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ABOUT COVER

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The primary aim of *World Journal of Gastrointestinal Surgery* (*WJGS*, *World J Gastrointest Surg*) is to provide scholars and readers from various fields of gastrointestinal surgery with a platform to publish high-quality basic and clinical research articles and communicate their research findings online.

WJGS mainly publishes articles reporting research results and findings obtained in the field of gastrointestinal surgery and covering a wide range of topics including biliary tract surgical procedures, biliopancreatic diversion, colectomy, esophagectomy, esophagostomy, pancreas transplantation, and pancreatectomy, *etc*.

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CASE REPORT

Massive simultaneous hepatic and renal perivascular epithelioid cell tumor benefitted from surgery and everolimus treatment: A case report

Han-Teng Yang, Fu-Rong Wang, Na He, Yuan-Hua She, Yong-Yue Du, Wen-Gui Shi, Jing Yang, Gang Chen, Shu-Ze Zhang, Feng Cui, Bo Long, Ze-Yuan Yu, Jun-Min Zhu, Geng-Yuan Zhang

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Abstract

BACKGROUND

Perivascular epithelioid cell tumor (PEComa) is a rare mesenchymal neoplasm that predominantly affects the kidney and uterus. The occurrence of this tumor in the liver, particularly with simultaneous involvement of the liver and kidney, is exceedingly uncommon. Pathological diagnosis is the gold standard. PEComas usually show positive immunohistochemical staining for melanocytic (HMB-45, Melan-A) and myoid (SMA, muscle-specific actin) markers.

CASE SUMMARY

We presented a noteworthy case of malignant PEComa affecting both the liver and kidney in a 53-year-old man with tuberous sclerosis complex (TSC). FAT2 and TP73 mutations in the kidney were identified and positive expression of diagnostic markers including HMB-45, Melan A, and TFE3 were detected. In addition, we demonstrated that hepatic artery perfusion chemotherapy was

ineffective for hepatic PEComa, while surgery remained the most effective approach. Everolimus showed an excellent efficacy in the postoperative treatment of the tumor.

CONCLUSION
Surgical treatment is preferred for malignant PEComa affecting liver and kidney, especially with TSC; everolimus is effective postoperatively.

Key Words: Perivascular epithelioid cell tumor; Hepatic; Renal; Surgery; Case report

Core Tip: Perivascular epithelioid cell tumor (PEComa) is rare. Herein, we present the case of a 53-year-old man with the concurrent presence of malignant PEComa in both the liver and kidney, accompanied by tuberous sclerosis complex. During treatment, the hepatic artery perfusion chemotherapy was ineffective for hepatic PEComa, while surgery remained the most effective approach. Everolimus showed an excellent efficacy during the postoperative treatment of the patient. This report provides valuable insights for the diagnosis and treatment of PEComa, paving the way for future clinical practice.

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INTRODUCTION

Perivascular epithelioid cell tumor (PEComa) is a rare mesenchymal neoplasm, belonging to a member of the family (PEComas) including lymphangioleiomyomatosis, classic angiomyolipoma (AML) and clear epithelioid cell tumors. It is most frequently detected in the retroperitoneum, peritoneal and pelvic cavities, and occasionally found in somatic soft tissues and the skin[1,2]. The female-to-male ratio is approximately 6: 1 and it is more common in young and middle-aged patients[3]. This tumor, an epithelioid AML, mainly consists of epithelioid cells, and lacks typical fat tissues[4]. It is typically accompanied with focal infiltration of blood vessel walls and the cells are often arranged radially around the vascular lumen[5]. PEComas usually show immunoreactivity for melanocytic (HMB-45, Melan-A) and myoid (SMA, muscle-specific actin) markers. Melan-A is expressed in more than 50% cases and approximately 25% of patients show desmin expression[4]. Compared with other markers, positive HMB-45 is often considered the gold standard for the diagnosis of AML histopathologically[6]. Most patients are asymptomatic or complain of non-specific gastrointestinal manifestations. The tumor is usually of a large size and holds the malignant potential. The more aggressive are less common[7]. Here, we present a case with the concurrent presence of malignant PEComa in both the liver and kidney, accompanied by tuberous sclerosis complex (TSC).

