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CASE REPORT

Retroperitoneal Myoepithelial Carcinoma: A Case Report and Literature Review

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Background: Myoepithelial carcinoma is rare, and myoepithelial carcinoma occurring outside the head and neck is even rarer. We reported one case of retroperitoneal myoepithelial carcinoma.

Case Summary: A 63-year-old woman who underwent computed tomography (CT) for progressive abdominal distension revealed a left retroperitoneal mass and subsequently underwent surgical treatment where the mass was completely removed with a postoperative diagnosis of retroperitoneal myoepithelial carcinoma. A follow-up CT review 40 days after surgery revealed a recurrence of the mass. After 8 months of chemotherapy and targeted immunotherapy, a follow-up review of the CT images revealed a gradual reduction in the mass. Four months after the cessation of chemotherapy and targeted drug combined immunotherapy, a follow-up review via CT revealed another recurrence and enlargement of the mass.

Conclusion: CT of retroperitoneal myoepithelial carcinoma revealed a massive cystic solid mass in the abdominal cavity and retroperitoneum. The solid region of the mass was significantly enhanced and the cystic region was without enhancement on enhanced CT; the mass involved the adjacent duodenum, partial jejunum, and left renal vein. PET–CT imaging revealed hypermetabolism in the solid region of the mass and no hypermetabolism in the cystic region.

Plain Language Summary: We studied a rare cancer called retroperitoneal myoepithelial carcinoma to understand how to treat it better.

Why: This cancer is tricky and often comes back after treatment. We wanted to find ways to help patients live longer.

What we did: We followed one patient's journey. Doctors used CT scans to see where the tumor was and how big it was. PET–CT scans showed how active the cancer cells were. The patient had surgery to remove the tumor, but it came back. They then tried chemotherapy, targeted drugs, and immunotherapy. Sadly, the cancer returned again.

What we found: This cancer is prone to coming back, but using multiple treatments together helps patients live longer. The scans were crucial for tracking the tumor and seeing how it changed.

What it means: Treating this cancer is challenging, but combining different treatments works better than using just one. Our findings show that doctors need to keep a close watch on patients and use all available tools to fight this disease. This case helps us understand how to improve care for others with this rare cancer.

Keywords: computed tomography, CT, myoepithelial carcinoma, retroperitoneal, rare diseases

Introduction

Myoepithelial carcinoma (MC), also known as malignant myoepithelioma (MME), is uncommon clinically and accounts for less than 1% of salivary gland tumors.[1] MC mainly occurs in the salivary glands of the head and neck. MC cases originating in other parts of the body, such as the lungs, stomach, ear canal, and scrotum, have been reported.[2–6] Although Aadya Kerkar et al[7] reported one recurrent retroperitoneal MC case which emphasized the need for cytomorphological awareness in 2024, we did not find other related reports about retroperitoneal MC cases. We report one case of primary retroperitoneal MC admitted to our hospital and analyze its clinical and imaging

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manifestations, pathological characteristics, and diagnosis and treatment. This report emphasizes the need for imaging awareness with the employment of ancillary testing for accurately diagnosing and following this rare tumor at an uncommon location. According to our best knowledge, such a case is perhaps being documented for the first time in the imaging literature. The existing literature is also reviewed with the differential diagnoses and making a distinction.

Case Presentation

The research protocol was approved by the Ethics Committee of Qingxian People's Hospital (No. 20241201). Because the patient has died, consent for publication was obtained from the patient's family. The case details have been approved for publication by Qingxian People's Hospital.

Chief Complaints

Progressive abdominal distension for more than 1 month.

History of Present Illness

The patient complained of abdominal distension without inducement for 1 month. The distension was slightly obvious after eating and occurred without nausea, vomiting, diarrhea, or fever. Owing to tolerable symptoms, the patient did not receive a diagnosis or treatment. Later, the patient's abdominal distension worsened, accompanied by intermittent vomiting. The vomit included food and bile ingredients, and the patient's abdominal distension was not significantly relieved. On March 14, 2023, the patient went to the local hospital for fasting water, gastrointestinal decompression, and supportive treatment with nutritional rehydration. The results of enhanced CT performed on March 15, 2023, revealed an irregular massive cystic solid lesion in the left middle abdomen. The patient was admitted to our hospital on March 17, 2023, for further treatment.

