ANNALI ITALIANI DI CHIRURGIA

Digital Edition
e-publish on-line
ISSN 2239-253X
Direttore Nicola Picardi

Malignant endometriosis of the abdominal wall

Ann Ital Chir, Digital Edition 2017, 6
pii: S2239253X17026895 - April 13
free reading: www.annitalchir.com

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Malignant endometriosis of the abdominal wall

Endometriosis is a disease represented by the presence of extra uterine endometrial tissue. It is a rare condition, and malignant transformation is seldom. We report a case with clear cell adenocarcinoma oncogenesis on abdominal wall scar that appeared after years of a caesarian section. After diagnosis, surgical treatment was performed twice, due to the fact that the margins were infiltrated with tumor cells, with replacement of the defect with a polypropylene mesh. The patient was cured and discharged with a favorable prognostic. To the best of our knowledge, there are few reported cases of clear cell adenocarcinoma arising from abdominal wall endometriosis. It is a rare condition that appears mostly after abdominal surgical interventions that clinicians must be aware.

KEY WORDS: Abdominal wall, Endometriosis, Cesarean section, Clear cell adenocarcinoma, Malignant transformation

The authors are grateful for the financial support from "Juliu Hatieganu" University of Medicine and Pharmacy, 3rd Department of Surgery, Cluj-Napoca, Romania, internal grant 4945/14/08.03.2016

Introduction

Endometriosis is a condition where the extra uterine endometrial tissue is present. The ectopic functional with stroma and glands may be a constituent of the myometrium, although in many instances are found in other sites (Table I)[1,2]. Rarely, the abdominal wall can be affected especially along previous scars after hysterectomy, caesarean section, salpingectomy [1,3,4]. Usually influenced by cyclic hormones, endometriosis has a locally invasive benign character, but malign transformation is a rare risk[1]. In literature were reported a limited number of cases of malignant transformation like: clear cell carcinoma, endometrioid adenocarcinoma, serous adenocarcinoma, and carcino-sarcoma [4,5]. We report a case of abdominal wall endometriosis after Pfannenstiel incision, with malignization to clear cell adenocarcinoma post caesarian resection.

Table I - *Sites of endometriosis*

Common sites	Rare and remote sites
Ovaries	Umbilicus
Pelvic peritoneum	Abdominal scar
Pouch of Douglas	Episiotomy scar
Uterosacral ligaments	Lungs
Rectovaginal septum	Pleura
Sigmoid colon/Appendix	Ureter
Pelvic lymph nodes	Kidney
Fallopian tubes	Arms
	Nasal mucosa
	Legs

Pervenuto in Redazione Dicembre 2016. Accettato per la pubblicazione Febbraio 2017
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Case Report

A 43-year-old Caucasian woman, with no relevant medical history, was admitted in surgery department for suprapubic pain described as burning sensation, sensitivity at palpation of the abdomen in the mentioned area, which started seven months before. The patient also mentioned pain and swelling in the suprapubic area during menstruation. At 21-years-old she delivered a baby through caesarean section.

The physical examination revealed soft abdomen, participating to respiratory movements, suprapubic pain at both superficial and deep palpation, with no sign of bowel obstruction.

Abdominal ultrasound revealed a median suprapubic solid tumor that measured 59/38 mm with mixt structure (Fig. 1), consisting in a basal echogenic area surrounded by a transonic one (Fig. 2). Using the Doppler ultrasound mode the solid portion of the tumor was shown to be irrigated by blood vessels (Fig. 3). Serum levels of CA-125 were not elevated. Other blood tests were performed but all were in normal range. Taken together, endometriosis diagnosis was decided followed up by surgical removal. An intraparietal cystic nodule was found intraoperatively that measured 8/5 cm. The liquid was aspirated and sent to laboratory for bacteriological examination. Excision of the tumor was performed followed by subcutaneous drainage. The pathology report revealed on macroscopic evaluation a cyst with evacuated content, surrounded by brown areas and adjacent to them, a small white region with elastic consistency. Microscopically, examination revealed endometrial stroma along with hemorrhagic areas, siderophages and lymphocytes (Fig. 4). The area corresponding to the macroscopic white part of the tumor was represented by clear cell adenocarcinoma with tumor infiltrated margins (Fig. 5). Immunohistochemistry, A1/A3, CK7 were intense

