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# Retroperitoneal Extraskeletal Mesenchymal Chondrosarcoma: A Very Rare Case Report

CASE REPORT

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ABSTRACT

Chondrosarcoma is an uncommon malignant neoplasm of cartilaginous origin. Extraskeletal chondrosarcomas are far less common than their intraosseous counterparts, representing approximately 2% of all soft tissue sarcomas. Clinically, extraskeletal mesenchymal chondrosarcoma (EMC) shows a very fast progression. It also has a high incidence of local recurrence and distant metastasis. The most common locations are the bladder, brain, meninges, mediastinum, and thighs. Other sites, including the retroperitoneum, are very uncommon. The diagnosis of mesenchymal chondrosarcoma can be very challenging, particularly in cases without conspicuous cartilaginous differentiation. In such cases, its distinction from other neoplasms cannot be safely established. Here we present a retroperitoneal tumor diagnosed as EMC in a 73-year-old female.

Keywords: Chondrosarcoma, multidetector computed tomography, retroperitoneal

## INTRODUCTION

Chondrosarcoma in which primary-type lesions grow is a type of malignant tumor developing a cartilage matrix. This type of chondrosarcoma accounts for 20%–27% of all primary malignant osseous neoplasms, making it the third most widespread primary malignant bone tumor. On the other hand, extraskeletal chondrosarcomas are a lot lesser and account for about 2% of all soft tissue sarcomas. In terms of histology, there are two types of extraskeletal chondrosarcomas: myxoid and mesenchymal. When these two types are compared, the mesenchymal type has a weaker prognosis and is more uncommon and aggressive (1, 2). While an upper limb location is comparatively rare, the lower limbs, orbit, and meninges are frequent extraskeletal sites (3, 4). Moreover, extraskeletal mesenchymal chondrosarcoma (EMC) of the retroperitoneum is very uncommon (5). In particular, when there is no conspicuous cartilaginous differentiation, the diagnosis of mesenchymal chondrosarcoma may be difficult. Therefore, differentiation between mesenchymal chondrosarcoma and other neoplasms is not very accurate on such occasions.

Here we present a case of a 73-year-old female with a retroperitoneal tumor diagnosed as EMC.

## CASE REPORT

A 73-year-old female presented with a 6-month history of a palpable mass in the lower abdomen. Examination of the abdomen revealed a large mass in the lower abdomen. The patient history was as follows: hysterectomy and ovariectomy performed 10 years previously. General physical examination and the remaining systemic examination revealed no other abnormalities. The laboratory data were normal. Routine abdominal ultrasonography revealed a heterogeneous, hypoechoic solid mass in the lower abdomen (Figure 1). Computed tomography (CT) (128-slice Somatom CT scanner; Siemens, Erlangen, Germany) of the abdomen and pelvis revealed a large, well-defined, lobulated, hypodense mass in the retroperitoneum, measuring approximately 100×140×170 mm in transverse, anteroposterior, and craniocaudal axes. No peripheral or central calcification was observed. Contrast-enhanced CT showed a smoothly marginated, heterogeneously and peripherally enhanced tumor adjacent to the retroperitoneum, unconnected with any other pelvic structures (Figures 2a-c). After written informed consent was obtained from the patient, she underwent total surgical resection of the mass. The tumor was completely resected, and any complication occurred. Histopathological assessment revealed EMC with undifferentiated spindle cells and focal areas of well-differentiated cartilaginous tissues (Figures 3a, b). Postoperative chemotherapy and radiotherapy were used for treatment. Recurrence was noted on CT scan after 12 months (Figure 4).

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## DISCUSSION

Retroperitoneal sarcomas, when compared to others, are uncommon tumor types, and they are generally seen in a locally advanced stage. Extraskeletal chondrosarcomas are relatively rare neoplasms and constitute only about 2% of all soft-tissue sarcomas. Histologically, there are two types of extraskeletal chondrosarcomas: myxoid and mesenchymal. EMC is a rare malignant tumor, but it progresses fast.