History of Past Illness

The patient had hypertension for 20 years and was treated with oral nimodipine tablets with stable blood pressure control. The patient denied a history of stroke, hepatitis, tuberculosis, or trauma; a history of blood product infusion; and a history of drug and food allergies.

Personal and Family History

The patient had no history of smoking or alcohol consumption and denied a familial genetic history or infectious disease.

Physical Examination

The patient had an abdominal bulge. A fixed circular mass of approximately 11×10 cm could be reached near the costal margin, exhibiting a smooth surface, poor mobility, and an unheard vascular murmur.

Laboratory Examinations

(1) Routine blood test: blood platelet 428×10^9/L (normal value $100–300\times10^9$/L), white blood cell 6.73×10^9/L (normal value $3.5–9.5\times10^9$/L), red blood cell 4.2×10^{12}/L (normal value $3.8–5.1\times10^{12}$/L), hemoglobin concentration 126 g/L (normal value 115–150 g/L).

(2) Tumor marker: Sugar antigens CA125 124.5 U/mL (normal value 0–22 U/mL), Sugar antigens CA199 < 2.00 U/mL (normal value 0–43 U/mL), Sugar antigens CA153 10.1 U/mL (normal value 0–20 U/mL), Sugar antigens CA724 0.721 U/mL (normal value 0–6.9 U/mL), Sugar antigens CA50 5.349 IU/mL (normal value 0–25 IU/mL), Carcinoembryonic antigen 0.89 ng/mL (normal value 0–5 ng/mL), Alpha fetal protein (AFP) 3.42 ng/mL (normal value 0–7 ng/mL), Cytokeratin 19 fragment 1.410 ng/mL (normal value 0–3.3 ng/mL), Neuron-specific enolase 7.06 ng/mL (normal value 0–16.3 ng/mL), Squamous cell carcinoma-associated antigen 0.5 ng/mL (normal value 0–1.5 ng/mL).

Imaging Examinations

CT Findings

CT images revealed a massive round cystic solid mass in the abdominal cavity and retroperitoneum, approximately 13.9 cm×12.2 cm×11.8 cm in size, which was mainly a cystic mass with multiple solid septations. The mass from the adjacent duodenum, some of the jejunum, and the left renal vein was unclear. Enhanced CT showed no enhancement of the cystic region of the mass, and significant enhancement of the solid region of the mass was not enhanced (Figure 1). The CT findings were as follows: a retroperitoneal mass and a high possibility of malignancy.

PET–CT Findings

No abnormalities were observed in the nasopharynx, oropharynx, or larynx structures or radioactivity. Salivary glands and thyroid glands were normal in size, morphology, density, and radioactivity.

A large retroperitoneal cystic solid mass was observed, and the solid region of the mass was hypermetabolic, with an SUV max of 7.5 and no hypermetabolism in the cystic region (Figure 2).

Multidisciplinary Expert Consultation

After multidisciplinary expert consultation, the patient's clinical symptoms were obvious and the relevant examination improved. There were no clear surgical contraindications, so surgical treatment was immediately selected.

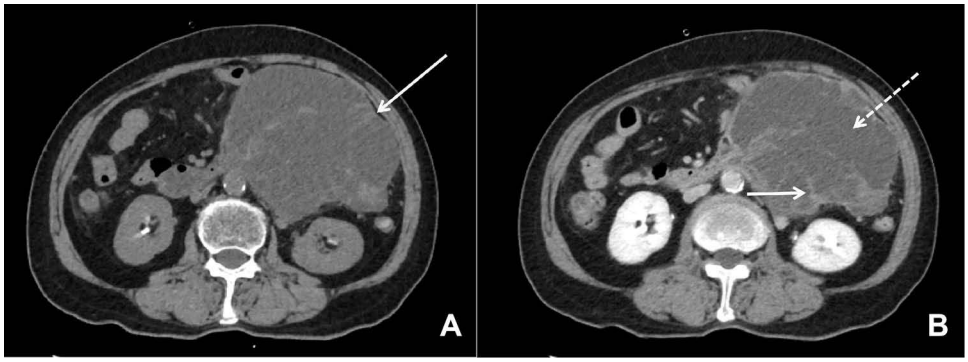

Figure 1 Transaxial non-contrast CT (**A**) and contrast-enhanced CT (**B**) images. (**A**) Transaxial non-contrast CT image showed a large retroperitoneal cystic solid mass with solid separation within the mass (solid arrow). (**B**) Transaxial contrast-enhanced CT image showed significant enhancement of the mass wall and solid septation (solid arrow) and no enhancement of the cystic region (virtual arrow).