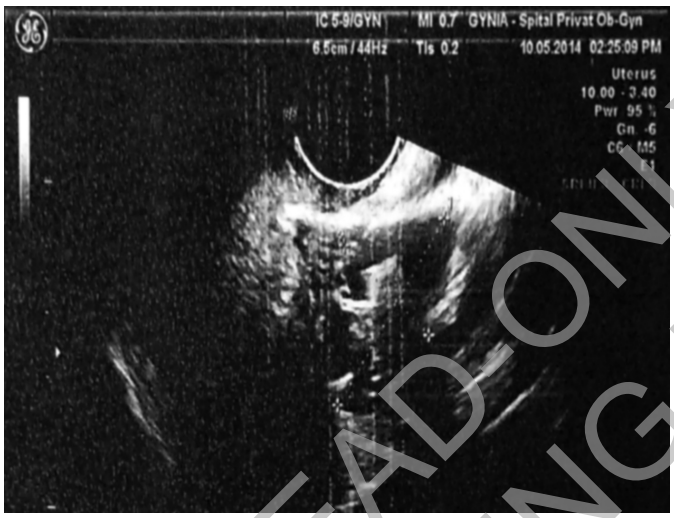

Fig. 1: Ultrasound image of the suprapubic solid tumor that measured 59/38 mm with mixt structure.

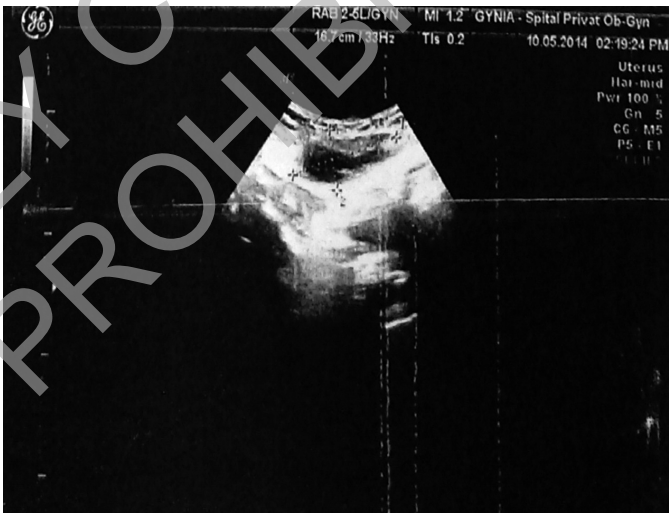

Fig. 3: Doppler ultrasound mode image of the solid portion of the tumor irrigated by blood vessels.

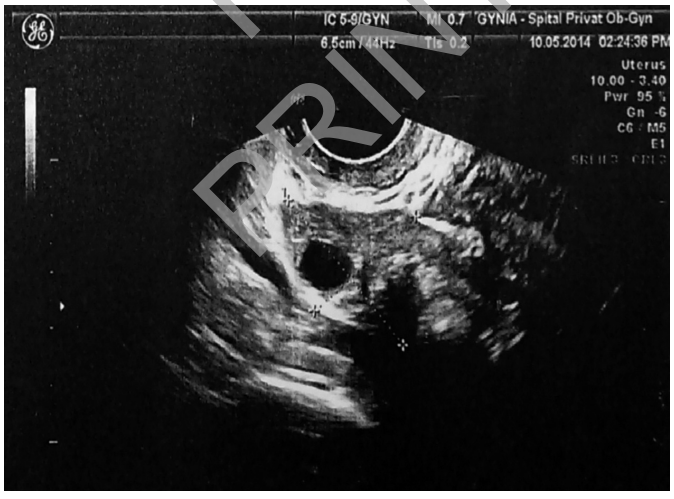

Fig. 2: Ultrasound image of a basal echogenic area surrounded by a transonic area.