It is possible for chondrosarcomas to occur in extraskeletal locations like soft tissues where ordinarily cartilage is not encountered, even though most of them come out of cartilaginous or bony structures. Lots of these lesions attack the head and the neck of the body, including the brain, orbit, and meninges. Musculoskeletal lesions generally affect the lower extremity, especially the thighs (3, 6). However, they are very uncommon with other sites, such as the retroperitoneum (5).

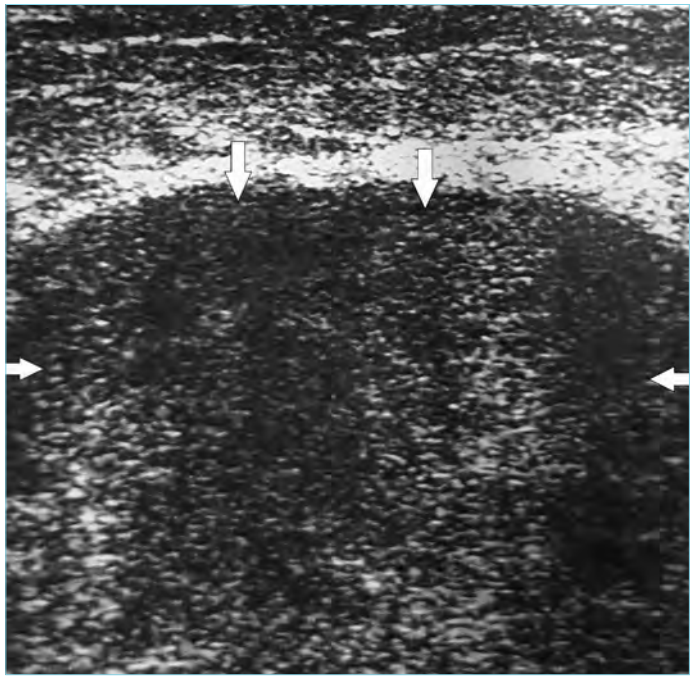

**Figure 1. Routine abdominal ultrasonography revealed a heterogeneous, hypoechoic, solid mass in the lower abdomen (*arrows*)**

The retroperitoneal space is surrounded by a peritoneal covering from the anterior part, by the posterior abdominal wall from the posterior part, by the 12$^{th}$ rib and vertebra from the superior part, by the base of the sacrum and iliac crest from the inferior part, and finally by the site borders of the quadratus lumbora muscles from the lateral part. In this space are connective tissue, the adrenals, the kidneys and ureters, the aorta and its branches, inferior vena cava and its tributaries, and lymph nodes (7). At the same time, the retroperitoneal soft-tissue sarcomas, such as the liposarcoma, lipoma, malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma, vascular tumors, peripheral nerve tumors, synovial sarcoma, extraskeletal osteosarcoma, and chondrosarcoma, occur in this area.

The peak incidence of retroperitoneal sarcomas is the sixth decade of life, even though they can be seen in any ages (8). The most common finding of mesenchymal chondrosarcomas is a slowly swelling painless soft-tissue mass; however, clinical symptoms of them are nonspecific, even though pain, early satiety, symptoms of bowel obstruction, neurologic symptoms of pain radiating to the lower limb or ipsilateral lower limb edema may indicate mesenchymal chondrosarcoma.

Particularly when there is no conspicuous cartilaginous differentiation, diagnosing mesenchymal chondrosarcoma may be difficult. Therefore, making a differentiation between it and other neoplasms cannot be safely done in such cases.

The radiographs belonging to these lesions frequently display a nonspecific soft-tissue mass. It may be possible to see areas of chondroid matrix mineralization, which is a lot more recurring in mesenchymal chondrosarcoma. Uncovering of a solid heterogeneous mass that regularly contains scattered areas of increased echogenicity and posterior shadowing because of calcific foci is