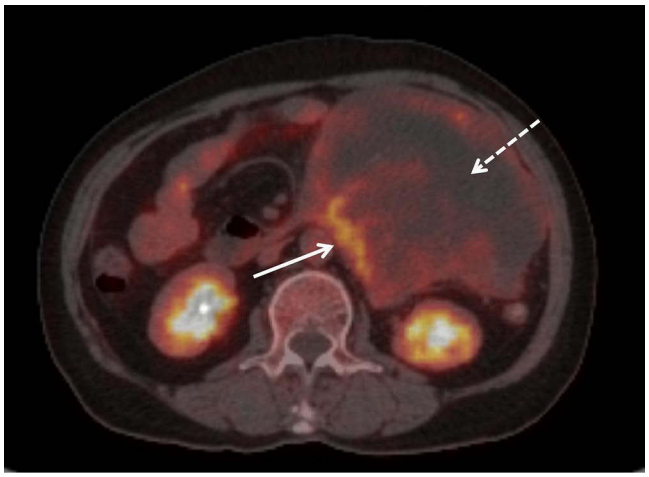

Figure 2 Transaxial fused 18F-FDG PET-CT image showed an active metabolism of the solid component of the retroperitoneal mass (solid arrow) and no FDG avid of the cystic component (virtual arrow).

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Final Diagnosis

(1) Retroperitoneal myoepithelial carcinoma. (2) Malignancies secondary to the abdominal cavity. (3) Secondary malignancy of the jejunum.

Treatment

The patient underwent retroperitoneal resection on March 29, 2023: the omentum was opened, revealing a cystic solid mass fixed to the retroperitoneum with a hard pedicle and a size of approximately 14 cm×12 cm×11 cm. The surface of the mass was smooth and dark brown, with brown liquid inside. Some of the cystic wall was convex, adhering to the duodenal jejunum, the lower edge of the pancreas, and the transverse colon. After the puncture and aspiration of part of the cystic fluid, the tumor body was smaller than the front, the puncture mouth was sutured, and the tumor body was completely removed. After surgery, anti-inflammatory, acid suppression, and nutritional rehydration support therapy were given, and the patient was discharged after reaching a stable condition.

Outcome and Follow-up

Pathological Findings

Pathological images under the microscope revealed fibrous tissue, visible tumor cell slices or diffuse invasive growth with extensive bleeding and cystic changes, multifocal necrosis, round, short, or epithelioid tumor cells, medium–severe cells, rich cytoplasm, powder, nucleus ovals, nucleoli, nuclear division, focal interstitial powder dye, and collagen degeneration (Figure 3).

