involvement.

In view of these findings, the patient was intervened by means of supra- and infra-umbilical laparotomy, and total hysterectomy plus double adnexectomy was practiced including the right ovarian mass, in addition to bilateral and para-aortic pelvic lymphadenectomy, omentectomy and appendectomy. Intraoperative anatomical pathology analysis indicated a right ovary borderline serous tumor. No infiltration of the sigmoid colon was observed, as previously suggested by the imaging tests.

At day 8 post-surgery, the patient underwent an emergency intervention owing to evisceration on the middle of the laparotomy wound, and new closure of the abdominal wall was performed.

Definitive anatomical pathology analysis described the different specimens, with epithelial inclusions without evidence of malignancy being found in the peritoneal tissue, i.e., epithelial inclusions lined with cuboidal or flattened epithelium, but without the architectural complexity or atypia of an ovarian neoplasm, and without stroma of desmoplastic appearance suggestive of infiltration, and benign inclusions of the endosalpingiosis type were thus considered. On the right adnexus, a borderline mixed-type (seromucinous) tumor with stromal infiltration foci and sparse, isolated images of lymphatic vessels invasion was identified. Most part of the neoplasm showed a papillary serous pattern, although, focally, it showed mucosecretory metaplasia (seromucinous mixed tumor). On the left adnexus, simple, endometriotic and siderophagic unspecific cysts were observed. The endometrium was hypotrophic without showing alterations, and neither did the cervix show signs of malignancy.

No tumor implants were found in the epiploic tissue, and adenopathies showed unspecific reactive changes. The cecal appendix had an atrophic appearance.

The patient was periodically followed by the medical oncology department and underwent yearly CT controls. She didn’t receive adjuvant therapy.

Three years after surgery, the patient developed a mass on the abdominal wall (Fig. 1). An imaging study with CT was performed (Fig. 2), which showed an heterogeneous mass...
at the umbilical and infra-umbilical level, located at the midline and with growth towards the left side, infiltrating the left anterior rectus abdominis muscle and, to a lesser extent, the right one and subcutaneous cellular tissue, extending up to the skin, with a size of 8 x 8.5 x 7.5 cm. It showed areas of lower attenuation within, which were suggestive of necrotic-cystic degenerative changes and with abundant peritumoral circulation in the subcutaneous cellular tissue. Smaller than 1 cm bilateral papillary nodules, consistent with metastases, were also appreciated.

A puncture of this mass was carried out, and anatomic pathology was consistent with infiltration by adenocarcinoma of solid and acinar cribriform pattern, with clear-cell predominance. Immunohistochemistry showed positive cytoke-ratin (CK) 7 and vimentin, and negative CK 20, CA 125, CA 19.9 and hormone receptors. Intestinal marker CDX2 was negative, as well as serous marker WT1. However, it showed positivity for nuclear transcription protein (TTF 1).

In view of this situation, chemotherapeutic treatment was started with 6 cycles of paclitaxel and carboplatin, with good tolerance. Subsequently, on the control CT, disease progression was appreciated (pulmonary, nodular, abdominal wall mass and probably hepatic). In the light of these findings, a new chemotherapeutic treatment was decided with bevacizumab and cyclophosphamide, with bad clinical tolerance and tumor progression and discontinuing the chemotherapeutic treatment was therefore finally decided, as well as to continue with domiciliary palliative care, with exitus subsequently being produced.

DISCUSSION

Abdominal wall-located clear-cell carcinoma can originate in peritoneum that has been trapped in the abdominal scar after laparotomy; therefore, it should be regarded as intra-abdominal carcinoma metastasis. Or it can be attributed to malignant transformation of an abdominal wall endometriotic focus.

Endometriotic tissue malignant transformation is very rare (it has an incidence of 0.7-1% in women with endometriosis and approximately 30 cases have been described in the literature) and generally it is associated with ovarian disease. Approximately 20% of cases occur at extragonadal zones such as the colon and vagina.

In 1925, Sampson established three criteria to define a neoplasm secondary to endometriosis: endometriosis should be closely associated with cancer, cancer histology should be consistent with endometrial tissue-origin, and no other lesion should be identified as primary neoplasm.

Few cases meet these three criteria. All abdominal wall cases that have been described had surgery with iatrogenic endometriotic diffusion.

In our case, the patient had no clinical signs or diagnosis consistent with endometriosis prior to surgery, although left adnexus subsequent anatomical pathology study revealed endometriotic cysts.

The transformation of endometriosis into abdominal wall carcinoma can invade all layers of the abdominal wall and grow up to 10 cm outside of it.

Most common extra-ovarian endometriosis-derived histological subtype is clear-cell adenocarcinoma (62%).

The diagnosis of this condition remains a challenge, since there are no symptoms or markers indicating the transformation of endometriosis into carcinoma. Disease-extension studies can detect endometriosis regions rapid growth and distant metastases, although the transformation into malignancy is suspected based on solid or mixed tumor behavior.

There is no effective treatment available for this type of condition, since there are no sufficient cases that enable the generation of a protocolized treatment. Some authors think that optimal treatment should be based on radical surgery with wide resection and subsequent association of carboplatin-based chemotherapeutic adjuvant treatment and radiotherapy, although there is no evidence of prognostic improvement.

Lymph node involvement might be correlated with poor prognosis.

Borderline seromucous tumors are characterized by a papillary architecture with mucinous epithelium that is similar to that of endocervical tumors. These tumors are associated with endometriosis.

Immunomarkers are essential for diagnosis in order to demonstrate the presence of endometrial stroma cells; the CD10 immunomarker, which is sensitive and specific in the diagnosis of extragonadal endometriosis, is used for this purpose.

CK 7, estrogen and progesterone-receptor-positive cells are found in these tumors, but they are negative for CK 20 and CDX2.

A CK 7-positive and CK 20-negative determination is associated with breast, endometrial, endocervical and ovarian tumors. CK 7 is a glandular adenocarcinoma marker and CDX2 marker would support a gastric tumor origin. TTF-1 is a non-mucinous and non-round-cell tumors specific marker.

In our case, the patient showed positivity for CK7 and TTF-1 and negativity for CK20 and CDX2, which would suggest gynecologic-origin disease in embryonic cells of endodermal origin.

Furthermore, if endometriotic cells’ nuclear expression is determined, p53 will be elevated in those cells with malignant transformation in comparison with benign endometriotic cells.

Endometriosis-derived ovarian tumors often occur as clear-cell carcinoma.

Skirnisdottir et al. describe the Napsin A marker, which is used in lung adenocarcinoma and is expressed in some thyroid, renal and endometrial carcinomas, as a useful marker in the diagnosis of ovarian-origin clear-cell adenocarcinoma.

CONCLUSIONS

Endometriotic disease, although highly infrequently, can undergo malignant transformation, and this should be taken into account when differential diagnosis is made in re-intervened patients with this underlying condition. Definitive diagnosis should be based on anatomic pathology study, either by tumor biopsy or fine-needle aspiration. Currently, owing to the low incidence of this transformation, there are no protocols available for its treatment, and the communication of different cases and implemented actions is therefore highly important.
REFERENCES