CASE PRESENTATION

Chief complaints

A 53-year-old man with a one-year history of intermittent abdominal distention and anorexia was hospitalized, who also experienced fatigue and shortness of breath for 2 months.

History of present illness

To control tumor progression and alleviate discomfort, we attempted to perform the hepatic artery perfusion chemotherapy with oxaliplatin, leucovorin and fluorouracil after obtaining informed consent from the patient and his family. Surprisingly, the size of the liver tumor increased from 14.0 cm × 16.0 cm before treatment (Figure 1) to 14.5 cm × 18.5 cm after treatment (Figure 2). And tumor vessels were observed (Figure 3). During hospitalization, the patient's chest tightness and shortness of breath worsened and blood pressure was elevated intermittently, reaching up to 145/96 mmHg. Blood pressure was controlled with oral administration of phenoxybenzamine (10 mg, twice a day) and nifedipine (30 mg, once a day).

History of past illness

The patient had no history of diabetes, hypertension or other cardiovascular diseases.

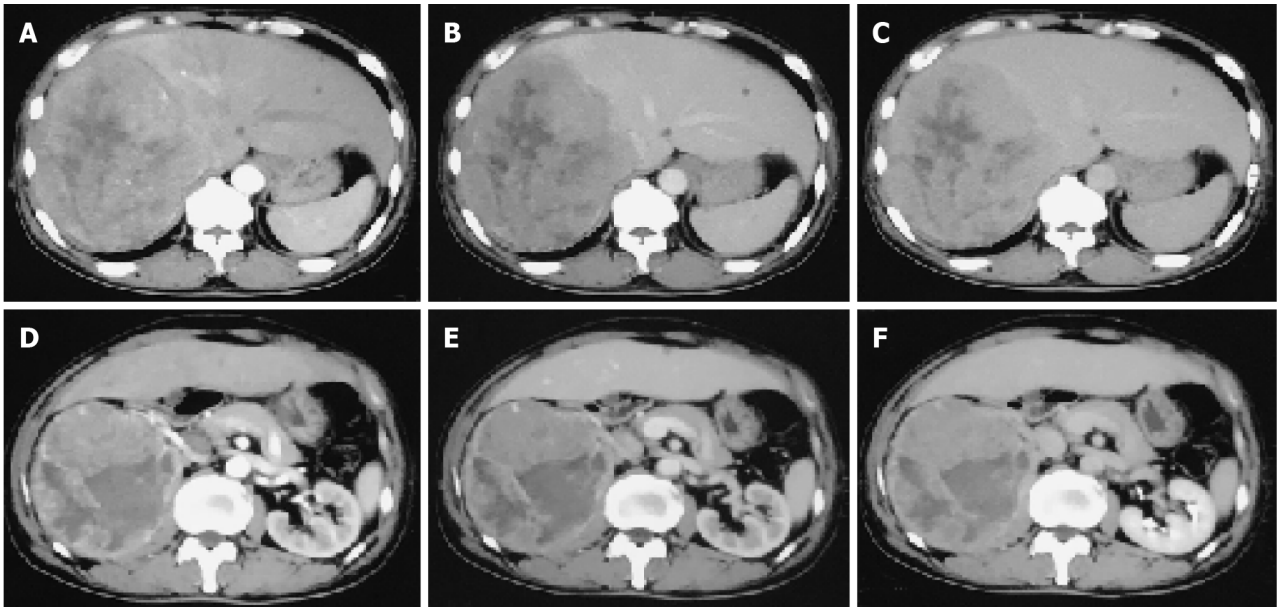

Figure 1 Images of the patient before hepatic artery perfusion chemotherapy. A: Contrast-enhanced computed tomography (CT) scans showed a well-circumscribed mass with multiple low-density patchy shadows, measuring 140 cm × 16.0 cm in the right lobe of the liver during the arterial phase; B: Contrast-enhanced CT scans in the right lobe of the liver during the portal phase; C: Contrast-enhanced CT scans in the right lobe of the liver during the delayed phase; D: Contrast-enhanced CT scans showed a distinct soft tissue mass with clear-boundaries and low-density foci, measuring 115 cm × 12.0 cm, was identified in the upper pole of the right kidney during the arterial phase; E: Contrast-enhanced CT scans in the upper pole of the right kidney during the portal phase; F: Contrast-enhanced CT scans in the upper pole of the right kidney during the delayed phase.

