Extragenital Malignant Mixed Mullerian Tumor in the Incisional Hernia – Primary Carcinosarcoma in the Abdominal Wall: Case Report

Marinko Žuvela1,2, Marjan Micev1,2, Milan Terzić2,3, Djordjije Šaranović1,2, Danijel Galun1,2, Miroslav Miličević1,2
1Clinic for Digestive Surgery, Clinical Center of Serbia, Belgrade, Serbia; 2University of Belgrade, School of Medicine, Belgrade, Serbia; 3Clinic of Gynecology and Obstetrics, Clinical Center of Serbia, Belgrade, Serbia

SUMMARY

Introduction This report presents a primary Mullerian carcinosarcoma localized in the incisional hernia i.e. anterior abdominal wall. There is no data in the literature about this localization of extragenital Mullerian carcinosarcoma.

Case Outline The patient had previous medical history of right-sided ovarian cystadenocarcinoma managed by hysterectomy, bilateral ovariectomy and chemotherapy. An incisional hernia occurred 1 year after the operation and Mullerian carcinosarcoma at the right border of the incisional hernia 16 years later. There was no tumor spreading into the abdominal cavity and pelvis. Full thickness of the abdominal wall resection and coexisting incisional hernia resulted in a large 25x20 cm abdominal wall defect managed by the modified components separation technique and implanting meshes.

Conclusion Major abdominal wall resection and abdominal wall reconstruction using the modified components separation technique reinforced with meshes could be one of possible solutions in the surgical treatment of primary malignant mixed Mullerian tumor localized in the abdominal wall.

Keywords: Müllerian carcinosarcoma; components separation technique; abdominal wall; incisional hernia

INTRODUCTION

Mullerian carcinosarcoma or malignant mixed Mullerian tumor (MMMT) is a very aggressive, malignant, female genital neoplasm composed of malignant epithelial and mesenchymal elements [1, 2]. Tumor stage, tumor type, grade of carcinomatous component and genital or extragenital tumor localization are prognostic factors influencing the overall survival [3, 4].

Primary extragenital Mullerian carcinosarcoma is a very rare disease and its localization in the anterior abdominal wall has not been reported in the literature. Due to different clinical presentations, small number of reported cases associated with limited surgical experience and absence of standardized treatment extragenital tumors have dismal prognosis. The aim of this report is to present a case of primary Mullerian tumor in the incisional hernia of the anterior abdominal wall.

CASE REPORT

A 70-year-old woman was admitted to the hospital with a huge palpable mass in an incisional abdominal wall hernia. Her medical history included midline laparotomy, total hysterectomy and bilateral adnexectomy, performed for a right ovarian tumor and myomatous uterus 18 years ago. Histopathological analysis demonstrated a well differentiated cystic endometrioid ovarian adenocarcinoma and intrauterine leiomyoma. Chemotherapy with Alceran was administered and the patient had a regular post-operative course. The asymptomatic midline incisional hernia occurred one year after the operation. A small tumor in the incisional hernia was diagnosed by clinical and ultrasound examination in January 2011. Abdominal CT was performed in May 2011 and confirmed a 62x50x38 cm right rectus muscle tumor next to the hernia sac. In October 2012 the patient was referred to a specialist for abdominal wall surgery. Clinical examination demonstrated a huge tumor in the incisional abdominal wall hernia. Three-phase contrast-enhanced MDCT detected a large inhomogeneous tumor mass in the anterior abdominal wall with infiltration of the rectus muscle, fascia and peritoneum. There were no signs of bowel infiltration. MDCT arterial phase demonstrated a tumor feeding artery arising from the right common iliac artery (Figure 1). Tumor markers: CA19-9 (353 U/mL) and CA 125 (287 U/mL) were significantly elevated. The patient was estimated as ASA II (diabetes type II and chronic diabetic nephropathy).

The patient was operated for the abdominal wall tumor and incisional hernia in December 2012. Intraoperative findings showed...
a sharply demarcated and partly incapsulated 12×10×10 cm abdominal wall tumor at the right border of the incisional hernia infiltrating the full thickness of the anterior abdominal wall. The skin and part of subcutaneous tissue were tumor-free. There was no tumor spreading in the abdominal cavity and pelvis, although a part of the omentum was attached to the tumor mass. Block resection of the full thickness of the right anterior abdominal wall with tumor (skin, subcutaneous tissue, 2/3 of the rectus muscle, parts of obliques and transverse muscles, and peritoneum), and partial omentectomy was performed (Figure 2). The size of the resected abdominal wall was 22×13 cm, however the size of abdominal wall defect was 25×20 cm due to the midline incisional hernia.

