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CASE REPORT

# Hepatic angiosarcoma mimicking hepatic epithelioid hemangioendothelioma: report of a case

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**Abstract** A 78-year-old male was admitted to our hospital for treatment of multiple hepatic tumors, which were suspected as hepatic epithelioid hemangioendothelioma (HEHE) by percutaneous tumor needle biopsy. With a diagnosis of HEHE, the patient underwent hepatic resection. In pathological findings, the tumor was composed of numerous endothelial cells without epithelioid cells, and was diagnosed as hepatic angiosarcoma (HAS). The patient received percutaneous radiofrequency ablation for recurrent HAS at 5 and 25 months postoperatively, and remains well with recurrence as of 28 months after the primary hepatic resection. In spite of improvement in radiological imaging, preoperative differential diagnosis between HAS and other malignant vascular tumors of the liver is still difficult. We herein report a case of HAS mimicking HEHE, treated successfully.

**Keywords** Hepatic angiosarcoma · Hepatic epithelioid hemangioendothelioma · Hepatic resection

## Introduction

Hepatic angiosarcoma (HAS) is a rare primary mesenchymal tumor, accounting for less than 2 % of all primary liver tumors, with a variety of findings on radiological imaging [1, 2]. We herein report a case of HAS mimicking hepatic epithelioid hemangioendothelioma (HEHE).

## Case report

A 78-year-old male was admitted to our hospital for treatment of multiple hepatic tumors, which were suspected as HEHE by percutaneous tumor needle biopsy with positive vascular markers including CD31, CD34, and factor VIII-related antigen. The patient had a past medical history of diabetes mellitus and chronic alcoholic pancreatitis with pancreatic pseudocysts treated at a local hospital. Serum tumor markers were as follows: α-fetoprotein 2.9 ng/ml, protein induced by vitamin K absence or antagonist-II 22 mAU/ml, carcinoembryonic antigen 3.5 ng/ml, and carbohydrate antigen 19–9 31 U/ml. Gadoxetic acid (Gd-EOB-DTPA)-enhanced magnetic resonance imaging (MRI) revealed high intensity tumors in segment 3 (Fig. 1a; arrow), 4 (Fig. 1b; arrow), 5 (Fig. 1c; arrow), 6 (Fig. 1c; arrowhead), and 7/8 (Fig. 1d; arrow) of the liver on T2-weighted images. On follow-up MRI 2 months later, the tumor in segment 3 (Fig. 2a; arrow) increased in size, but the tumors in 4 (Fig. 2b; arrow), 5 (Fig. 2c; arrow), 6 (Fig. 2c; arrowhead), and 7/8 (Fig. 2d; arrow) became slightly smaller. Dynamic enhanced computed tomography (CT) detected only well-circumscribed enhancing tumors with a diameter of 30 mm in segment 3, and 40 mm in segment 7/8. With a diagnosis of HEHE, the patient underwent extended posterior segmentectomy including the dorsal half of the anterior segment, subsegmentectomy of segment 3, partial resection of segment 4 and 5, and cholecystectomy. The duration of the operation was 342 min and intraoperative blood loss was 460 g. In the resected specimen, macroscopic findings revealed multiple, solid, hepatic tumors with the largest diameter of

