Hepatic perivascular epithelioid cell tumor treated by transarterial embolization plus radiofrequency ablation

A case report and literature review

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Abstract

Background: Perivascular epithelioid cell tumors (PEComas) are extremely rare mesenchymal entities with potentially malignant properties; the liver cases are not encountered frequently. Owing to themalignant potential, these tumors are treated by surgical methods to ensure total resection. In the present report, a case of liver PEComa treated by embolization combined with radiofrequency ablation (RFA) has been described.

Case summary: A 40-year-old female was admitted for the detection of a liver mass during an annual physical examination. The patient did not have any liver disease background, enhanced computed tomography (CT), and magnetic resonance revealed a huge mass in the right lobe. Pathology gave the diagnosis of PEComa, for disagreement of open surgery, a combination of transarterial embolization (TAE) and RFA were applied for treatment and the outcomes were acceptable, the patient was under follow-up to observe the long-term effect.

Conclusion: Interventional procedures such as TAE and RFA are feasible and effective for such lesions and may serve as an alternate when resection is not indicated. Prospective studies are warranted to verify the long-term outcomes.

Abbreviations: CT = computed tomography, HCC = hepatocellular carcinoma, PEComas = perivascular epithelioid cell tumors, RFA = radiofrequency ablation, TAE = transarterial embolization.

Keywords: HMB45, liver, perivascular epithelioid cell tumor, radiofrequency ablation, SMA, transarterial embolization.

1. Introduction

Perivascular epithelioid cell tumors (PEComas) comprise a family of solid tumors that exhibit perivascular epithelioid cell differentiation with low morbidity. In 2002, the World Health Organization presented identifying guidelines for the mesenchymal neoplasms, containing a series of morphologically and immunophenotypically similar lesions.1 These tumors may involve multiple organs, including lung, kidney, liver, somatic soft tissue, or skin.2–4 A subset of these lesions behaves in a malignant manner. Therefore, the therapeutic priority is given to both total surgical resection and pathological diagnosis simultaneously and previously reported cases yielded promising outcomes.5–10 However, open surgery may not be favorable due to contraindications or patients’ disagreement. Transarterial embolization (TAE), also known as bland embolization, might be employed as a palliative treatment for unresectable hepatocellular carcinoma (HCC), whereas radiofrequency ablation (RFA) serves as a curative treatment in early cases; a combination treatment is also conducted specifically. Nevertheless, such an application in PEComas is not yet reported. Herein, the therapeutic process and preliminary results during the treatment of a liver PEComa are presented.

2. Case presentation

Institutional review board approved this case report, and informed consent was obtained before treatment. A 40-year-old female was admitted to the Peking University First Hospital as an outpatient presenting a liver mass that was detected during an annual physical examination in September 2013. The patient did not show any symptom-related discomfort or history of hepatitis B or C, cirrhosis background, and alcohol abuse. The laboratory tests including blood routine, liver, and renal function, and tumor biomarkers including AFP, CEA, and CA19–9 were found to be within the normal range. An abdominal computed tomography (CT) scan performed at the local hospital revealed a circular, space-occupying lesion with a maximum diameter of 7.5 × 5.0 cm. The lesion was localized primarily in segment 8 of the right lobe that was hypodense on a normal scan with apparent early arterial-phase enhancement and delayed-phase washout (Fig. 1A, B). An enhanced magnetic resonance scan revealed an inhomogeneous signal in the mass and thick draining veins in the venous phase (Fig. 2). Based on these imaging findings and medical history, the suggested primary tumor was adenoma or atypical HCC;
therefore, a needle biopsy was performed to facilitate a definite diagnosis. The pathological and results revealed a disorderly arrangement of epithelioid and spindle-like cells. On the other hand, immunohistochemistry demonstrated a strongly positive human melanoma black and smooth muscle actin staining (Fig. 3A, B), thereby suggesting a diagnosis of PEComa. Despite a comprehensive consultation, the patient was unwilling to accept open surgery. Thus, we selected TAE instead of TACE for primary treatment according to the imaging features and the absence of specific sensitive chemotherapeutics. Due to the lesion size larger than 7cm in diameter, RFA was not the first choices of treatment. The TAE procedure was normal, angiography revealed a tumor with abundant blood supply from the superior mesenteric artery, with an enlarged feeding artery, and draining vein to the inferior vena cava. A 2.8F microcatheter (ProgreatTerumo, Japan) was applied for super selection ascribed to the abundant blood supply (Fig. 4A). A mixture of 10mL lipiodol plus 6mL ethanol and 500 to 700μm polyvinyl alcohol particles were successively delivered through the catheter to ensure total embolization. A 2nd angiography demonstrated a cut-off of the target vasculature without tumor staining (Fig. 4B), and the patient was discharged after a 3-day observation period without any discomfort.

