CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 41 (2017) 323–326

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Extragenital malignant mixed mesodermal tumor: A case report

Mauro Del Papa [a], Gabriele D'Amata [a,*], Fulvio Manzi [a], Luca Musmeci [a], Marco Crovaro [a], Carlo Buonocore [a], Gaetano Florio [a], Andrea Giannetti [b]

[a] Department of General Surgery, Ospedale "Leopoldo Parodi Delfino", Colleferro, Italy
[b] Department of Pathology, Ospedale "Leopoldo Parodi Delfino", Colleferro, Italy

ARTICLE INFO

Article history:
Received 2 November 2017
Accepted 5 November 2017
Available online 9 November 2017

Keywords:
Carcinosarcoma
Mixed mesodermal tumor
Primary peritoneal carcinosarcoma
Malignant mixed mullerian tumor
Case report

ABSTRACT

INTRODUCTION: Primary malignant mixed mesodermal tumor (MMMt, also called malignant mixed Mullerian tumor and designated in the WHO classification of female genital tract neoplasms as carcinosarcoma) is an infrequent tumor that develops usually in the uterus and more rarely in the ovary. Extragenital tumor, including primary peritoneal MMMt, is an extremely rare and aggressive neoplasm with only few case reported in the literature.
PRESENTATION OF CASE: We report a case of a 70-year's old female who presented with nausea and abdominal discomfort for 6 months. Workup revealed an abdominal mass. Patient was treated with surgical removal in a general hospital.
DISCUSSION: Most peritoneal carcinosarcomas originate in the pelvic peritoneum, followed by decreasing frequency in the serosal surface of the colon, retroperitoneum, anterolateral abdominal peritoneum, and omentum. Surgical excision is the most effective treatment in carcinosarcomas. A complete cytoreduction, with resection of cancer to a status of no evidence of disease by the surgeon's unaided eye should be attempted.
CONCLUSION: Owing to the rarity of the disease, limited data regarding the management of peritoneal MMMT exists. Recommendations for the treatment of MMMT are based on individual cases only. In our case, the patient is alive with a follow-up of 15 months and she did not receive any cycle of chemotherapy.

1. Introduction

Primary malignant mixed mesodermal tumor (MMMt, also called malignant mixed Mullerian tumor and designated in the WHO classification of female genital tract neoplasms as carcinosarcoma) is an infrequent tumor that develops usually in the uterus and more rarely in the ovary. Extragenital tumor, including primary peritoneal MMMt, is an extremely rare and aggressive neoplasm with only few case reported in the literature [1]. The neoplastic elements of extragenital MMMT presumably arise directly from the mesothelium or submesothelial stroma and hence parallel the biphasic pattern of the genital (uterine or ovarian) counterpart. Since the first report in 1955 by Ober and Black [2], to our knowledge there have been only 30 well documented reports of extragenital malignant mixed Müllerian tumors [3]. Pelvic peritoneum seems to be the most common site for extragenital MMMTs. Extragenital MMMTs have also been shown to arise in other sites such as the serosal surface of the colon, retroperitoneum, cul-de-sac, rectal peritoneum, anterolateral abdominal peritoneum, diaphragm peritoneum, and omentum [4,5]. We report a case of a 70-year's old female who presented with nausea and abdominal discomfort for 6 months. Workup revealed an abdominal mass. Patient was treated with surgical removal. The following case has been reported in line with the SCARE criteria [6].

2. Case presentation

A 70-year's old woman presented for episodes of abdominal pain and nausea for 3 months. The patient's body mass index was 25 kg/m^2. Her past medical history was unremarkable. Abdominal examination revealed a large palpable, relatively mobile, non-tender mass in the right flank and mesogastrium. She had lost 4 kg of weight in 3 months. Computer tomography of her abdomen showed an abnormal mass with areas of necrosis below the transverse mesocolon, measuring approximately 17 cm × 11,5 cm × 9.5 cm (Fig. 1). The mass was adherent to the right rectum muscle and was strictly involving some loops of the small intestine. The patient underwent subsequent exploratory midline laparotomy. The mass was resected en bloc with the small intestine loops involved and an ileocolic anastomosis was performed (Fig. 2). The rest of the abdominal cavity, including ovaries,

* Corresponding author at: Ospedale "Leopoldo Parodi Delfino", Piazza Aldo Moro 1, 00034 Colleferro, RM, Italy.
E-mail address: gabridamata@gmail.com (G. D'Amata).

https://doi.org/10.1016/j.ijscr.2017.11.003

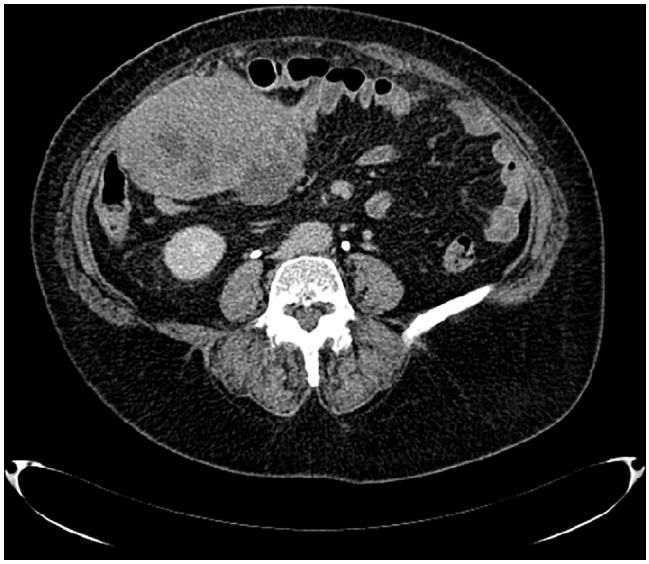

Fig. 1. Computer tomography of her abdomen showed an abnormal mass with areas of necrosis below the transverse mesocolon, measuring approximately 17 cm × 11,5 cm × 9.5 cm.