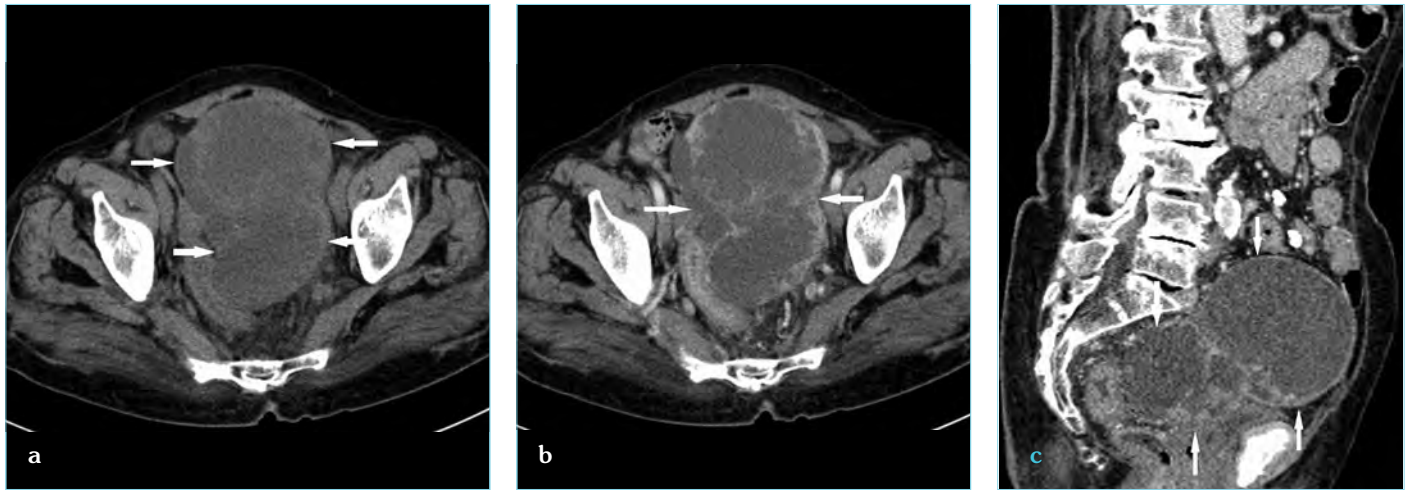


**Figure 2. a-c. An axial nonenhanced computed tomography (a) of the abdomen and pelvis revealed a large, well-defined, lobulated, hypodense mass in the retroperitoneum. Axial (b) and sagittal (c) contrast-enhanced CT showed a smoothly marginated, heterogeneously and peripherally enhanced tumor (*arrows*)**