The abdominal wall defect was managed by the modified components separation technique as follows: a) dissection of the external from internal oblique muscle without skin dissection was performed at the right abdominal side; b) skin and subcutaneous tissue dissection from the anterior fascial layer in the direction from midline to the left spigelian line was followed by a left external oblique muscle aponeurosis longitudinal incision 1 cm lateral to the spigelian line and its dissection from the internal oblique muscle to the level of posterior axilar line; c) right internal oblique and transverse muscles were sutured to the left rectus muscle with its anterior and posterior sheaths using interrupted mattress non-resorbable sutures (suture line of the left and right myofascial flaps was localized at the level of the right mammillary line); d) herniorraphy was reinforced with 2 meshes in the onlay position, 1 nonresorbable (Parietene standard polypropylene mesh 30×10 cm, Covidien-Sofradim, Trevoux, France) and 1 resorbable (Vycril mesh 26×21 cm, Johnson-Johnson, Ethicon); e) nonresorbable mesh was sutured to the right external oblique muscle aponeurosis and lateral border of the left anterior rectus muscle sheath with running suture (the mesh protected myofascial flaps suture line and potential weakness in the region of the remaining right internal oblique muscle); f) resorbable mesh was sutured to the left external oblique muscle aponeurosis and lateral border of the left anterior rectus muscle sheath with running suture (the mesh protected potential weakness in the region of the left internal oblique muscle); g) skin and subcutaneous tissue were sutured over meshes (Figure 3).

Histopathology confirmed malignant mixed mesodermal Mullerian tumor (carcinosarcoma) composed of moderately differentiated endometroid adenocarcinoma (type I) with focal squamous differentiation and abundant homologues type sarcoma which was consistent with a low grade endometrial stromal sarcoma. Mitotic index of sarcomatous component and proliferation status based on the labeling index of Ki-67 protein were estimated as 9/10 HPF.
Tumor marker levels decreased after the operation (CA19-9 29 U/mL and Ca 59 U/ml). There were no post-operative complications and the patient was discharged 7 days after the operation. Chemotherapy with paclitaxel (Taxol) and carboplatin (CBDCA) was given. There was no tumor or incisional hernia recurrence during the 7-month follow-up period.

DISCUSSION

Malignant mixed Müllerian tumor (MMMT), also called malignant mixed mesodermal tumor or Mullerian carcinosarcoma, occurs in less than 1% of elderly menopausal females. This complex tumor usually occurs in the uterus, rather than in the ovaries, fallopian tubes and the upper portion of the vagina, and consists of carcinomatous and sarcomatous components [4].

According to the sarcomatous components, MMMT is divided into homologous type arising from tissues which are present in the uterus (endometrium and myometrium) and heterologous type arising from tissues absent in the uterus (bone, cartilage and skeletal muscle) [1, 4]. Histogenesis of MMMT might be closely related to the so-called secondary Mullerian system, composed of the pelvic and lower abdominal mesothelium and the subjacent mesenchyme. MMMT can be associated with extragenital endometriosis, synchronous or metachronous gynecological malignancies, colon cancer and primary peritoneal serous carcinoma [5, 6, 7]. There are reported cases of MMMT occurring after tamoxifen therapy in breast cancer [8]. The most common sites of metastasis are lungs and the abdominal cavity.

Extragenital primary Mullerian carcinosarcoma is extremely rare. Ober and Black [9] in 1955 reported the first extragenital MMMT, however there have been less than 50 reports in the English literature about this entity. The majority of reported extragenital MMMT are located in the peritoneum and almost half of cases in the pelvic peritoneum [6, 10, 11, 12]. There are only few reports of primary...
extragenital Mullerian tumor arising from the mesentery, omentum, spleen, urinary bladder, and retroperitoneum [3, 13-17].

This report presents a primary MMMT localized in the incisional hernia i.e. anterior abdominal wall. There is no data in the literature about this localization of extra-genital MMMT. The patient had previous medical history of right-sided ovarian cystadenocarcinoma managed by hysterectomy, bilateral ovariectomy and chemotherapy. An incisional hernia occurred 1 year after the operation and MMMT at the right border of the incisional hernia 16 years later. There was no clear evidence of a histological relationship between the ovarian neoplasm and extra-genital MMMT in the abdominal wall. Similar to this case, Garde et al. [18] reported extragenital MMMT occurrence after operation for ovarian malignancy, while Huang et al. [3] and Arora et al. [6] reported synchronous extragenital MMMT and ovarian carcinoma. In the present patient, primary extragenital MMMT involved all myofascial layers of the right anterior abdominal wall including parietal peritoneum and a part of the major omentum, while skin and subcutaneous tissue were tumor-free. It should be noted that the parietal peritoneum and a part of the adhered major omentum were involved only in the region of the abdominal wall tumor. There was no tumor spreading into the abdominal cavity and pelvis, and there was no peritoneal dissemination or ascites. The largest tumor in-growth was at the myofascial layers of the abdominal wall and not into the abdominal cavity.