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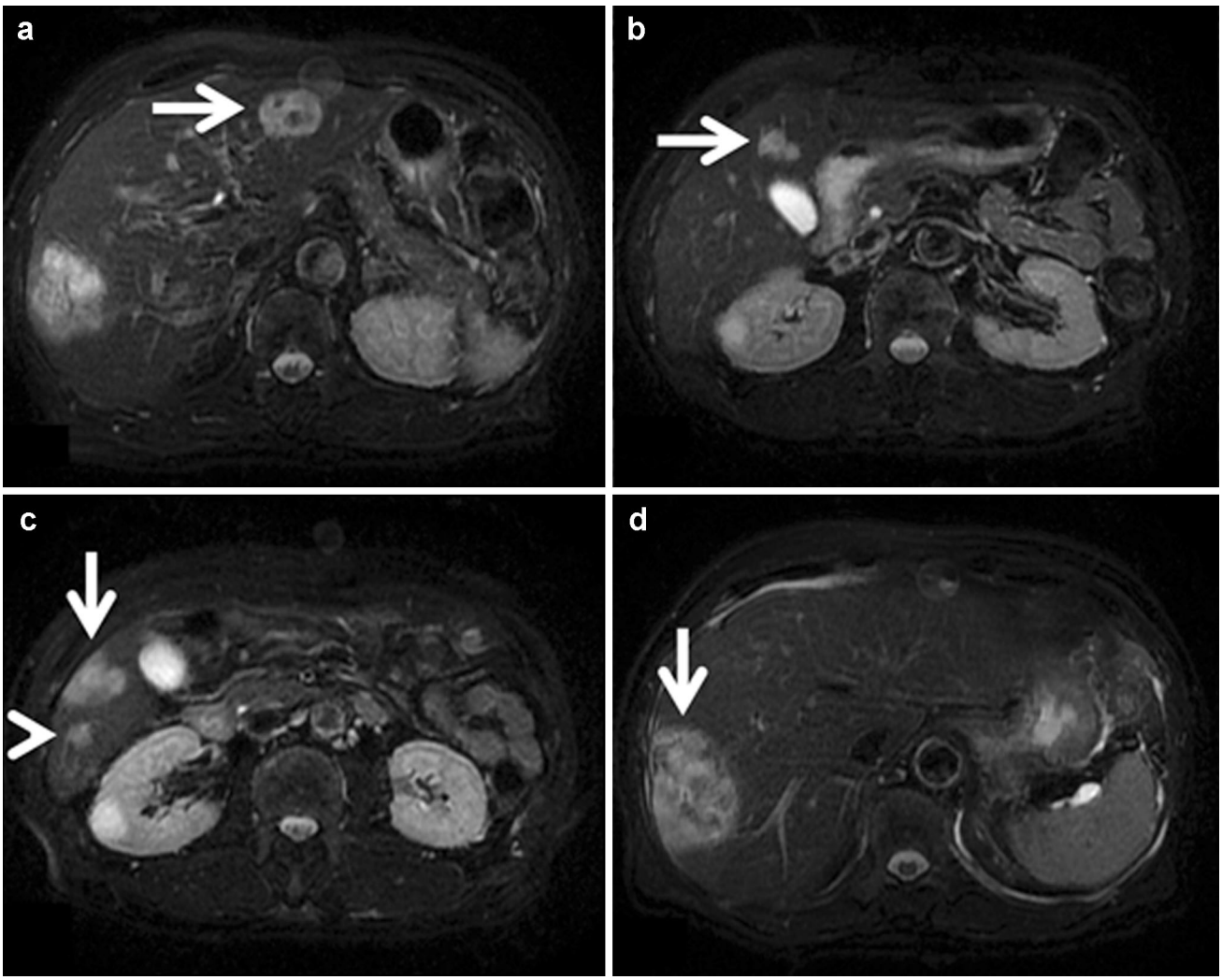


**Fig. 1** Magnetic resonance imaging revealed high intensity tumors in segment 3 (**a**; *arrow*), 4 (**b**; *arrow*), 5 (**c**; *arrow*), 6 (**c**; *arrowhead*), and 7/8 (**d**; *arrow*) of the liver on T2-weighted images

42 × 37 × 35 mm. In pathological findings, the tumor was composed of numerous endothelial cells without epithelioid cells (Fig. 3a), with positive vascular markers including CD31 (Fig. 3b), CD34 (Fig. 3c), and factor VIII-related antigen (Fig. 3d). Pathological diagnosis of the liver tumor was hepatic angiosarcoma. The patient made a satisfactory recovery, and was discharged 9 days after resection. The patient received percutaneous radiofrequency ablation (RFA) for recurrent HAS in segment 8 at 5 months, and segment 2 and 8 at 25 months after hepatic resection, respectively. After the RFA, the patient remains well with recurrence as of 28 months after the primary hepatic resection.

## Discussion

HAS accounts for one-third of primary hepatic sarcomas and commonly affects patients between 50 and 70 years of age [3]. HAS is composed of variably differentiated endothelial cells that are positive for vascular markers including CD31, CD34, and factor VIII-related antigen stain. Arsenic, thorium dioxide, and polyvinyl chloride are known risk factors, but most of HAS are classified as idiopathic [1].

