

A Rare Case of Mesenteric Carcinosarcoma

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Abstract

Carcinosarcoma is an aggressive malignant neoplasm of the female genital tract that is biphasic and composed of carcinomatous and sarcomatous components. Carcinosarcomas that arise outside of the genital tract are extremely rare and there have been only few reported cases in the clinical literature.

We report the case of an 85 years old patient who was presented with for weight loss, constipation, enlarged abdomen, bilious vomiting, and selective loss of appetite for meat for 1 month.

The CT examination revealed a tumor mass in the mesentery, extended to its root, with heterogeneous structure and iodophilia. A biopsy was performed, and the histopathological result was that the tumor was a carcinosarcoma.

Keywords: Mesenteric Tumor; Carcinosarcoma; MMMT; CT

Abbreviations

CT: computed tomography; MMMT: Malignant Mixed Mesodermal Tumor

Introduction

The small bowel mesentery is a fan-shaped fold of peritoneum that suspends the small intestine from the posterior abdominal wall [1,2]. The two layers of peritoneal reflection forming the mesentery contain a variable amount of fat through which run the major arteries, veins, and lymphatics of the small intestine [1,2].

Primary tumors arising in the mesentery are relatively rare [1-4]. On the other hand, secondary involvement of the small bowel mesentery is frequent, and most of the masses are malignant at histopathologic examination. [1,2,4]. Patients with mesenteric neoplasms usually present with nonspecific symptoms of abdominal pain, weight loss, a palpable abdominal mass, or diarrhea [1,2].

CT plays an essential role in identifying the precise localization of these neoplasms, as well as to highlight belonging to a specific structure and, thus, to guide the treatment of the patient.

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Carcinosarcoma is an aggressive malignant neoplasm of the female genital tract that is biphasic and composed of carcinomatous and sarcomatous components [5].

Carcinosarcomas that arise outside of the genital tract are extremely rare and there have been only 25 reported cases in the clinical literature [6-8]. These cases include tumors located in the visceral peritoneum of the cecum and rectosigmoid colon, parietal peritoneum, and retroperitoneum [6,7].

The extragenital locations are part of the so-called secondary Mullerian system, consisting of the mesothelium and subjacent mesenchyma of the pelvis and lower abdomen [9]. The secondary Mullerian system originates from coelomic epithelium, as the primary Mullerian system.

Primary peritoneal neoplasms composed of both malignant epithelial and stromal elements have variously been referred to as extragenital malignant mixed mesodermal tumors [10], malignant mixed mullerian tumors [11], carcinosarcomas [12] and mixed tumors of mullerian type [7,13,14]. This lack of consistent terminology reflects both the rarity of these lesions, mostly described in single case reports, as well as the uncertainties concerning the histogenesis [7,13].

Since the first report in 1955 by Ober and Black [15], to our knowledge there have been only 30 well documented reports of extragenital malignant mixed Müllerian tumors [16]. Pelvic peritoneum seems to be the most common site for extragenital MMMTs [7]. 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 [17].

Case Report

We present the case of a female patient, aged 86 years, with known hypertension, who was hospitalized for weight loss, about 20 kg last year, and sudden constipation for two months ago. One month before admission, accused enlarged of the abdomen, bilious vomiting, with selective loss of appetite for meat.

The clinical exam reveals a mediocre general condition, edemas, non-palpable lymph node, hypotonic and hypertrophic muscular system; also, it was noticed abdominal volume increase with ascites fluid, and slow intestinal transit.

The anamnesis, corroborated with the clinical data, raises the suspicion of peritoneal carcinomatosis. An evacuation paracentesis was performed, which revealed increased global cellularity (frequent 30% cremation), frequent lymphocytes, and frequent cells suggesting neoplastic cells.

The ultrasound examination revealed ascites fluid in moderate amount, and an hypoechoic abdomino-pelvic tumor with irregular shape, irregular contour, about 4.4 cm in antero-posterior diameter, and 10 cm in transverse diameter, with no Doppler signal, without being able to tell the true origin.

Therefore, an abdominal CT was performed, which revealed a tumor mass located at the mesenter, extended to its root level, with maximum axial diameters of 15/11 cm, with heterogeneous structure and iodophilia, which includes intestinal loops, without causing stenosis.



Figure 1: Abdominal CT - native examination: large mass on the mesentery.



Figure 2: Abdominal CT - arterial phase: heterogeneous important iodophilia of the tumor mass on the mesentery.

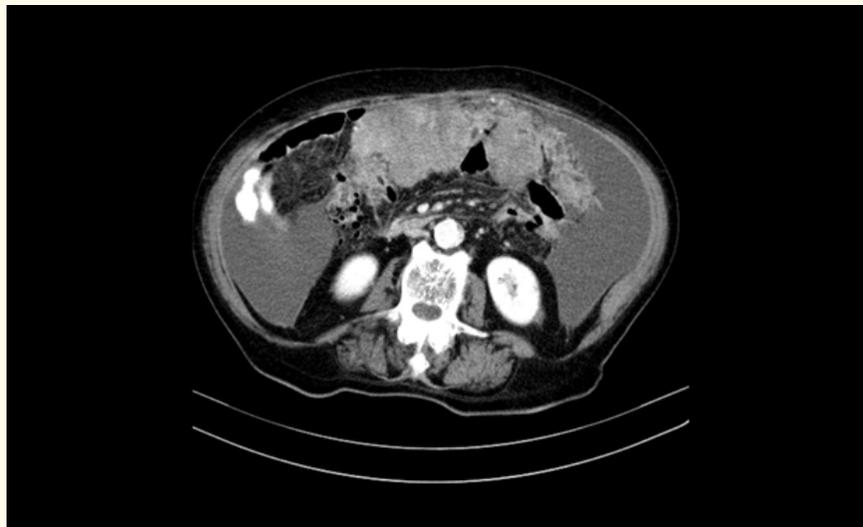


Figure 3: Abdominal CT - venous phase: heterogeneous important iodophilia of the tumor mass on the mesentery.