It could be assumed that the development of the incisional hernia in the abdominal wall after the first operation might be related to implantation of previous undetected or unrecognized peritoneal endometriosis. This assumption is based on the morphological aspect and strong diffuse CD10 immunoexpression of abundant sarcomatous component of MMMT consistent with a low grade stromal sarcoma. Not surprisingly, one could also consider very late recurrence of previously operated ovarian carcinoma [19]. However, on detailed revision of histology samples of the well differentiated ovarian endometrioid carcinoma neither elements of associated stromal proliferation nor previous endometriosis or endosalpingiosis in resection specimen could be detected. Furthermore, one can speculate the possibility of metastatic dormant carcinoma cells stimulating the adjacent peritoneal stroma to develop the mesenchymal component of the tumor and in-growth into the abdominal wall, according to the so-called “combination” histogenetic theory of these tumors [20]. The metaplastic conversion of poorly differentiated carcinomatous components to sarcomatous differentiated

Figure 4. Histological examination (a) biphasic malignant neoplasm with coexistent carcinomatous and sarcomatous areas (H&E, original magnification 5×), (b) close admixture of both cell populations (H&E, original magnification 20×), (c) immunohistochemical stainings demonstrated vimentin immunoexpression (original magnification 20×) in sarcomatous as well as in endometrioid carcinomatous components and (d) strong CD10 immunoexpression (original magnification 20×) predominantly in the mesenchymal portion of the tumor.
areas is highly unlikely as both malignant components are not dominantly poorly differentiated. In the absence of evidence to support such possibilities, the strong Mullerian metaplastic potential of the inserted peritoneum in the incisional hernia could give rise to de novo MMMT.

Full thickness of the abdominal wall resection and coexisting incisional hernia resulted in a large 25×20 cm abdominal wall defect managed by the modified components separation technique and by implanting meshes. The abdominal wall reconstruction was based on the Ramirez “rectus-sharing” modification of the components separation technique for the management of abdominal wall defects following transverse rectus abdominus myocutaneous (TRAM) flap breast reconstruction [21, 22]. Unlike the Ramirez modified technique for TRAM a flap of external oblique was released from the internal oblique muscle at the side of the resected abdominal wall. Releasing the left and right rectus-internal-transverse muscles complex achieved myofascial flaps that were sutured at the level of the right mamilary line. Myofascial flaps movement in this case was much greater than in the Ramirez “rectus-sharing” modification. In order to achieve the best results in the abdominal wall reconstruction (obtained by combination of components separation technique and mesh hernioplasty) [23, 24] we reinforced our herniorrhaphy with 2 meshes: a nonresorbable mesh which protected the right internal oblique/transverse muscle and left rectus muscle suture line, as well as the area of the remaining right internal oblique muscle (parts of the right internal oblique and transverse muscle were resected with the tumor); and a resorbable mesh which protected the released internal oblique muscle at the left side. Using autologous tissue in combination with meshes a dynamic abdominal wall support was achieved.

Chemotherapy with Taxol/CBDCA protocol was administered and six months after the operation there are no signs of the disease and incisional hernia recurrence.

Surgical experience in the management of extragenital MMMT is limited due to the small number of reported cases and the fact that the majority of cases have been in the advanced tumor stage. The standardization of surgical treatment for extragenital MMMT is not possible and therapy should be adapted to each individual patient. In all patients with extragenital MMMT chemotherapy should be applied.

In conclusion, this is the first reported case of MMMT in the abdominal wall. Major abdominal wall resection and abdominal wall reconstruction using modified components separation technique reinforced with meshes could be one of possible solutions in the surgical treatment of primary MMMT localized in the abdominal wall.

ACKNOWLEDGMENT
This work has been supported by the grant No RS 41030 of the Ministry of Education, Science and Technological Development of the Republic of Serbia.

REFERENCES
Екстрагенитални малиги мешовити Милеров тумор у инцизионој хернији – 
примарни карциносарком у трбушном зиду: приказ болесника

Маринко Жувела1,2, Марјан Мицев1,2, Милан Терзић2,3, Ђорђије Шарановић1,2, Данијел Галун1,2, Мирослав Милићевић1,2

1Клиника за дигестивну хирургију, Клинички центар Србије, Београд, Србија;
2Универзитет у Београду, Медицински факултет, Београд, Србија;
3Клиника за гинекологију и акушерство, Клинички центар Србије, Београд, Србија

КРATAK САДРЖАЈ

Увод Овај приказ представља примарни Милеров (Müller) карциносарком локализован у инцизионој хернији предњег трбушног зида. У литератури нема података о овој локализацији екстрагениталног Милеровог карциносаркома. 

Приказ болесника Болесници је претходно оперисана због цистаденосаркома на десном јајнику, када су примењене хистеректомија, обострана оваријектомија и постоперацио на хемитерапија. Инцизиони хернија се јавила годину дана после операције, а Милеров карциносарком у инцизионој хернији се развио 16 година касније. Није било ширења тумора у трбушну дупљу, нити у малу карлицу. Комплетна ресекција тумором захваћеног трбушног зида и инцизионог кила оставише су оштећен предњи трбушни зид величине 25 x 20 см који је решен модификованим техником компонентне сепарације и имплантацијом мреже. 

Закључак Поплутно одстрањивање трбушног зида захваћеног тумором с реконструкцијом применом модификоване технике компонентне сепарације уз уградњу мреже може бити једна од опција хируршког лечења болесника с Милеровим карциносарком локализованом у трбушном зиду. 

Кључне речи: Милеров карциносарком; техника компонентне сепарације; трбушни зид; инцизионе хернија

Примљен • Received: 12/06/2014
Прихваћен • Accepted: 22/08/2014