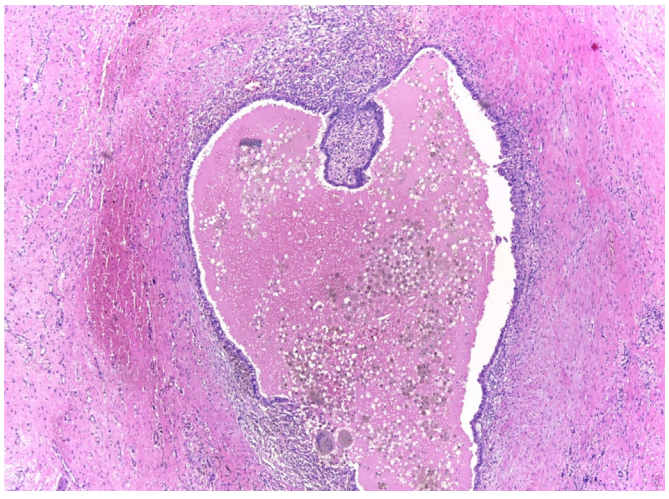

Fig. 4: Microscopic image of endometriosis.

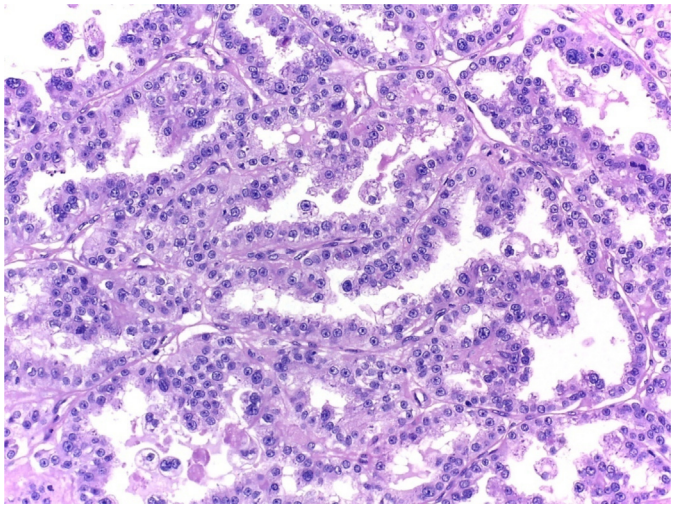

Fig. 5: Microscopic image of clar cell papillary adenocarcinoma.

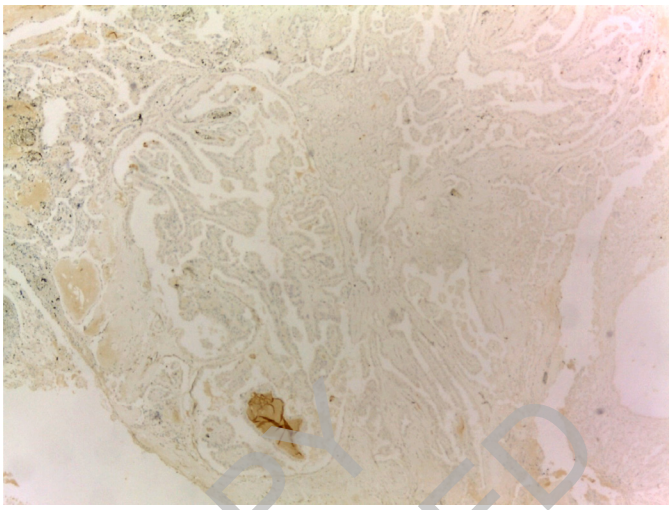

Fig. 8: Immunohistochemistry Wilms coloration 4x: negative.