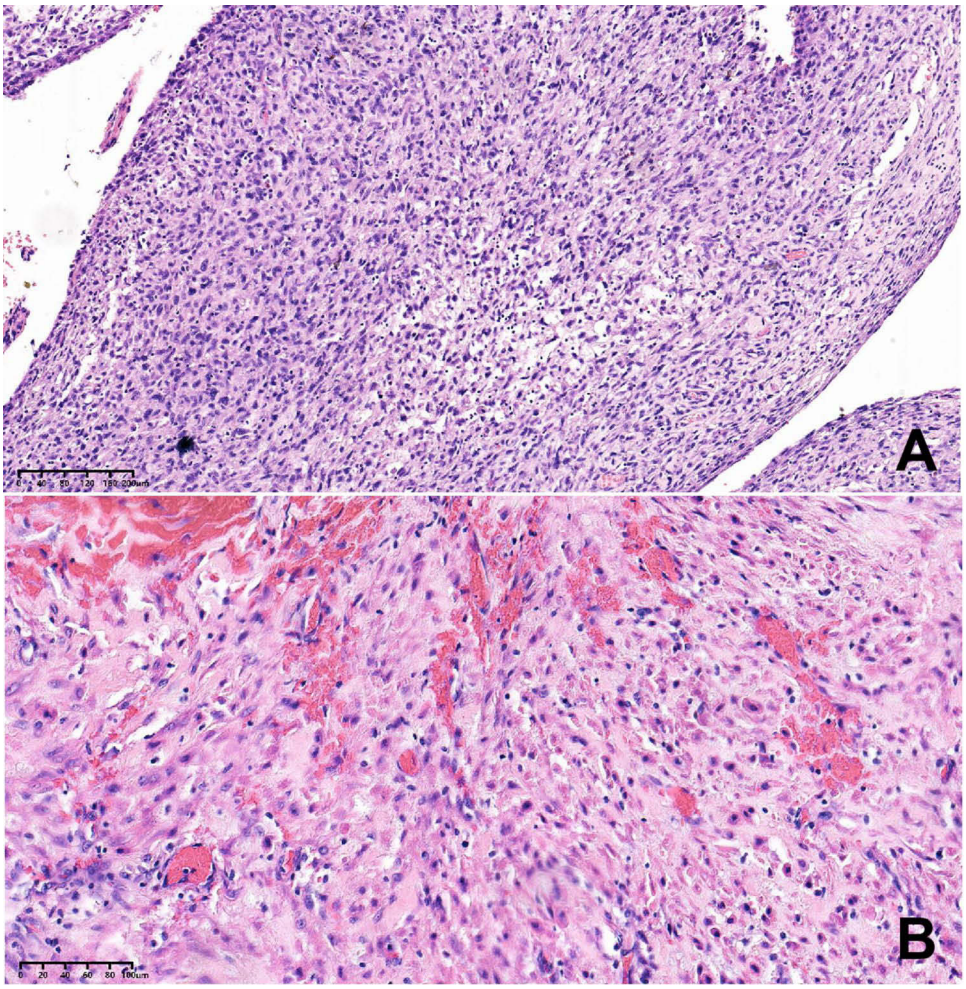

Figure 3 Pathological images under the microscope (**A** and **B**). (**A**) Hematoxylin-eosin staining (200 × 200) staining showed the tumor cells were round, short spindle-shaped, or epithelial-like. (**B**) Hematoxylin-eosin staining (400 × 400) staining showed medium-severe cell heterogeneous, rich cytoplasm, oval nucleus, obvious nuclei, more common nuclear division, focal interstitial powder staining, collagen degeneration, rich small and medium blood vessels in the background.

The immunohistochemical results were as follows: P63 (diffuse +), calponin (partial +), and CK (+) (Figure 4).

In summary, the lesion was a malignant tumor with myoepithelial differentiation, which is consistent with myoepithelial carcinoma.

