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COMMUNICATION

A rare case of primary extragenital retroperitoneal carcinosarcoma with review of the literature

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Abstract Carcinosarcomas are rare, highly aggressive neoplasms that most commonly arise from the female genital tract but can unusually present in extragenital locations. To the best of our knowledge, only three cases of primary retroperitoneal, extragenital carcinosarcomas have been documented in the English literature to date. A 72-year-old woman presented with onset of abdominal pain and urinary retention. Computed tomography scan revealed a mass in the retroperitoneal space with hydronephrosis and partial obstruction of the left ureter. Lost to follow-up at that time, the patient returned 6 months later with a left leg deep vein thrombosis. On exploratory laparotomy at that time, the retroperitoneal mass was found to completely engulf the left ureter, iliac artery, and vein. Resection was not possible, but biopsy confirmed the presence of an extragenital carcinosarcoma.

Keywords Carcinosarcoma · Mullerian · Retroperiteonal · Extragenital

Case report

A 72-year-old Caucasian female presented with new onset left-sided abdominal pain and urinary retention. The patient's medical history was insignificant except for remote total abdominal hysterectomy and bilateral salpingoophorectomy done for symptomatic fibroids.

A computed tomography (CT) scan revealed a 5-×7-cm mass in the retroperitoneal space with left hydronephrosis due to mass effect (Fig. 1). Ureteric stenting was performed, but the patient was then lost to follow-up. Six months later, the patient presented again with new onset of left leg deep vein thrombosis. A repeat CT scan revealed extensive expansion in the retroperitoneal space with complete engulfment of the left renal collecting system as well as partial engulfment of the left iliac vein and artery. A CT-guided biopsy was suspicious but not conclusive for carcinosarcoma. The decision was made to obtain an open biopsy with possible resection.

During exploratory laparotomy, the mass was found in the lower left retroperitoneal space with extension up to the aorta and was deemed unresectable. A 3-×2-cm open tissue biopsy was obtained, and pathology confirmed the presence of extragenital carcinosarcoma.

The patient made a satisfactory postoperative recovery. She received radiation therapy but had a poor response to this. In a matter of 14 months from laparotomy, the patient developed massive (13 cm) liver (Fig. 2) and lung metastasis and was transferred into Hospice care.

Pathology: gross and microscopic

The needle biopsy of the retroperitoneal mass grossly consisted of multiple light tan, cylindrical fragments of soft tissue. Microscopically, the biopsy showed an infiltrative malignant neoplasm consisting of a glandular epithelial component and a stromal spindle cell component. There was a background of collagenous fibrosis and focal necrosis.

All authors have contributed significantly to this manuscript and are in agreement with the content of this manuscript.

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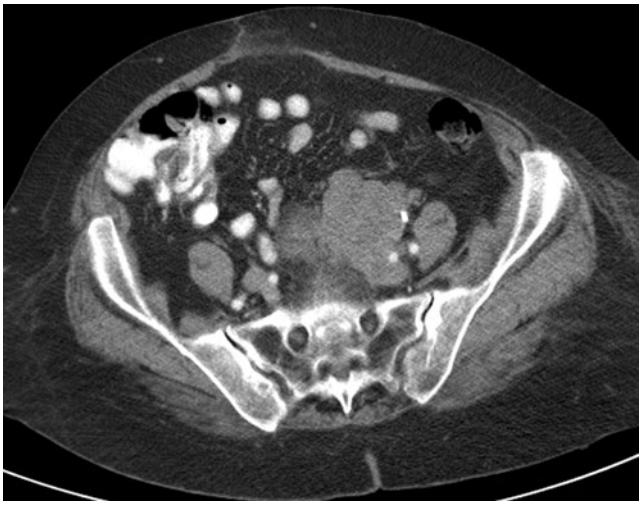

Fig. 1 CT on initial presentation showing the left retroperitoneal mass

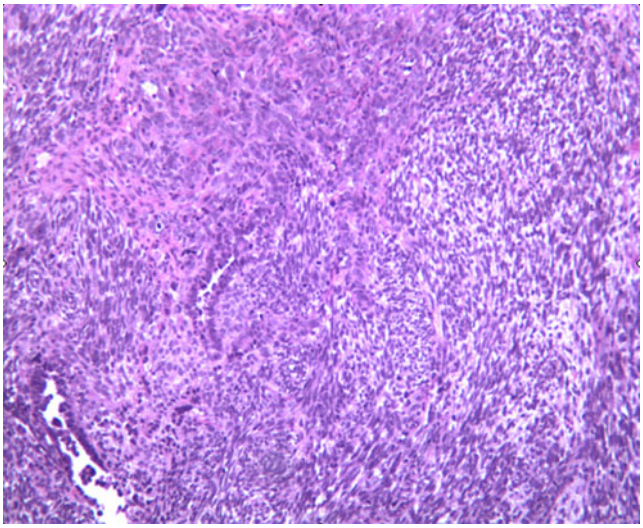

Fig. 3 H&E stain of the tumor

The open biopsy of the retroperitoneal mass revealed an irregular, pink, soft, friable tumor fragment measuring 3.0×2.5×2.2 cm. The specimen had a biphasic pattern consisting mostly of malignant spindle cells and elements of epithelial cells (Fig. 3).