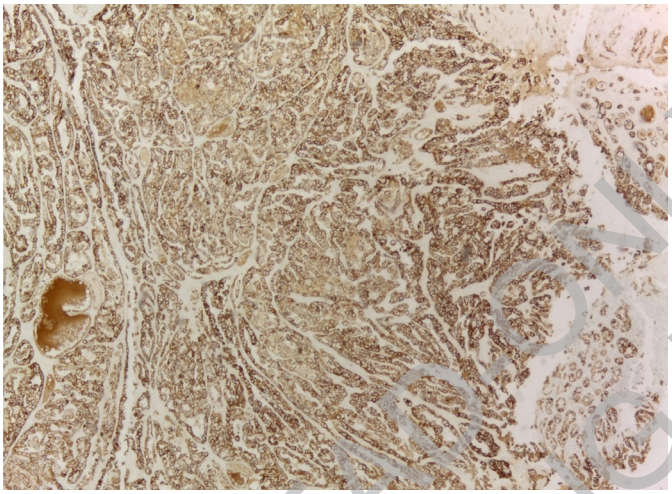

Fig. 6: Immunohistochemistry A1/A3, CK7: highly positive on tumor proliferation 4x.

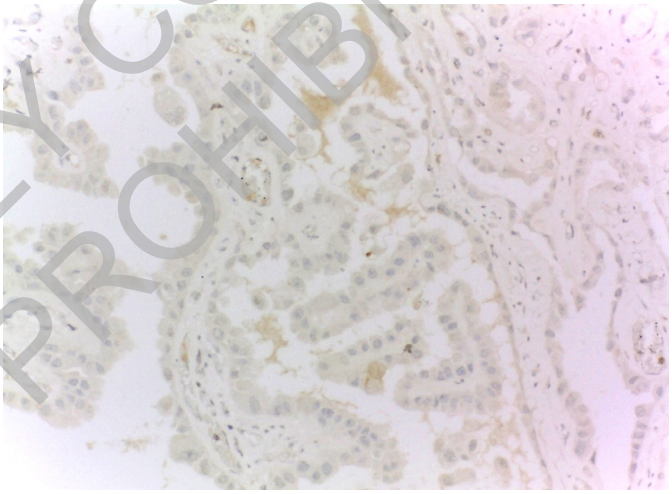

Fig. 9: Immunohistochemistry Wilms coloration 20x: negative.

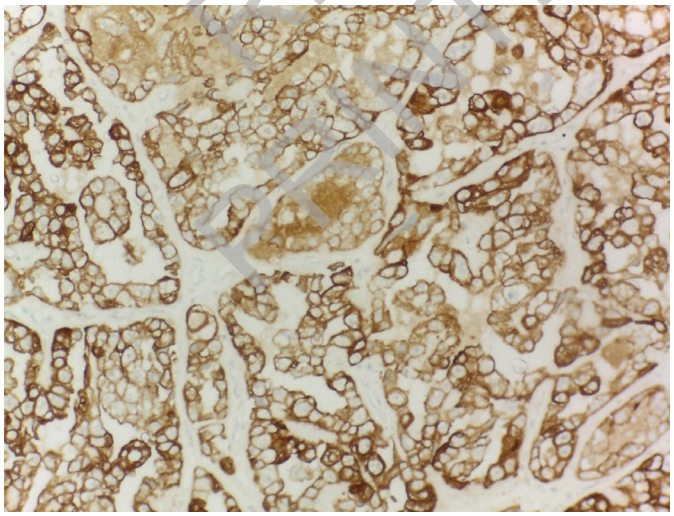

Fig. 7: Immunohistochemistry A1/A3, CK7: highly positive on tumor proliferation 20x.

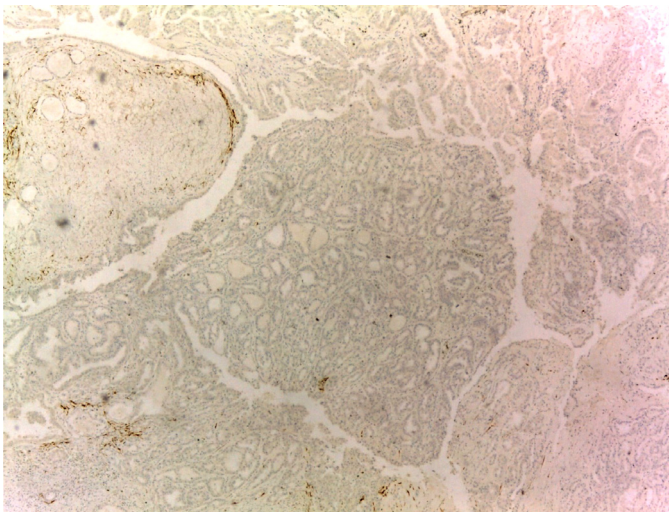

Fig. 10: Immunohistochemistry Calretinin coloration 4x: negative.