HAS has a variety of findings on enhanced CT and MRI [2]. In spite of improvement in radiological imaging, preoperative differential diagnosis between HAS and other

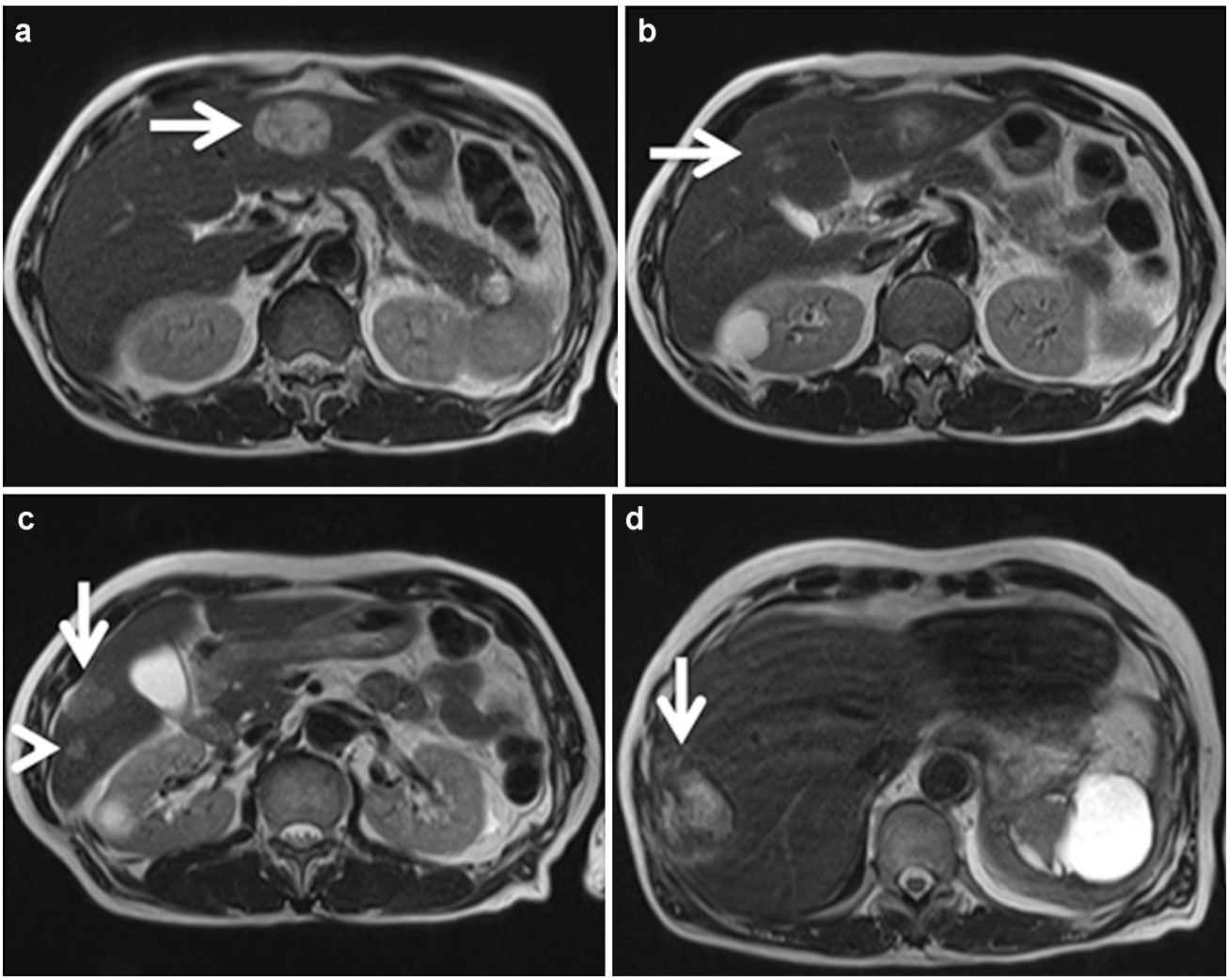


**Fig. 2** On follow-up magnetic resonance imaging, the tumor in segment 3 (**a**; *arrow*) grew, while the tumors in 4 (**b**; *arrow*), 5 (**c**; *arrow*), 6 (**c**; *arrowhead*), and 7/8 (**d**; *arrow*) became slightly smaller

malignant vascular tumors of the liver including HEHE, and hemangiopericytoma is still difficult.