Immunohistochemical stains were positive for Vimentin (Fig. 4), representing the sarcomatous portion of the tumor. The epithelial component was strongly positive for Pankeratin (Fig. 5). The cells demonstrating Vimentin and Pankeratin staining were closely intermingled; They did not come from different portions of the mass, which is consistent with a mesodermal mixed tumor. There was weak staining for Calretinin and CK20. The neoplasm was negative for S100 and showed nonspecific weak staining for leukocyte common antigen. Correlating our findings with the clinical history of a rapid growing well-defined nodular tumor, the diagnosis was consistent with a homologous type primary carcinosarcoma. On the initial CT and at the time of the open biopsy, the tumor appeared to be arising from the retroperitoneum; it appeared to be invading into and not spreading out of the ureter or kidney. It also did not appear to be arising from the peritoneum or from inside the peritoneal cavity. Based on this data, we identified the tumor as a primary retroperitoneal carcinosarcoma.

Discussion

Malignant mesodermal mixed tumors (MMMT), also known as mullerian carcinosarcomas, are tumors of the

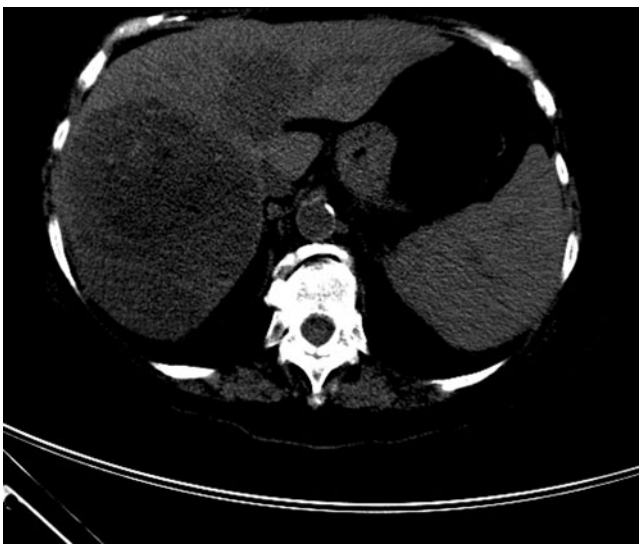

Fig. 2 CT showing large liver metastasis and splenomegly found on follow-up CT

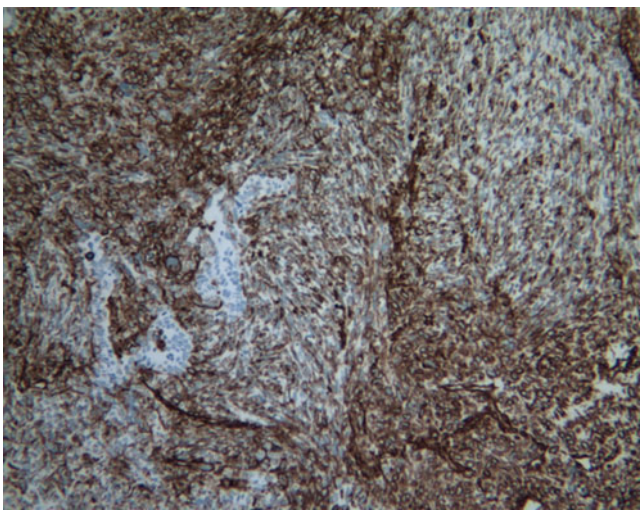

Fig. 4 Vimentin stain, revealing the sarcomatous portion of the tumor

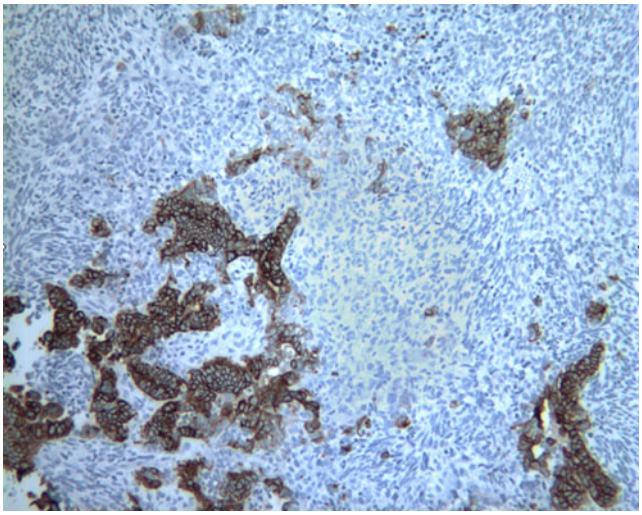

Fig. 5 Pankeratin stain, highlighting the carcinomatous portion of the tumor

female genital tract, which contain both carcinomatous and sarcomatous elements. These malignancies usually arise from the uterus, comprising 2–3% of uterine malignancies [1]. They arise less commonly from the cervix, fallopian tube, and ovary [12]. Extragenital mullerian carcinosarcomas are even more uncommon, representing only about 29 reported cases since first being reported by Ober and Black in 1955 [6, 11]. The pelvic wall, peritoneum, and omentum, ureter, and kidney have been described as extragenital locations [5, 10, 13]. To date, we can find only three other cases of primary retroperitoneal carcinosarcoma that have been reported in the English literature [2, 4, 12] (Table 1).