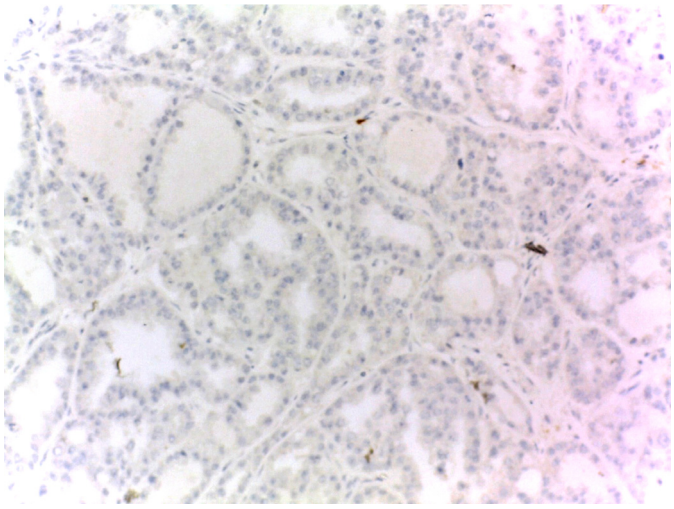

Fig 11: Immunohistochemistry Calretinin coloration 20 x: negative.

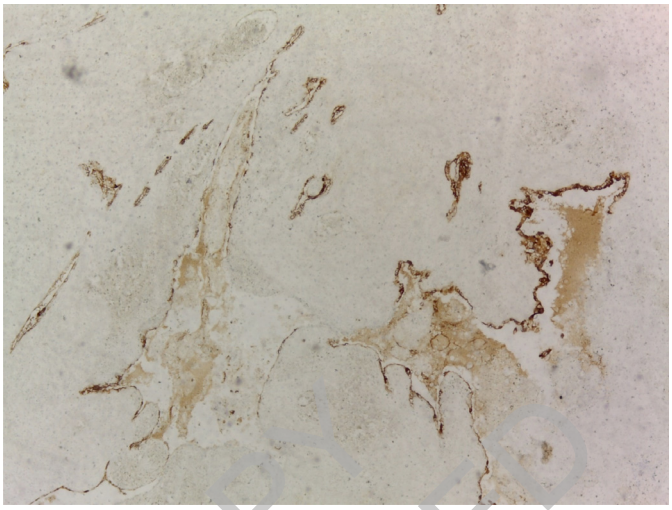

Fig. 14: Immunohistochemistry CA125 coloration 4x: focal positive on 5% of the tumor profliferation.

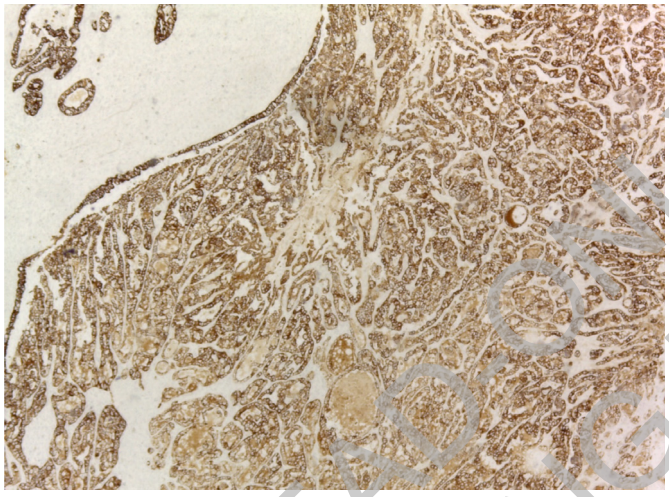

Fig. 12: Immunohistochemistry CK 5 coloration 4x positive on 50-60% of the tumor prolieration.