fallopian tubes and uterine corpus was normal. The postoperative course was unremarkable. The patient did not receive any cycle of chemotherapy and was alive with a follow up of 15 months. The histopathology revealed findings consistent with a malignant mixed mesodermal tumor, measuring cm 12. The radial, proximal and distal margins were negative. No metastasis in 2 lymph nodes was noted. Immunohistochemical analysis was performed to identify the immune profile of the tumor cells. Monoclonal mouse antihuman antibodies against cytokeratin (clone AE1/AE3), Calretinin, C-kit/Cd117, desmin (clone D33), miogenin (clone F5D) were used. The tumor cells arranged in a solid and vague papillary pattern (presumably epithelial element) showed strong reactivity for epithelial markers (cytokeratin and epithelial membrane antigen). Alternatively, the neoplastic cells arranged singly (presumably mesenchymal element) showed strong reactivity for miogenin and desmin. These cells (presumably sarcomatous element) showed negative reactivity for the epithelial markers (cytokeratin and epithelial membrane antigen). This combined morphology was of a carcinosarcoma, with aspects of heterologous osteocondroblastic and rabdomiosarcomatous differentiation (Fig. 3).

3. Discussion

Carcinosarcoma comprises coexisting carcinoma and sarcoma. Based on pathological diagnosis, 2 or more malignant components are required for carcinoma diagnosis. The carcinoma tests positive for epithelial tissue markers cytokeratin (such as cytokeratins 7,8,18,19, and pan-cytokeratin AE1/AE3) and the sarcoma component is positive for mesenchymal tissue marker vimentin, desmin and miogenine. Primary peritoneal neoplasms composed of both malignant epithelial and stromal elements have been referred to as extragenital malignant mixed mesodermal tumors [7], malignant mixed mullerian tumors [8], carcinosarcomas [9] and mixed tumors of mullerian type [10].

The mechanism of carcinosarcoma in uncleared. The peritoneal surfaces are host to a range of benign and malignant lesions commonly encountered in the müllerian duct derivatives of the female

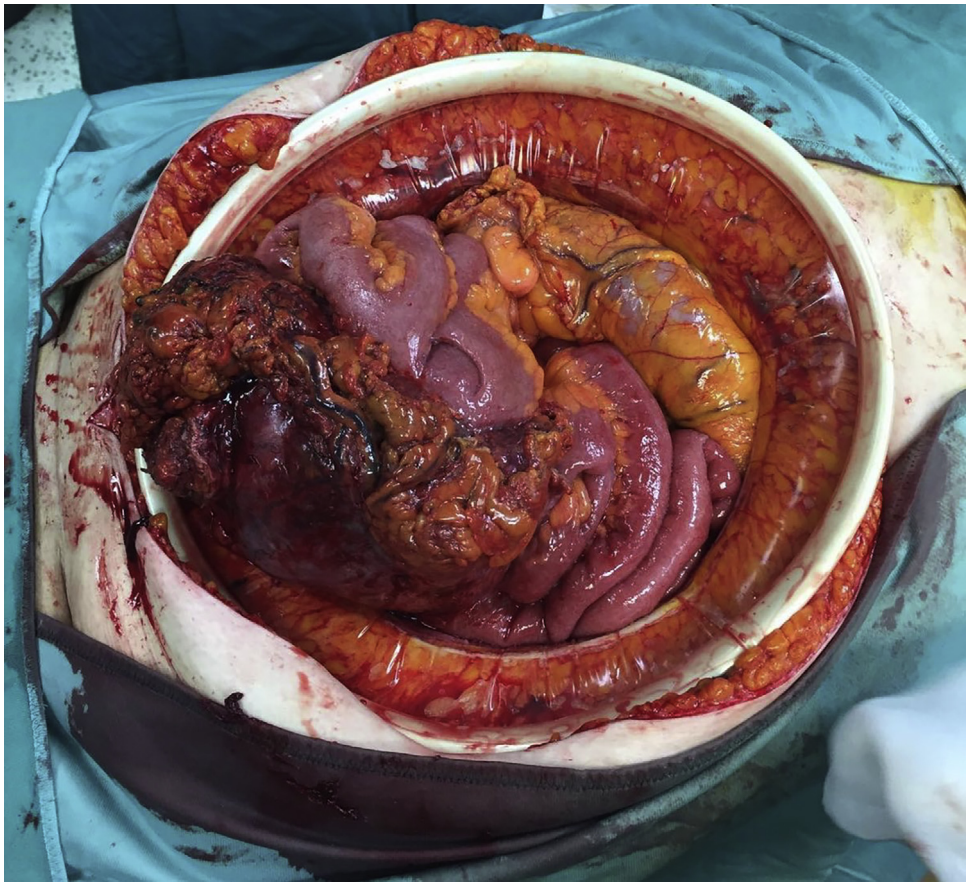

Fig. 2. Intraoperative finding: en bloc resection of the mass with small intestine loops involved.