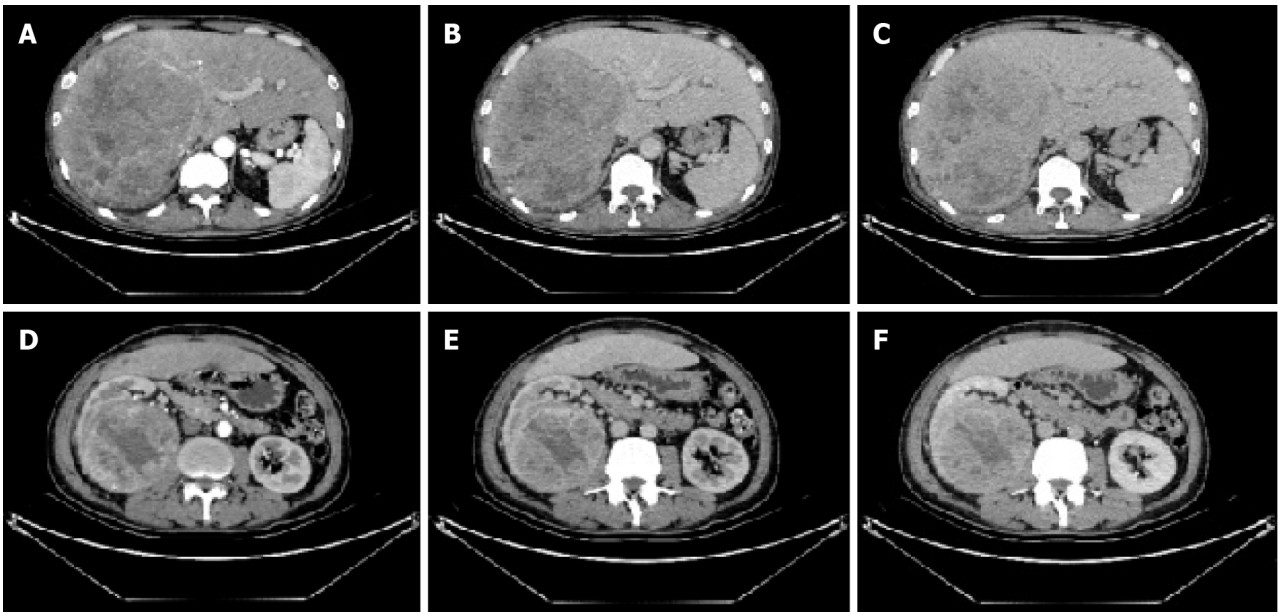

Figure 2 Images of the patient after hepatic artery perfusion chemotherapy. A: Contrast-enhanced computed tomography (CT) scans showed a well-circumscribed mass with multiple low-density patchy shadows, measuring 145 cm × 18.5 cm × 26.4 cm in the right lobe of the liver during the arterial phase; B: Contrast-enhanced CT scans in the right lobe of the liver during the portal phase; C: Contrast-enhanced CT scans in the right lobe of the liver during the delayed phase; D: A distinct soft tissue mass with clear-boundaries and low-density foci, measuring 116 cm × 11.9 cm × 12.2 cm, was identified in the upper pole of the right kidney during the arterial phase; E: Contrast-enhanced CT scans in the upper pole of the right kidney during the portal phase; F: Contrast-enhanced CT scans in the upper pole of the right kidney during the delayed phase.

Personal and family history

The patient denied any family history of malignant tumors and TSC.

Physical examination

Physical examination revealed space-occupying lesions with non-tender, flexible texture, poor mobility and blurred boundaries in the right upper abdomen. Many depigmented macules were found in multiple regions of the body,