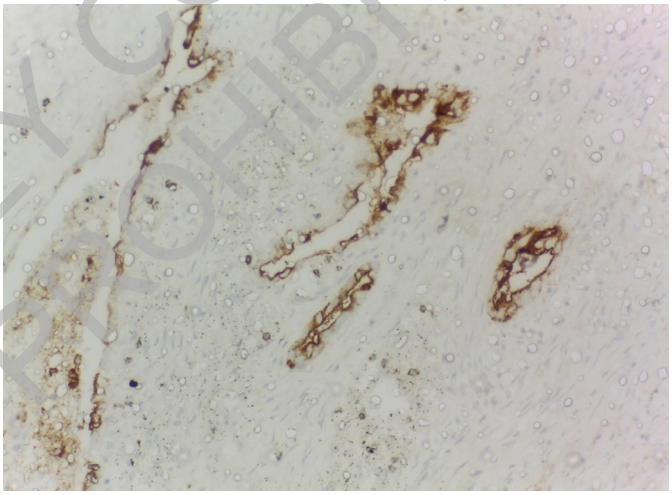

Fig. 15: Immunohistochemistry CA125 coloration 20x focal positive on 5% of the tumor profliferation.

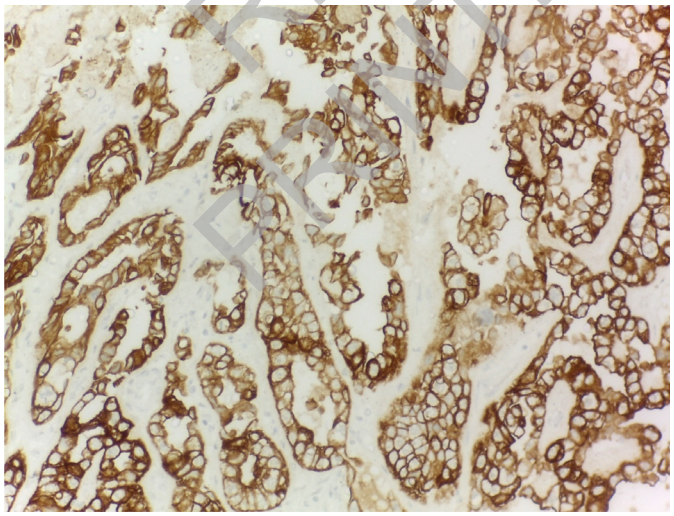

Fig.13: Immunohistochemistry CK 5 coloration 20x: positive on 50-60% of the tumor prolieration.

positive (Figs. 6, 7), Wilms (Figs. 8, 9) and Calretinin were negative (Figs. 10, 11), CK5 was positive on 50-60% on the tumor proliferation (Figs. 12, 13) and CA 125 was focal positive on 5% on the tumor proliferation (Figs. 14, 15). Concluding, the final diagnosis was endometriosis cyst and clear cell papillary adenocarcinoma.

Follow-up after 5 months by gynecological examination and diagnosis hysteroscopy showed no pathological aspects. The delay was caused by the patient's personal reasons. Due to the fact that the margins were tumor infiltrating, decision for surgical reintervention was taken. The operatory team decided to perform a wide excision of the anterior abdominal wall, incorporating the old scar. Also, it was decided the replacement of the abdominal wall defect with a polypropylene mesh with supraprotetic double drainage (Graur 2010). The following histology report revealed infiltrating lymphocytes, with no malign proliferation on none of the sections.

Under antibiotic with Ceftriaxone and Metronidazole, non-steroidal anti-inflammatory and pain management treatment, the patient recovered eventfully. The woman was discharge with a well general state, with no fever, hemodynamically stable and no signs of bowel obstruction. The patient had several follow-ups, at 6 months interval. The latest follow-up showed no signs of local relapse and the biological tests were within normal parameters.