Also, there was found mesenteric adenopathies, up to 0.7 cm, and ascites fluid in medium amount.

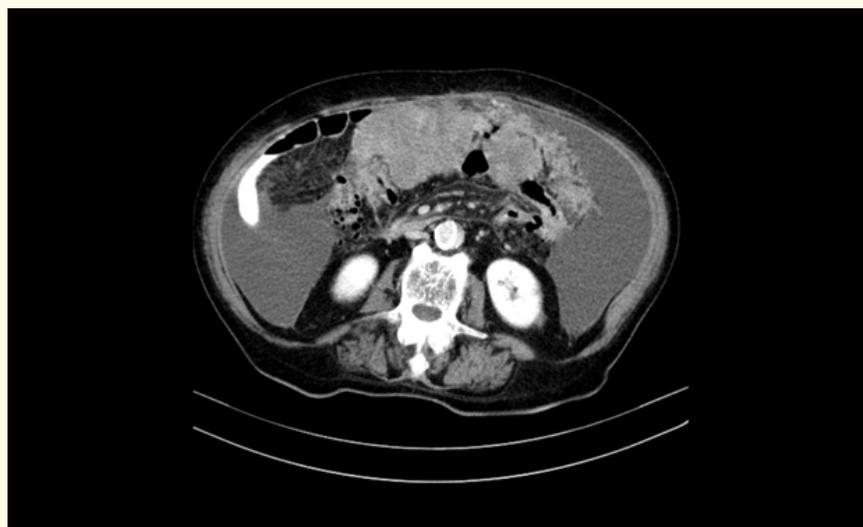


Figure 4: Abdominal CT - parenchymal phase: ascites in moderate amount and adenopathies.

Evacuation paracentesis, symptomatic treatment, and maintenance of transit was made, under which evolution is was favorable, with maintaining a stable weight.

There is an oncological and surgical consultation that recommend for diagnostic biopsy and laparoscopic assessment.

An exploratory laparoscopy with tumor biopsy was performed, with favorable postoperative evolution.

The patient was discharged in good general condition, painless abdomen, without signs of peritoneal irritation, and intestinal transit present.

The result of biopsy was fragment tissues with massive tumor infiltration, suggestive of a carcinosarcoma (having the chondrosarcoma-like component), necrotic.

Discussion

Early diagnosis of mesenteric small bowel tumors is a diagnostic challenge for both clinicians and radiologists because the majority of patients present with nonspecific symptoms [18], and because of its deep location, the mesenteric small bowel is difficult to investigate. Therefore, the diagnosis is often delayed [19].

In their review of a large series of patients with mesenteric pathology seen at CT, Whitley, *et al.* [20] found 101 cases of mesenteric neoplasms, but only one was a primary mesenteric tumor. Most primary lesions are mesenchymal in origin, and the majority was histologically benign [1].

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 [7].

Carcinosarcoma or malignant mixed mullerian tumor (MMMT) is an aggressive malignant neoplasm that shows both carcinomatous and sarcomatous components, and therefore can arise in almost every organ in the body [21]. Most patients die within one year, with median postoperative survival time of 14 months, and a range of 7 days-73 months [5,11].

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 [7].

Extragenital primary peritoneal MMMTs are very rare and only few cases have been reported in literature [22].

They are highly aggressive neoplasms that occur in elderly postmenopausal women [23], like in our case.

The mechanism of carcinosarcoma is unclear. Several theories have emerged in the attempt to explain the biphasic appearance of the tumor [24]. Histogenetically, primary peritoneal MMMT is thought to originate from the secondary Müllerian system. Mullerian carcinosarcomas may be categorized as homologous or heterologous, depending on the histologic characteristics of the sarcomatous elements [7].

When components foreign to the uterus, including cartilage, like in our case, and bone, are observed, the tumor belongs to the heterologous type [7].

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 [7].

Extragenital MMMT was found to be associated with synchronous or metachronous gynecologic tumors of mullerian duct origin [25], previous irradiation [17], and extragenital endometriosis [26]. In the present case, the patient had no history of an operation for any other tumors or previous irradiation. Microscopically, there was neither associated endometriosis, and the tumor revealed chondrosarcoma-like components.

Owing to the rarity of primary peritoneal carcinosarcomas, limited data exists regarding their treatment management [27]. The basic treatment is surgical excision, but most cases of carcinosarcomas present metastasis at the time of presentation, making surgery difficult or impossible [28]. Systemic chemotherapy is recommended in all cases regardless of stage due to the early spread of tumors. Platinum in combination with ifosfamide were the preferred agents [29]. There is not enough data to support the role of radiotherapy in extragenital carcinosarcoma [22].

The review of the literature shows that the patients underwent surgery followed by chemotherapy, when it was possible [27].

Conflict of Interest

There is no conflict of interest.

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