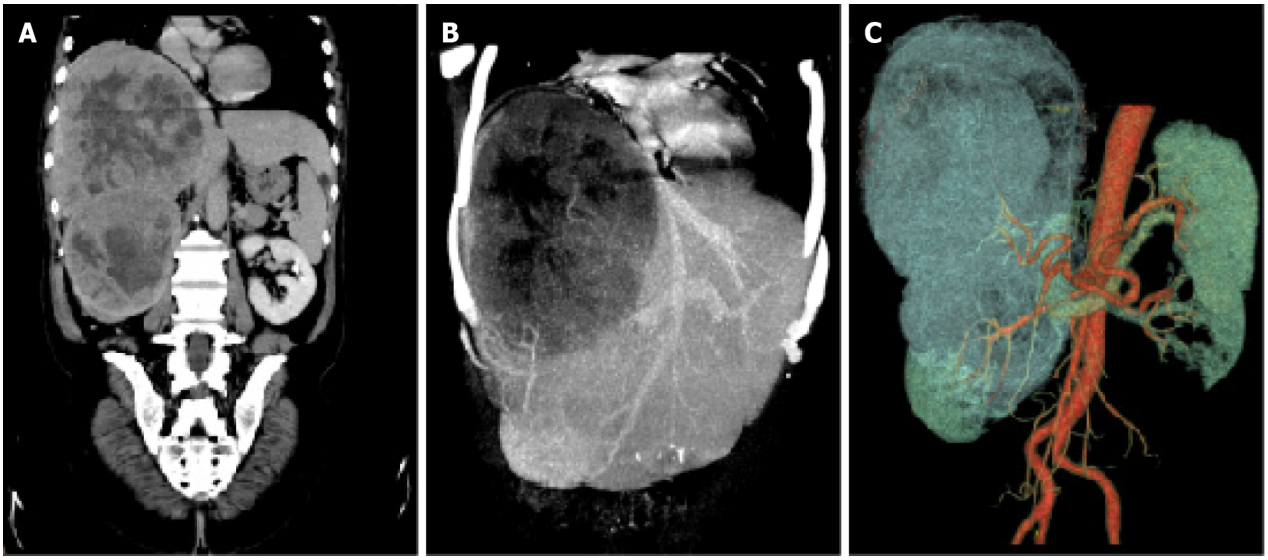

Figure 3 Images of tumor vascular supply. A: The computed tomography reconstructions displayed a large well-circumscribed mass with uneven density in the right lobe of the liver and a clear-boundary and round-like neoplasm in the right kidney at the coronal level; B: In the maximum intensity projection images, there was no visualization of the right hepatic vein and the middle hepatic vein appeared compressed and deformed due to the presence of the tumor; C: The volume rendering technique images revealed that the hepatic tumor was supplied by branches of the proper hepatic artery and the renal neoplasm received blood supply from branches of the renal artery.

especially in the abdomen (Figure 4).

Laboratory examinations

Laboratory results showed an albumin concentration of 30.5 g/L, lactate dehydrogenase level of 665 U/L (120-250), interleukin-6 level of 207.00 pg/mL (0.00-7.00), PO_2 of 66 mmHg (80-100), SO_2 of 93% (95%-98%), methoxynorepinephrine level of 72.9 pg/mL, methoxyepinephrine level below 18.0 pg/mL and dopamine level of 35.9 pg/mL. Tumor markers revealed elevated levels of CA199 48.9 U/mL (0.00-27.00 U/mL), CA72-4 9.42 U/mL (0.00-6.90 U/mL) and neuronal-specific enolase 119.0 ng/mL (0.00-15.20 ng/mL).

Imaging examinations

He had undergone contrast-enhanced computed tomography (CT) scans of the abdomen, which confirmed the presence of neoplasms located in the right lobe of the liver and the upper pole of the right kidney (Figure 1).

FINAL DIAGNOSIS

Based on various clinical data, including patient complaints, physical, laboratory and imaging examinations, and pathology and immunohistochemistry findings, the patient was diagnosed with malignant PEComa.

TREATMENT

The patient underwent open surgical resection, including right hemihepatectomy, right nephrectomy and right adrenalectomy. Gross examination of the specimens revealed a liver tumor measuring approximately 24 cm × 17 cm × 14.5 cm and a kidney tumor measuring approximately 12 cm × 10 cm × 11.7 cm, both with gray-yellow or gray-red surfaces and focal hemorrhage (Figure 5). Finally, the patient recovered well and was discharged on the 15th postoperative day.