Carcinosarcomas are classified by what type of sarcomatous tissue is present. Two types have been described: homologous type if a sarcoma is of uterine origin and heterologous type if extra-uterine tissue is present, such as muscle, cartilage, or bone [4]. Heterologous mixed mullerian tumors carry a worse prognosis than homologous. It is thought that the ability of the tumor to form heterologous tissue types denotes more aggressive potential [4].

The origin of extragenital mullerian tumors is uncertain. An association with radiation has been suggested [4], but lack of firm evidence for this correlation has been discounted by some authors [3]. Three main hypotheses have been proposed. First, these malignancies may arise out of ectopic foci of endometriosis. Although there have been reports of carcinosarcomas arising from endometriosis [1], there are other cases in which no endometrial tissue was able to be documented [13]. No endometrial tissue was demonstrated in our case. Another hypothesis features the so-called secondary mullerian system [12]. The secondary mullerian system, first described by Lauchlan, posits that the abdominal and pelvic peritoneum and retroperitoneal mesenchyme retain the potential for mullerian differentiation, probably due to their close embryologic relation to the mullerian ducts [7, 8]. Therefore, these tissues of the secondary mullerian system could possibly spontaneously differentiate into mullerian malignancies. Lastly, mullerian carcinosarcomas could begin as poorly differentiated epithelial neoplasms that undergo transformation into mesenchymal differentiation [9]. The transformation of carcinomas into tumors with sarcomatoid elements has been previously demonstrated in ovarian carcinomas [9].

Most extragenital carcinosarcomas reported have occurred in middle-aged to older individuals [3]. The CA-125 tumor marker was noted to be elevated in at least one case and returned to normal after the tumor was removed [3]. However, although CA-125 can be elevated when the carcinomatous element of the carcinosarcoma is

Table 1 Three cases reporting primary retroperitoneal carcinosarcoma

	Case 1	Case 2	Case 3
Author	Ferrie and Ross [2]	Herman and Tessler [4]	Shintaku and Matsumoto et al. [12]
Presentation	Palpable mass	Weakness, anorexia and vague abdominal pain	Abdominal fullness, pain, diarrhea and nausea
Tumor size	11 cm diameter, 650 g	700 g	20 cm diameter, 1,040 g
Resectable	Yes	No	Yes
Surgical intervention	En bloc resection	Debulking with left nephrectomy	En bloc resection with TAH-BSO and omentectomy.
Tumor location	Retroperitoneum below right kidney	Retroperitoneum involving the left kidney and ureter	Left retroperitoneum extending to lateral pelvic wall
Chemotherapy	None reported	Adriamycin, Cytoxan, DTIC, Vincristine	Intraperitoneal carboplatin then adjuvant epirubicin hydrochloride and carboplatin
Patient age	45	72	51
Metastases	None	Bilateral pulmonary nodules	None
Sarcomatous elements	Malignant stroma of endometrial type	Chondrosarcoma	Spindle cell

predominating, it is not an effective diagnostic tool and is useful primarily for follow-up [14]. The prognosis for extragenital carcinosarcomas is poor, with most patients dying within 1 year [3, 6]. Knowledge of the most effective treatment is limited due to the paucity of cases [12], and chemotherapy and radiation are of limited effectiveness [3]. However, carboplatin–paclitaxel has been shown to be effective against uterine MMMT [15] and may offer some benefit in tumors of this type as well. Surgical resection remains the mainstay of treatment, even if the goal of therapy is palliation. Given the propensity of extragenital carcinosarcomas for invasion of surrounding structures, it is recommended that tumor removal is performed by an oncologic surgery team, often involving members of different surgical subspecialities.

Conclusion

Carcinosarcomas are rare tumors that histologically contain the epithelial and mesenchymal elements of both a carcinoma and a sarcoma. Although rare, a high level of suspicion must be maintained for any fast-growing tumor of the retroperitoneal space. Found primarily in the female reproductive tract, carcinosarcomas can arise from a variety of extragenital locations including the retroperitoneal space. Surgical excision is the treatment of choice in cases amenable to resection. Early detection and aggressive surgical intervention should be the goal of any clinician faced with this rare but aggressive neoplasm.

Conflict of interest The authors declare that there are no conflicts of interest.

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