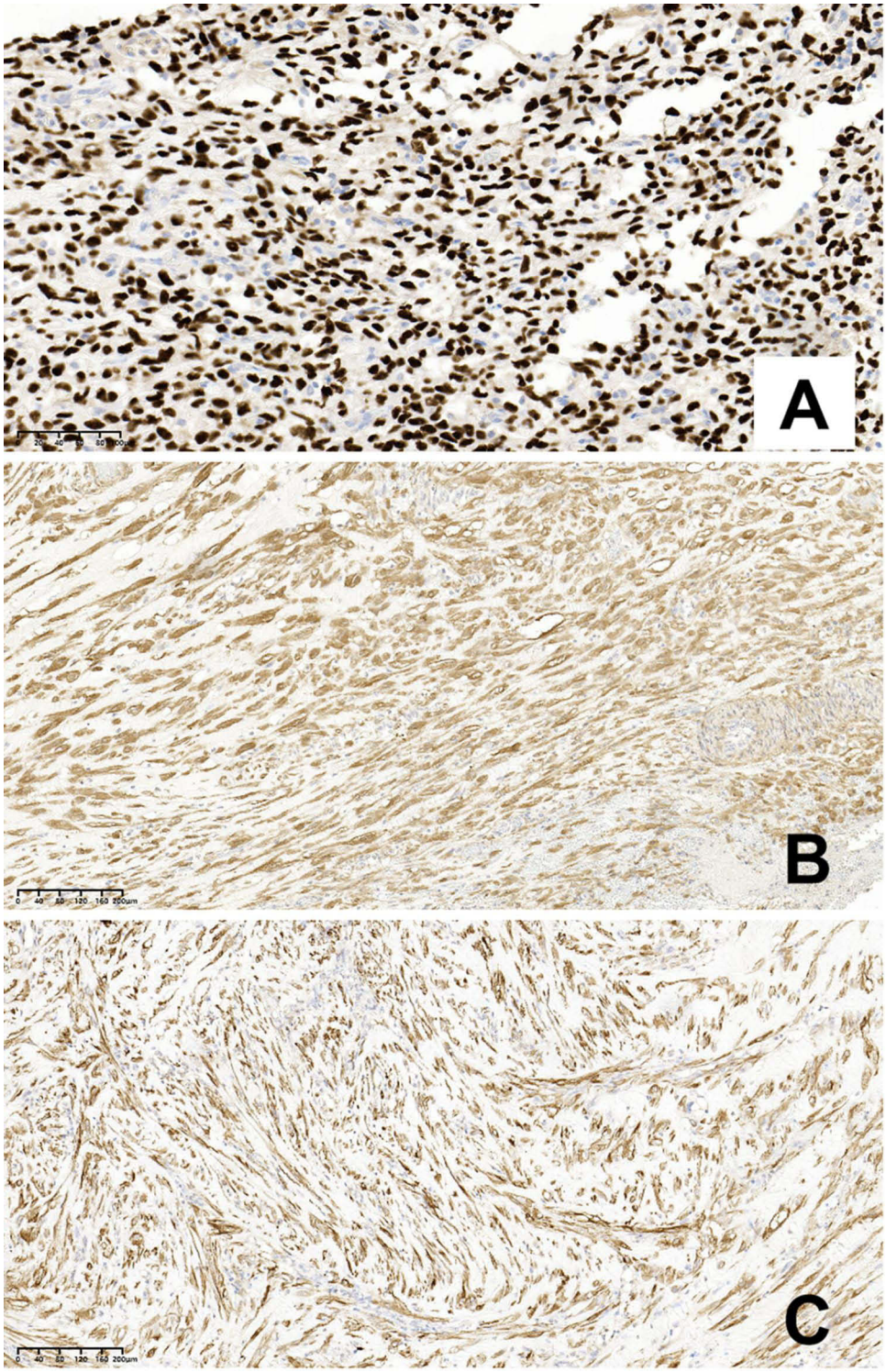

Figure 4 Immunohistochemical images (**A-C**). (**A**) Immunocytochemistry: Diffuse and intense immunoreactivity for epithelial membrane antigen (200 × 200). (**B**) Immunocytochemistry: Patchy and focal positivity for Calponin (200 × 200). (**C**) Immunocytochemistry: Tumor cells showing positivity for CK (200 × 200).

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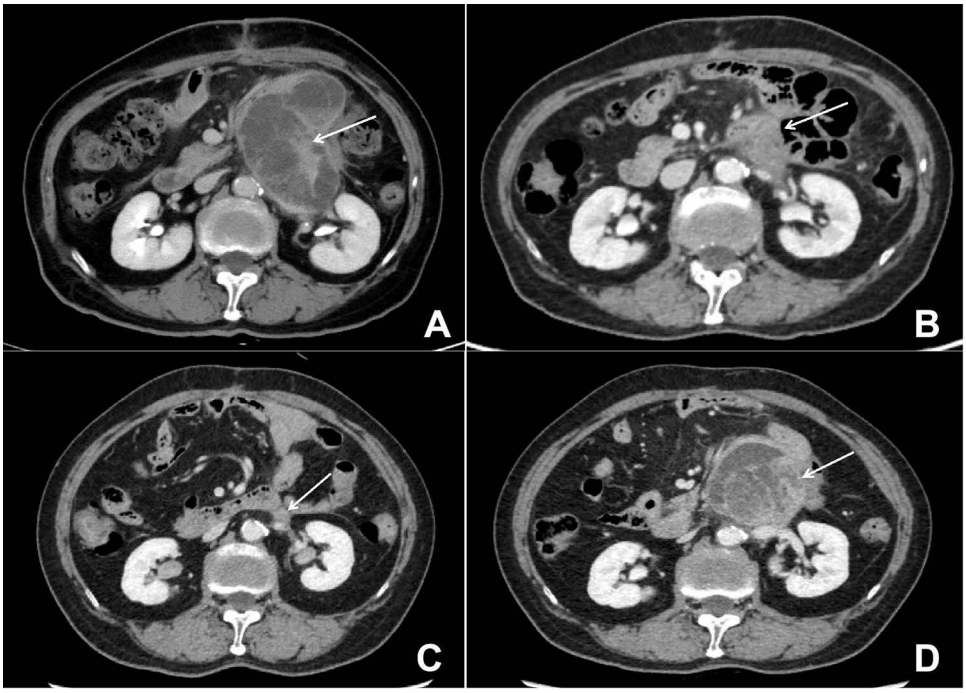