OUTCOME AND FOLLOW-UP

Histopathological analysis of surgical specimens showed that the tumors in the liver and kidney were about of the same type. The tumor cells were diffusely distributed, forming a sheet-like structure and characterized by epithelioid cell differentiation. The nuclei were enlarged, vacuolar, and showed overt atypia. In addition, the cytoplasm were abundant. The cells were strongly positive for HMB45 and Melan A, weakly positive for TFE3, but negative for Vimentin, SMA, Desmin, Calponin, S-100, EMA, PAX2, PAX8, CD10, CD34, CD117 and CK7. The Ki67 proliferation index was approximately 2% (Figure 6). The second-generation sequencing identified FAT2 and TP73 mutations in the kidney (Table 1).

Table 1 The second-generation sequencing technologies applied to the surgical specimens from the liver and kidney revealed mutated gene loci

Gene	Transcript	Mutated base	Changed amino acid	Functional region	Mutation frequency (%)
FAT2	NM_001447.2	c.5986C>T	p.H1996Y	EX9	1.8
TP73	NM_005427.3	c.94C>T	p.P32S	EX3	1.1

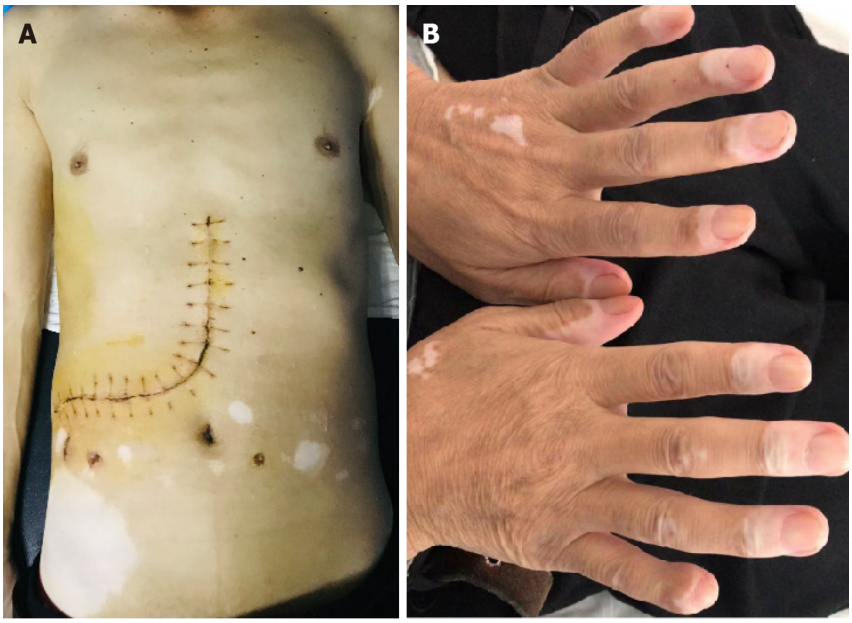

Figure 4 Depigmented macules. A: They were observed in parts of axillary region and abdomen; B: They were observed in parts of fingers and hand dorsum.

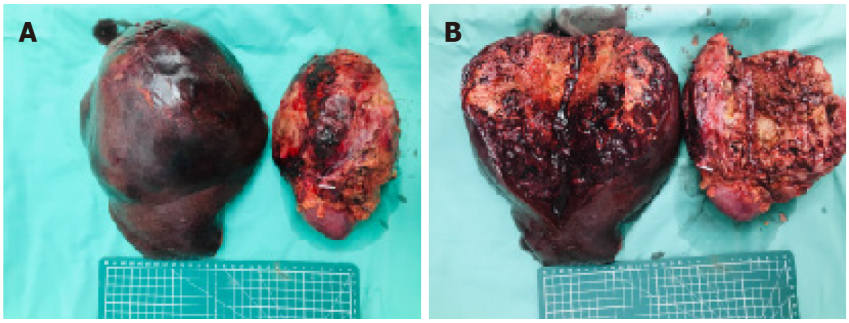

Figure 5 Images of surgical specimens. A: The neoplasm in the liver was approximately 24 cm × 17 cm × 14.5 cm in size and the neoplasm in the kidney measured approximately 12 cm × 10 cm × 11.7 cm on the gross examination of the surgical specimens; B: Upon sectioning, both tumors exhibited focal haemorrhage and displayed gray-yellow or gray-red colors.