Most HAS are multiple and metastatic at presentation, and majority undergo hepatic resection. Prognosis of patients with HAS is poorer than that of those with other malignant vascular tumors of the liver. Median overall survival of patients with HAS after hepatic resection is 6 months and 1-year overall survival rate is 28.5 %, which is better than those who did not undergo hepatic resection (median overall survival 1 month) [4]. Few cases of survival beyond 2 years after hepatic resection for HAS have been reported, and are listed in Table [5–10]. Of these, 10 of 12 patients had a solitary tumor, and 11 patients underwent anatomical hepatic resection (Table 1). Four patients received preoperative needle biopsy, but only one patient was diagnosed as HAS. The treatment strategy for HAS has not been standardized [11], and complete resection of the solitary tumor is recognized as the only effective

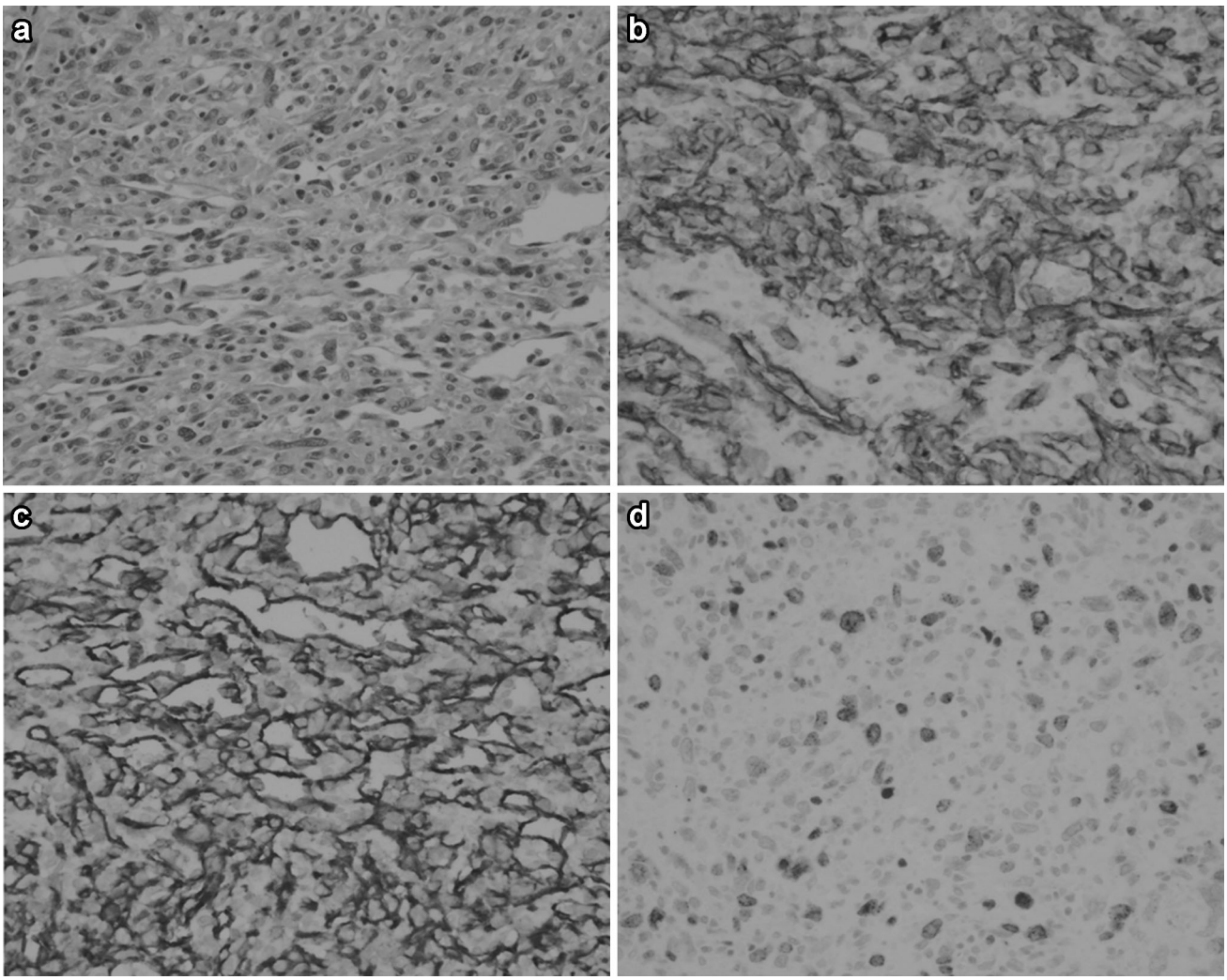


**Fig. 3** The tumor is composed of numerous endothelial cells without epithelioid cells (**a**), and is positive for vascular markers including CD31 (**b**), CD34 (**c**), and factor VIII-related antigen (**d**)

therapy. To the best of our knowledge, this is the second reported case of survival beyond 2 years after hepatic resection for multinodular hepatic angiosarcoma. The first reported long survivor with multinodular hepatic angiosarcoma after hepatic resection was a 65-year-old man. The patient had 2 nodules whose diameters were 13 and 3 cm, and underwent left hepatectomy. However, the patient had intrahepatic recurrence 10 months after the primary hepatic resection, and a second hepatic resection was performed. Unfortunately, the patient died 40 months

**Table 1** Cases of hepatic angiosarcoma with survival beyond 2 years after resection

| No | Age (years) | Gender | Preoperative diagnosis | Preoperative biopsy | Solitary/multiple | Size (cm) |
|---|---|---|---|---|---|---|
| 1. [5] | 39 | Male | NA | – | Solitary | 13 |
| 2. [6] | 47 | Female | NA | – | Solitary | 15 |
| 3. [7] | 54 | Female | HAS | Done | Solitary | 5 |