Figure 5 Transaxial contrast-enhanced CT images (**A-D**). (**A**) On 09-May-2023, transaxial contrast-enhanced CT showed a recurrent retroperitoneal mass (solid arrow) of about 9.2cm × 4.7cm × 6.1cm. (**B**) On 17-Jul-2023, transaxial contrast-enhanced CT showed the retroperitoneal mass (solid arrow), about 2.1cm × 2.6cm × 3.3cm. (**C**) On 08-Jan-2024, transaxial contrast-enhanced CT showed a significant reduction of the retroperitoneal mass (solid arrow) with a size of approximately 0.9cm. (**D**) On 08-May-2024, transaxial contrast-enhanced CT showed another recurrence of the retroperitoneal mass (solid arrow), about 6.1cm × 6.4cm × 7.1cm.

The genetic findings included a heterozygous BCL2L11 (BIM) mutation and an EML 4-ALK fusion variant. The patient was reviewed with CT on May 9, 2023, which revealed a recurrence of the retroperitoneal cystic solid mass with a size of 9.2 cm×4.7 cm×6.1 cm. The patient subsequently received two rounds of chemotherapy and two rounds of targeted drugs combined with immunotherapy, with the following specific regimen: 200 mg ocrelizumab + 400 mg bevacizumab + 40 mg doxorubicin hydrochloride liposome + 100 mg nedaplatin. A review of CT data obtained on July 17, 2023, revealed that the retroperitoneal mass was significantly smaller than before. The tumor size was approximately 2.1 cm×2.6 cm×3.3 cm. The patient received five rounds of chemotherapy and six targeted combination immunotherapies after enhanced CT on January 8, 2024, and CT images revealed a significant reduction in the retroperitoneal mass (approximately 0.9 cm). On May 8, 2024, the patient's re-examination via CT revealed a recurrence of the retroperitoneal capsule, with dimensions of 6.1 cm×6.4 cm×7.1 cm (Figure 5). On July 18, 2024, the patient died of heart and kidney failure.

Discussion

Myoepithelial carcinoma (MC) was proposed by the WHO in 1991 as a separate type in the histological analysis of salivary gland tumors.[8] Moreover, MCs are distinguished from epithelial–myoepithelial carcinomas (epithelial–myoepithelial carcinomas, EMCs), both of which are malignant epithelial tumors. The pathological difference between the two is that MCs have only myoepithelial components, whereas EMCs contain glandular epithelial components in addition to myoepithelial components. In this case, only myoepithelial cells, not glandular epithelial component cells, were microscopic, so MC was diagnosed. The incidence of MC is not accurately known because of its rarity. Imaging characteristics of MCs have not been well-described in the literature, possibly due to their low incidence.[9] In our study, enhanced CT findings of retroperitoneal MC showed a round cystic solid mass with no enhancement in the cystic region and obvious enhancement in the solid region of the tumor. The solid region of the tumor was hypermetabolic and no hypermetabolism in the cystic region on PET-CT. Therefore, there is no specific imaging characteristic for the differential diagnosis of retroperitoneal MC, and there is an overlap between the benign and malignant presentation of these tumors, making it impossible to differentiate them using imaging alone. Hence, a biopsy is necessary to obtain pathological results.[10]