Approximately 1 year after discharge, a follow-up abdominal CT revealed no aberrant lesions in the surgical area (Figure 7). During the 1.5-year follow-up period, the patient took oral everolimus without experiencing obvious discomfort.

DISCUSSION

Malignant PEComa is extremely rare, accounting for only 4.6% cases of AML[7]. Compared to classic AML, PEComa is typically larger in size and contains less-fat, often accompanied by necrosis and hemorrhage[8]. When 2 or more of these characteristics are present, such as epithelioid cells ≥ 70%, tumor size > 5 cm, mitoses per 10 high-powered fields ≥ 2, aberrant mitosis and necrosis, and tumor-infiltrating growth and metastasis, the tumor is considered to behave in a highly malignant manner[9,10]. In a study of 36 patients, Tirumani *et al*[11] found that malignant PEComa tends to be a well-defined heterogeneous neoplasm. However, the diagnosis of PEComa can be challenging due to its rarity and lack of

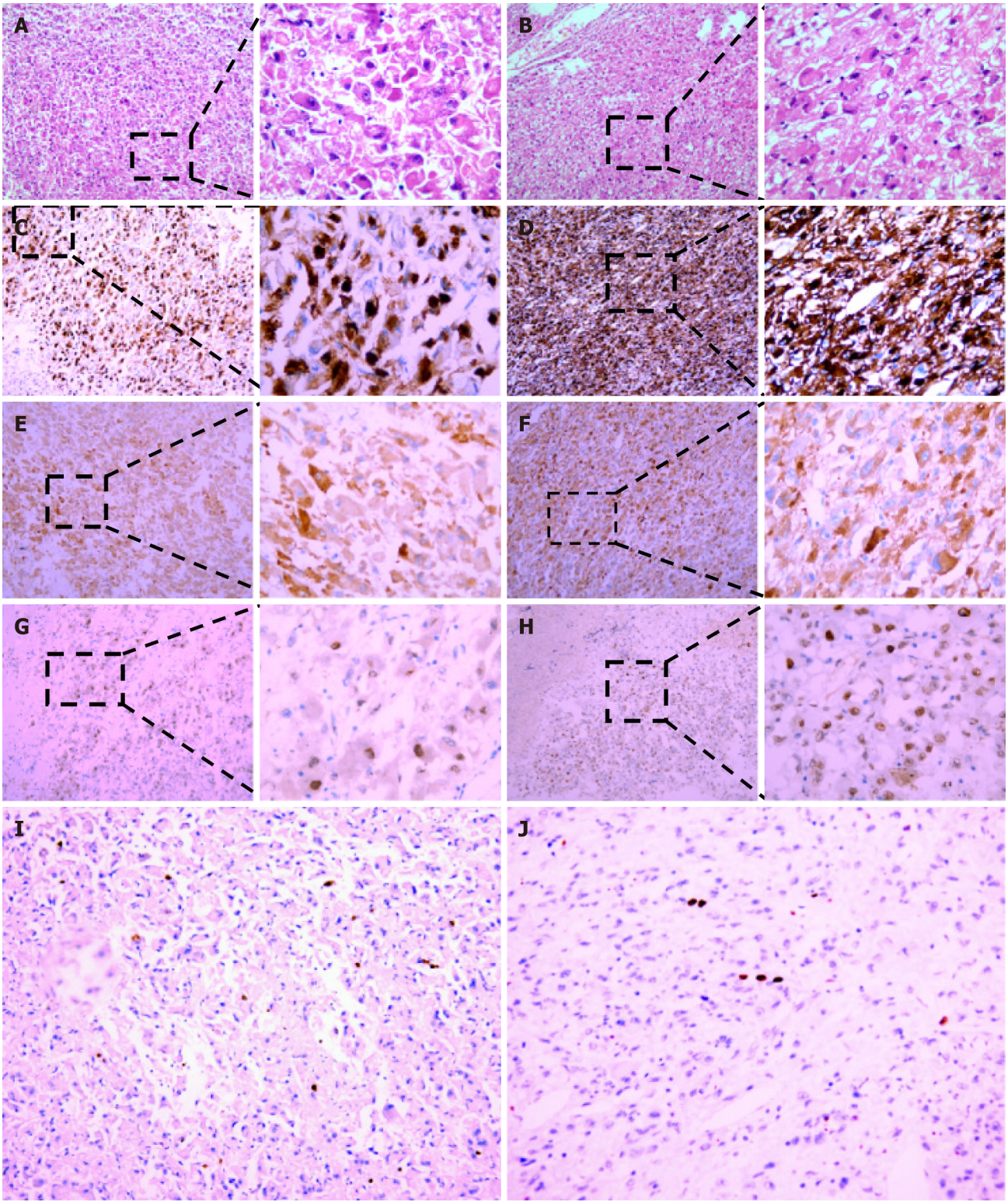

Figure 6 Pathological findings of surgical specimens. A and B: The histopathological examination of surgical specimens from liver and kidney lesions showed tumor lesions mainly composed of dysplasia epithelioid cells with rich and red-stained cytoplasm and large, atypical and vacuolated nuclei, and prominent nucleolus (hematoxylin-eosin, original magnification × 100 and × 400); C and D: The surgical specimens from the liver and kidney lesions showed strong and diffuse granular cytoplasmic immunoreactivity for HMB-45 in epithelioid tumor cells (original magnification × 100 and × 400); E and F: The surgical specimens from the liver and kidney lesions exhibited intense and partly cytoplasmic expression of melan-A, with a staining pattern similar to that of HMB-45 (original magnification × 100 and × 400); G and H: The surgical specimens from the liver and kidney lesions revealed weak and punctate nuclear immunoreactivity for TFE3 in the epithelioid tumor cells (original magnification × 100 and × 400); I and J: The surgical specimens from the liver and kidney lesions displayed weak expression of Ki67 in the nuclei of the epithelioid cells (original magnification × 100).