| 4. [8] | 46 | Male | HCC | Done | Solitary | 8 |
| 5. [8] | 45 | Male | HCC | – | Solitary | 7 |
| 6. [8] | 65 | Male | Cystadenocarcinoma | – | Multiple | 13 and 3 |
| 7. [8] | 52 | Female | HCC or adenoma | – | Solitary | 3 |
| 8. [9] | 67 | Male | Sarcoma | Done | Solitary | 15 |
| 9. [9] | 70 | Female | NA | – | Solitary | 11 |
| 10. [10] | 46 | Male | NA | – | Solitary | 14 |
| 11. [10] | 68 | Male | Sarcoma | – | Solitary | 16 |
| 12.* | 78 | Male | HEHE | Done | Multiple | 4, 3, 2, 2, and 1 |

| No | Surgery | Recurrence | Outcome |
|---|---|---|---|
| 1. [5] | Right hepatectomy | None | 38 months (alive) |
| 2. [6] | Right hepatectomy | None | 120 months (alive) |
| 3. [7] | Right hepatectomy | None | 64 months (alive) |
| 4. [8] | Non-anatomical resection | NA | 41 months (dead) |
| 5. [8] | Subsegmentectomy (S7, S8) | None | 84 months (alive) |
| 6. [8] | Left hepatectomy | Liver | 40 months (dead) |
| 7. [8] | Lateral segmentectomy | NA | 69 months (dead) |
| 8. [9] | Right hepatectomy | Peritoneum, lung | 32 months (alive) |
| 9. [9] | Subsegmentectomy (S5, S6) | Spine, brain, lung | 37 months (alive) |
| 10. [10] | Right hepatectomy | Peritoneum | 44 months (dead) |
| 11. [10] | Right hepatectomy | Peritoneum, lung | 31 months (alive) |
| 12* | Extended posterior segmentectomy, subsegmentectomy (S3) Non-anatomical resection | Liver | 28 months (alive) |

*NA* not available, *HAS* hepatic angiosarcoma, *HCC* hepatocellular carcinoma, *HEHE* hepatic epithelioid hemangioendothelioma, *S* segment

* Present case

after the primary hepatic resection due to tumor progression. In HAS patients, early discovery and close follow-up after surgical resection are necessary because of its poor therapeutic outcome.

**Compliance with ethical standards**

**Conflict of interest** The authors declare that they have no conflict of interest.

**Human/animal rights** All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008(5).

**Informed consent** Informed consent was obtained from the patient.

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