MC has moderate to high malignant potential.[6] MC has a high risk of local recurrence. Some studies[3,8] have shown that the local recurrence rate of MC ranges from 42% to 63%, and the median time to relapse is 47 months.[3] This patient experienced local recurrence more than 1 month after undergoing abdominal surgery. The patient had a retroperitoneal mass after five rounds of chemotherapy and six doses of targeted drugs combined with immunotherapy, which proved the significant efficacy of comprehensive treatment for MC. Some studies have reported that the therapeutic effect of chemoradiotherapy on MC is limited and not obvious.[11] However, in this case, after comprehensive treatment of the mass was significantly reduced, the curative effect was significant. We speculate that retroperitoneal mass control may be associated with targeted drug combination immunotherapy, namely, the 200 mg terellizumab + 400 mg bevacizumab treatment plan for the presence of the BCL2L11 (BIM) heterozygous mutation. EML 4-ALK fusion variant MC has a significant curative effect and can significantly reduce the size of the tumor. Consequently, our data suggest that targeted combined immunotherapy may have a role in palliating advanced retroperitoneal MC, which has not been reported in previous literature. In this case, the retroperitoneal MC recurred and progressed again after 4 months of comprehensive treatment. We believe that this was related to the higher degree of malignancy and higher risk of local recurrence of the retroperitoneal MC. During this period of treatment, the patient underwent four CT scans to monitor the mass changes and provide important imaging information guidance for the adjustment of a treatment plan.

Retroperitoneal MC should be differentiated from the following diseases. (1) Retroperitoneal neuroblastoma. In children, the disease is relatively common and mostly originates from the adrenal gland. Non-enhanced CT images revealed a homogenous density of round retroperitoneal mass on CT, and internal calcification was relatively common. Enhanced CT revealed progressive mild and moderate enhancement at the edge and interior of the lesion, and no enhanced necrotic cyst area could be seen at the center of the lesion.[12] Retroperitoneal neuroblastoma shows enhanced CT findings similar to those of retroperitoneal MC. However, calcification is more common with retroperitoneal neuroblastoma while no calcification occurs in a retroperitoneal MC. The age of the patient and the calcification of the lesion can be used to help identify the two diseases; however, final identification still depends on pathological and immunohistochemical examination. (2) Retroperitoneal liposarcoma (RPLS). RPLS is the most common retroperitoneal soft tissue sarcoma. RPLS varies greatly according to its classification, in which highly differentiated liposarcoma is rich in fat components, which is a typical imaging sign. Necrotic liquefaction can occur in dedifferentiated liposarcoma and pleomorphic liposarcoma, resulting in uneven enhancement in enhanced CT images and uneven metabolism in PET-CT images.[13] This increases the difficulty of differentiating RPLS from a retroperitoneal MC. (3) Retroperitoneal seminomas. Most patients with this disease have a history of testicular tumors. CT images reveal a large retroperitoneal promiscuous density mass in the lesion.[14] The final diagnosis is obtained through a combination of medical history and pathological examination. (4) Primary retroperitoneal teratomas (PRTs). Regarding imaging, the retroperitoneal mass contains fluid, fat, and calcification, while the mass is separated from other organs, such as the kidney, adrenal gland, and pancreas.[15] The components of PRT are complex and diverse and often less invasive than those of MC, which can be used as a differentiating point between the two diseases.

Conclusion

In conclusion, MC is a rare malignancy, with primary retroperitoneal MC being even rarer. Surgical resection is the preferred MC treatment, but the tumor is prone to recurrence after surgical resection. Comprehensive treatment is helpful for inhibiting tumor growth and prolonging the survival of patients. Postoperative pathology and immunohistochemistry are necessary, and imaging examination is of great value in the diagnosis and follow-up of retroperitoneal MC.

Abbreviations

CT, computed tomography; MC, myoepithelial carcinoma; MME, malignant myoepithelioma; EMCs, epithelial myoepithelial carcinomas; RPLS, retroperitoneal liposarcoma; PRTs, primary retroperitoneal teratomas.

Ethical Approval

Full institutional ethical approval was obtained before commencing this retrospective study.

Disclosure

Tong Wei and Hongmin Quan are co-first authors for this study. The authors report no conflicts of interest in this work.

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