Two weeks later, the patient was readmitted for hyperpyrexia with the highest temperature at 39.3°C accompanied by hepatalgia. Routine blood and biochemical tests revealed a normal range of the white blood cell count, a slightly elevated AST level at 45 IU/L, and a decreased albumin at 28.6 g/L. A repeat of the abdominal CT scan revealed an abscessed lesion with an air-fluid level within the original mass (Fig. 5A). Consequently, a percutaneous transhepatic puncture was fashioned via an inserted 8.5F drainage catheter (Cook, Bloomington, Indiana) for abscess draining. The symptoms were remitted promptly, and the patient was discharged after a 5-day hospital stay with normal temperature and results of laboratory tests. After 14 days, the tube was removed by the outpatient clinic postultrasonic confirmation of complete drainage.

The 3-month review of CT scan in January 2014 revealed small nodular residual lesions along the edge of the original mass (Fig. 5B). Hence, an RFA procedure was implemented via percutaneous transhepatic approach with a CelonProSurge needle (Olympus Winter & Ibe, Germany), and no adverse event occurred. A 1-month follow-up CT scan showed complete necrotic lesions without arterial phase enhancement (Fig. 6). Beginning with the 1st round of treatment, the patient was under follow-up for over 3 years until present. A half-year interval review by enhanced CT or ultrasound did not reveal any recurrence or metastasis; however, repeated TAE or RFA would be taken into account as necessary.

3. Discussion

Given the peculiar rarity, the precise morbidity and mortality of liver PEComa cannot be well-established from the current data, and thus, the pathogenesis of this disease remains controversial[7]. The summary of the previously reported cases led to identifying basic features such as a female predominance and an abundant blood supply, which were evident during enhanced imaging examinations. These characteristics are speculated to associate with hormonal changes, similar to hemangioma or adenoma[8]. No specific clinical presentations or laboratory results for diagnosis were displayed in the present case; the tumor was detected incidentally, and the patient was asymptomatic. However, the symptomatic disease can occur in
some cases suffering from ambiguous pain or abdominal distension.[9,10]

An imaging study for the preoperative diagnosis of PEComa comprises of ultrasonography, enhanced CT, or magnetic resonance. Some common manifestations have been described previously, including a well-defined border, obvious enhancement following contrast administration with early washout, hypervascularity, and arteriovenous fistula.[11,12] These phenomena are consistent with our findings; however, the diagnostic accuracy is low attributed to nonspecificity with other lesions such as HCC, hepatic adenoma, and focal nodular hyperplasia. Despite angiography, as in the present case, a typical hallmark was yet absent.
Ultimately, a diagnosis of PEComa mainly relies on the pathological and immunohistochemistry results that indicate a tumor comprising of an epithelioid appearance and spindle-like cells with eosinophilic cytoplasm haphazardly arranged around the thin-walled vessels. These tumors are usually highly vascularized but dysplastic, which may explicate the imaging presentations of apparent enhancement or abnormal feeding and draining vessels. The immunohistochemistry demonstrated almost all PEComas with strong positive human melanoma black and smooth muscle actin staining, which might indicate a characteristic phenotype of differentiation.

Most PEComa cases have exhibited benign behavior, although malignant cases have also been documented in recent years. Therefore, PEComa should be identified as a potentially malignant disease and treated positively. Surgical resection currently remains the primary treatment, and promising outcomes have been reported. In the present case, TAE plus RFA was selected because: 1st, the patient was resistant to resection from the heart despite comprehensive consultation; 2nd, the imaging findings showed rich blood supply that indicated transarterial therapy. Moreover, no sensitive chemotherapeutic agents were documented for PEComa still date, and hence, ethanol was selected; 3rd, after the necrotic tissue was discharged, the residual nodule was small and could be a candidate for RFA to achieve the curative effect. This combination has already been applied in some early stage HCC patients, and the outcomes were promising.

Concerning the interventional procedure, several factors may influence the therapeutic outcomes, such as the tumor size, trait of blood supply, and embolic agents used. Generally speaking, a tumor mass with rich blood vascular will be easier to get a total embolization, thus give rise to complete necrosis. The embolic agents including type and size are supposed to be suitable in light of the result of angiography. One unanticipated and notable condition in this case was that liquefaction occurred following the first round of treatment, which may be induced by a rapid and radical necrosis and seemed as an adverse event. Thus, we speculate that the prompt disintegration may be attributed to the sponginess of the tumor tissue, with increased sensitivity to ischemia. However, whether this represents a dogma caused by ethanol injection or merely an accidental infection event remains uncertain.

Conversely, none of the bacterial culture and routine blood test results supported an infectious disease, and thus, to identify the dependence of blood supply to PEComas and sensitivity to TAE necessitate additional studies. To our knowledge, there have been no previous studies reporting similar lesions treated by the interventional approach. The present case suggested TAE and/or RFA as the alternatives for patients not suitable for resection.

In conclusion, PEComa of the liver is a rare disease that has been increasingly recognized in recent years, pathology and immunohistochemistry are required to make a confirmative diagnosis. Although resection remains the primary curative method, the interventional procedure, which could not be established as a standard therapeutic regimen hitherto, is a feasible and effective treatment that may serve as an alternative when resection is not indicated. Thus, prospective studies are warranted to verify the long-term outcomes of interventional therapy.

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Figure 6. One-month review after radiofrequency ablation (RFA) showed a total necrotic lesion without arterial phase enhancement (arrow).