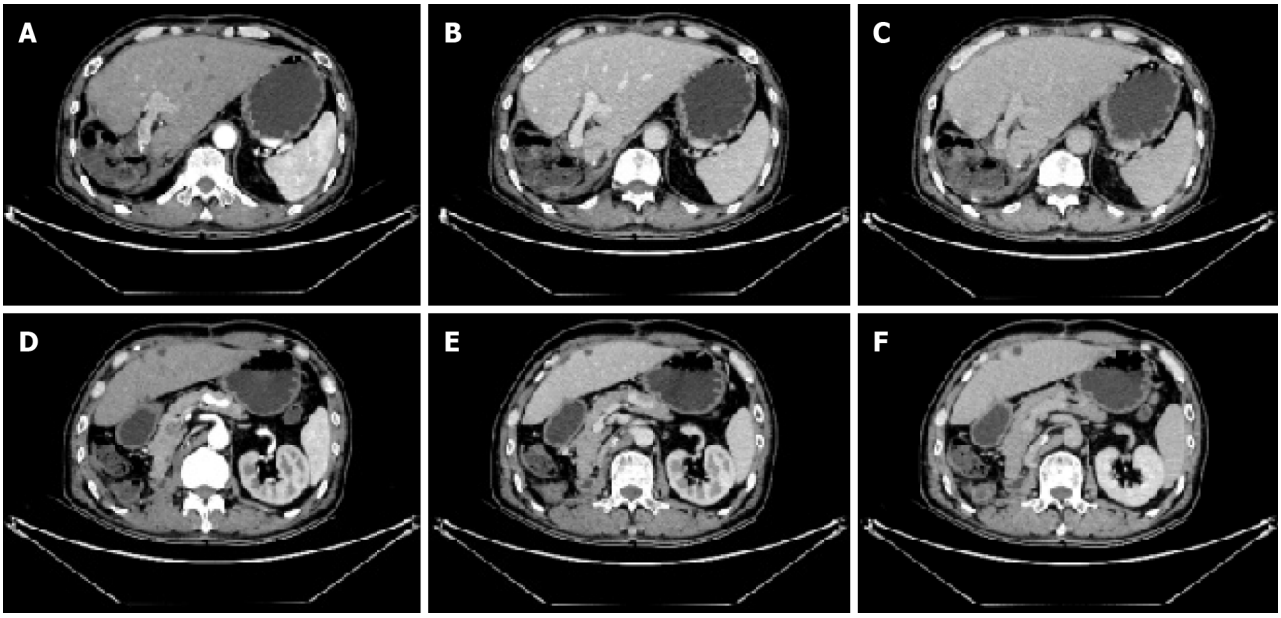

Figure 7 Images of the patient 1 year after discharge. A: Contrast-enhanced computed tomography (CT) scans revealed banded low-density shadows in the liver surgical area during the arterial phase; B: Contrast-enhanced CT scans in the liver surgical area during the portal phase; C: Contrast-enhanced CT scans in the liver surgical area during the delayed phase; D: Contrast-enhanced CT scans revealed banded low-density shadows in the right kidney surgical area during the arterial phase; E: Contrast-enhanced CT scans in the right kidney surgical area during the portal phase; F: Contrast-enhanced CT scans in the right kidney surgical area during the delayed phase.

typical imaging characteristics. Kidneys and uterus are the most commonly affected organs, while involvement of the liver, especially simultaneous involvement of both the liver and kidney, is exceedingly rare[9,12]. Renal PEComa can be difficult to differentiate from renal cell carcinoma, lymphoma and transitional cell carcinoma. Uterine PEComa should be discriminated from fibroids and leiomyosarcomas. Hepatic PEComa is easily misdiagnosed as hepatocellular carcinoma, hepatic hemangioma, hepatic sarcoma, gastrointestinal stromal tumor, or other types of tumors. The diagnostic accuracy of hepatic PEComa using CT combined with MRI is only 20%[9]. Fine-needle aspiration and immunohistochemical staining for HMB45 and Melan-A are necessary for a definitive diagnosis. Patients with PEComa often presented to the clinic, due to weight loss, fatigue, abdominal discomfort or shortness of breath. Approximately one-third of patients may develop metastatic lesions in the liver, lung, and peritoneum/mesentery[4]. It is consistent with the findings of Nese *et al* [13]. In their study, the recurrence and mortality rates of PEComa patients were 17% and 33%, respectively, during a median follow-up of 24.5 months[13]. The malignancy of PEComa may be associated with vascular invasion and certain genetic changes. However, the detailed mechanism remains to be further explored.