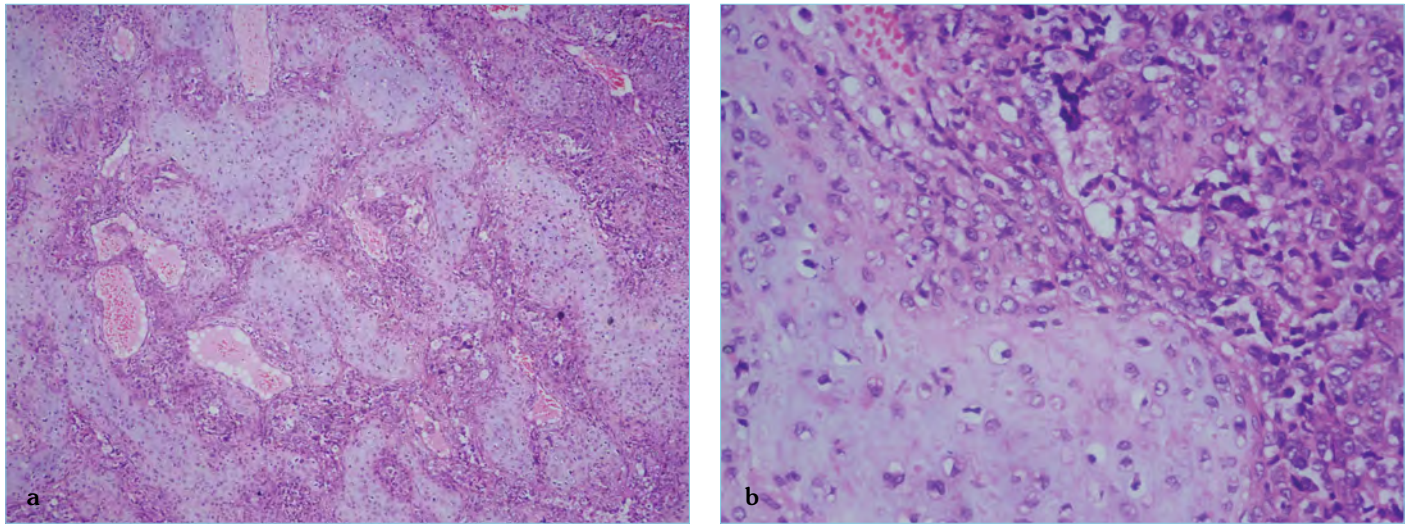


**Figure 3. Histopathological assessment revealed undifferentiated spindle cells and focal areas of well-differentiated cartilaginous tissues [hematoxylin and eosin stain, magnification ×100 (a), ×400 (b)]**

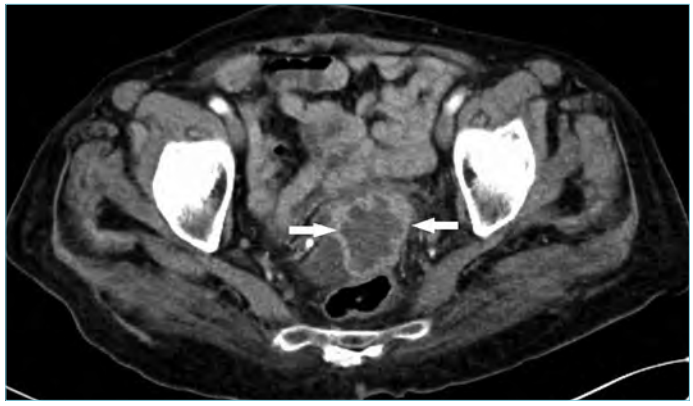

**Figure 4. A contrast-enhanced axial CT scan showed recurrent tumor (*arrows*)**

generally done by ultrasound. The patients who are doubted to have developed a mass are examined preferably via CT, through which one can see the dimension and size of the tumor and its connection to the vessels, organs, and skeletal structures around it (9). The attenuation of EMC and muscle on CT scans are alike, but EMC, which has a lower water content caused by the intermixture of small cells and more limited cartilaginous tissue, ordinarily has an intermediate signal intensity on T2-weighted MR images. Becoming visible as areas of high signal intensity on T2-weighted MR images, it is possible to notice areas of necrosis. Via the MR images obtained after intravenous administration of contrast material, it is possible to display prominent and diffuse enhancement, which is also heterogeneous (10). With CT and MRI images, it may still not be possible to make a difference between EMC and other neoplasms. Therefore, an exact diagnosis can be ensured depending on histological features. The basic characteristics of mesenchymal chondrosarcoma is, when looked at using a microscope, a biphasic pattern composed of sheets of undifferentiated round, oval, or spindle-shaped cells and small, usually well-defined, islets of well-differentiated, benign-appearing cartilaginous tissue (3).

The favorite treatment of the tumor is to resect with a surrounding margin of normal tissue, which seems to be the single way of curative treatment. However, more studies are needed for the use of combined modality treatment for this kind of tumor, since their recurrence after surgery is at a high level. Local recurrence is the most widespread form of recurring of EMC. To improve the cure rates and relapse-free survival, adjuvant radiotherapy is suggested.

## CONCLUSION

Mesenchymal chondrosarcoma is a rare and more aggressive version of conventional chondrosarcoma; nearly 30%–50% of mesenchymal chondrosarcomas originate extraskeletally. Localization to the retroperitoneum is very rare. Retroperitoneal chondrosarcomas are present in the form of an abdominal mass and can be monitored with ultrasonography. MR and CT images demonstrate several overlapping features.

**Informed Consent:** Written informed consent was obtained from patient.

**Peer-review:** Externally peer-reviewed.

**Authors' Contributions:** Conceived and designed the experiments or case: MK. Performed the experiments or case: MK, MRÖ. Analyzed the data: MK, MRÖ. Wrote the paper: MK. All authors have read and approved the final manuscript.

**Conflict of Interest:** No conflict of interest was declared by the authors

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