Discussions

Incidence of this rare pathology is only 0.5-1.0%, appearing mostly adjacent to surgical scars, most frequent after caesarian sections with a latency of 16.75 years [3,5,7]. The endometriosis appearance probability is 0.03-1% with the possibility to malign transformation around 0.3-1% [8]. Frequently the sites of malignant transformation are the ovaries, rarely in other sites, for instance the rectovaginal septum, colon, vaginal wall and abdominal wall [8-10]. The pathogenesis of malignant transformation is not yet fully understood. Japanese Society of Clinical Oncology proposed a model consisting of three theories for the carcinogenesis. First one speculates that the endometriotic epithelium is the precursor of cancer in a similar way the normal surface of the ovary epithelium is for ovarian tumors. Second hypothesize that malign transformations resemblance tumorigenesis of endometrial neoplasia that is affected by the estrogens, which finally leads to endometrioid tumor [4]. The last, refers to the fact that the microenvironment has a severe impact on carcinogenetic process due to continuous exposure to oxidative stress and inflammation which enhance the genesis of malignant tumors defined by resistance [8].
In literature were described four criteria for the diagnosis of malignant changes in endometriosis. The first three, proposed by Sampson in 1925 included that [1] the tumor must present both benign and neoplastic cells [3], the tumor histology should be compatible with the origin of endometriosis and [4] the lack of other primary tumor sites. The last, theorized by Scott in 1953, was represented by the presence of cytology changes in architecture of the endometriotic glands along with atypical endometriosis changes. In our case, all four criteria were fulfilled [4,5,9].
Diagnosis must take in consideration surgical history and a comprehensive anamnesis. Firstly, a soft tissue ultrasound can be performed along with fine needle aspiration for further histology investigation. There are no disease specific markers, CA-125 may be slightly elevated in some cases. Anterior study revealed the fact that CRP serums levels were considerably increased in patients with endometriosis than in healthy ones, in contrast with the anti-Müllerian hormone levels that were indirectly proportional. Further investigations for reliable biomarkers for endometriosis and its subtypes of malignant transformation are conducted, for instance human epididymis protein 4 combined with CA-125 [8].
Characteristically for this pathology, local invasion is one of dissemination ways. However, there were described few cases of lymph nodes metastases [7,8,11]. Their location depends on the anatomic region of the primary tumor. If the tumor is located in the infraumbilical area then the inguinal chains will be the first station of metastases and if the tumor is in the supraumbilical area, axillary chains may be involved [8].
Regarding the treatment, in an absence of management guidelines there is not a standardized one[4]. In our case surgical resection was chosen due to benign tumor suspicion. After the histology report, which revealed the malignant transformation, wide abdominal resection was performed along with replacement of the defect with polypropylene mesh. Recurrences after treatment are atypical in these cases, but have been reported, with poor prognostic including evolution to exitus[11]. Chemotherapy and radiotherapy should be taken into consideration depending on the tumor histology, however, other literature data contest this affirmation [3,5,7,12].

Conclusions

Endometriosis appearing after caesarian section is thought to be caused by iatrogenic factors. Its malignant complications are rare with a pathogenetic mechanism still unknown and not fully understood. With a vast symptomatology, along with the lack of a specific diagnosis and treatment, malignant transformation of abdominal wall endometriosis remains a diagnosis of exclusion and a great challenge for the medical practice. Due to its complexity, further investigations in this direction are imperative to be made.

Riassunto

L'endometriosi è una patologia rappresentata dalla presenza di tessuto endometriale in sede extrauterina. Si tratta di una condizione rara che raramente evolve con trasformazione maligna.
Viene riferito un caso con oncogenesi di un adenocarcinoma a cellule chiare su cicatrice della parete addominale insorto anni dopo un parto cesareo. Dopo la diagnosi si è proceduto due volte al trattamento chirurgico poiché i margini della prima resezione erano infiltrati da cellule tumorali, e la ricostruzione del difetto è stata realizzata con una rete di polipropilene. Dopo la guarigione chirurgica la paziente è stata dimessa con prognosi favorevole.
Per quanto ci è noto vi sono poche segnalazioni bibliografiche di adenocarcinomi a cellule chiare insorti da una endometriosi della parete addominale. Si tratta di una

situazione rara, per lo più legata a precedenti accessi chirurgici addominali e di cui i clinici devono esser consapevoli.

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