TSC is a rare autosomal dominant disorder characterized by inactivating mutations in the *TSC1* and *TSC2* genes encoding the proteins hamartin and tuberin. Patients with TSC are predisposed to multiple tumor or cyst diseases, occurred frequently in central nervous system, skin, and kidney[14]. The incidence of TSC was reported to be around 1 in 6000-10000 individuals, and TSC-related lesions increase with age[6,15]. Renal involvement is present in approximately 60%-80% of TSC cases, with renal AML being the most common[14,16]. But the probability of having TSC in patients with renal AML is less than 20%[17]. Hepatic AML is uncommon, accounting for 6%-10% of TSC cases[18]. However, the incidence of hepatic AML has been reported to be as high as 13%-21% in female TSC patients with bilateral renal AML [19]. For the malignant PEComa affecting both the liver and kidney, reports are even rarer. It is well established that most PEComas are characteristic of TSC genetic alterations[20]. TSC2 mutations were detected in 80% of TFE3-negative PEComas[21]. In addition, mTOR inhibitors, such as everolimus or temsirolimus, are beneficial for some patients with advanced or recurrent PEComas[4,7,17,22].

In this study, we present a rare case of malignant PEComa affecting both the liver and kidney in a patient with TSC. FAT2 and TP73 mutations in the kidney were identified. Due to lower mutation frequencies of these genes, no relevant important targets for PEComa were detected. The tumor showed positive expression for markers including HMB-45, Melan A, and TFE3. TSC1 and TSC2 mutations were not detected in this case. The hepatic artery perfusion chemotherapy is not effective and the surgery is the most effective. During a 1.5-year postoperative follow-up period, the patient recovered well after long-term oral administration of everolimus.

CONCLUSION

The malignant PEComa affecting both the liver and kidney, especially accompanied by TSC, is exceedingly rare. Herein, we provided a valuable case study suggesting that the interventional therapy is undesirable and the surgical treatment is a better choice. In addition, we demonstrated that everolimus is highly effective in the postoperative treatment of this

tumor, indicating the tight association of PEComa with TSC. However, the pathogenic mechanism of PEComa remains poorly defined and further studies are required.

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FOOTNOTES

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