Scientific Research Publishing

Case Reports in Clinical Medicine, 2024, 13, 477-484
https://www.scirp.org/journal/crcm
ISSN Online: 2325-7083
ISSN Print: 2325-7075

Rapidly Progressing Hepatic Angiosarcoma Terminally Cared for at Home: A Case Discussion

—Hepatic Angiosarcoma Taken Palliative Care at Home

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How to cite this paper: Yamane, H., Yoshimitsu, A., Itagaki, T. and Yamane, M. (2024) Rapidly Progressing Hepatic Angiosarcoma Terminally Cared for at Home: A Case Discussion—Hepatic Angiosarcoma Taken Palliative Care at Home. *Case Reports in Clinical Medicine*, **13**, 477-484.
https://doi.org/10.4236/crcm.2024.1311056

Received: September 30, 2024
Accepted: November 2, 2024
Published: November 5, 2024

Abstract

Background: Hepatic angiosarcoma is a rare disease with a poor prognosis due to its tendency for distant sites. Few opportunities exist for palliative treatment of hepatic angiosarcomas at home. Here, we report a rare case of palliative treatment of hepatic angiosarcoma at home. **Case Presentation:** An 87-year-old male patient complained of upper abdominal pain and anorexia, persisting for 2 months. Computed tomography revealed multiple tumors in the liver, spleen, left kidney, and bone. Hepatic angiosarcoma was diagnosed based on the liver biopsy results. Due to his advanced age and dementia, the patient and his family decided to receive palliative treatment at home, thereby initiating home medical care at our clinic. During the first visit, oral opioid medication was introduced, and home oxygen therapy (HOT) was initiated because of complaints of cancer pain and respiratory distress. As oral intake became difficult, the patient was switched to a patch opioid, and suppositories were used for ton use. The patient was treated with morphine and HOT; however, the improvement in respiratory distress was below acceptable. The patient died at home on the ninth day after his visit. **Conclusion:** To the best of our knowledge, this is the first case report on palliative care for hepatic angiosarcoma at home. Owing to the rapid progression of this disease, home physicians must know its characteristics and provide appropriate medical care.

Keywords

Hepatic Angiosarcoma, Home-Based Palliative Care Service, Home Oxygen Therapy, Invasive Tumor

1. Introduction

Angiomas represent approximately 3% of adult soft tissue sarcomas, with primary hepatic angiosarcoma being a rare and invasive subtype, accounting for 2% of all angiomas [1]. Hepatic angiosarcoma is a rapidly fatal tumor, with most patients dying succumbing to liver failure or hemorrhage within 6 months [2]. At diagnosis, 28% of patients present with metastatic disease [3]. The rarity of this tumor, combined with the absence of specific tumor markers, makes diagnosis particularly challenging.

Home medical care provides numerous advantages to homebound patients, particularly older individuals. Home medical care has been reported to foster trust between physicians and patients, allowing physicians to understand the patient's home environment and other aspects of their lives, which can aid in making decisions about medical interventions and examinations [4]. However, home physicians' prognostic predictions for patients with terminal cancer often differ from the actual prognosis [5]. In this study, we report our experience of caring for rapidly progressing hepatic angiosarcoma at home.

2. Case Description

We present the case of an 87-year-old male with a medical history of hypertension, dementia, and benign prostatic hyperplasia of an unknown period. He presented with a complaint of upper abdominal pain and anorexia that persisted for 2 months. His blood test results during admission are summarized in **Table 1**. Computed tomography (CT) revealed multiple liver tumors (**Figure 1(A)**), with some tumors characterized by ring-like contrast effects. In addition, there were findings suggestive of metastasis to the spleen, left kidney, and bilateral lungs (**Figure 1(B)**, **Figure 1(C)**). Numerous osteolytic changes were observed in the spinal and pelvic bones (**Figure 1(D)**). Positron emission tomography-CT (PET-CT) revealed a diffuse liver tumor with a maximum standardized uptake value (SUV) accumulation of 7.9. Moreover, metastasis to the bones, spleen, and left kidney tumors showed an SUVmax of approximately 7.4. No significant accumulation occurred in the lung nodules. A percutaneous liver tumor biopsy was performed on segment 5 of the liver. A dense proliferation of spindle-shaped tumor cells accompanied by partial necrosis was observed (**Figure 2(A)**). Immunohistochemical test results were positive for CD31 and CD34, indicating hepatic angiosarcoma (**Figure 2(B)**). The patient and his family opted for palliative care at home because of his advanced age and dementia, declining systemic treatment. The patient was referred to our clinic by a home physician, who initiated home-based palliative care services.

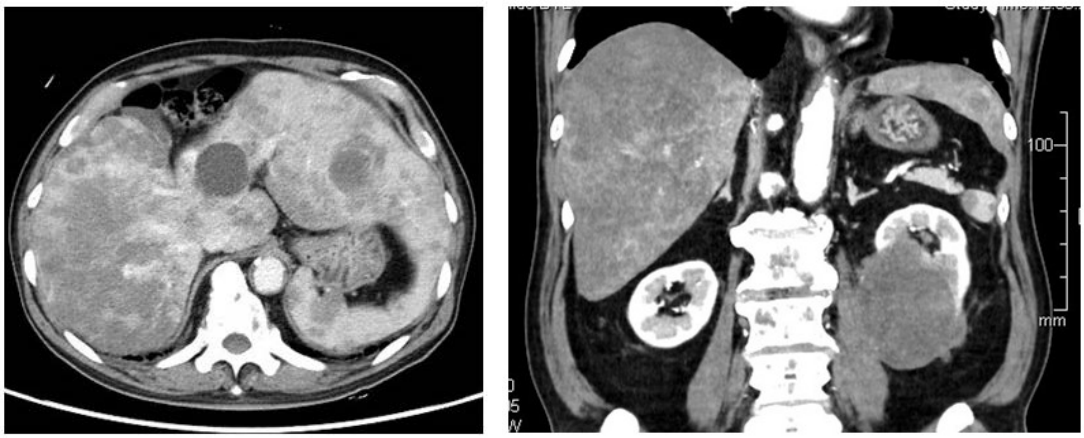
On the first day of the visit, the patient complained of upper abdominal pain and had a Numerical Rating Scale (NRS) [6] of approximately 5 and a Glasgow Coma Scale score of 13 (E4V4M5). The patient was able to drink water but had difficulty eating because of muscle weakness due to disuse syndrome. Oxycodone Hydrochloride Hydrate was administered for pain management, improving the NRS to approximately 3 with once-daily oral administration of Oxycodone Hydrochloride

Hydrate; however, the patient gradually developed difficulty taking oral medication. In addition, worsening of the symptoms of respiratory distress and a decrease in oxygen saturation were observed (90% at room air).

Table 1. Laboratory data of the patient.

White blood cell	6840	/uL
Haemoglobin	8.7	g/dL
Plate	10.4	10^4/uL
Total protein	6.4	g/dL
Albumin	3.1	g/dL
Total bilirubin	1.1	mg/dL
Direct bilirubin	0.3	mg/dL
AST	61	U/L
ALT	57	U/L
LDH	395	U/L
ALP	242	U/L
BUN	25	mg/dL
Creatinine	1.94	mg/dL
CRP	7.85	ng/dL
CEA	3.2	ng/mL
CA19-9	10.7	U/mL
AFP	1.9	ng/mL
HBs ag	Negative	
HBs ab	Negative	
HCV ab	Negative	

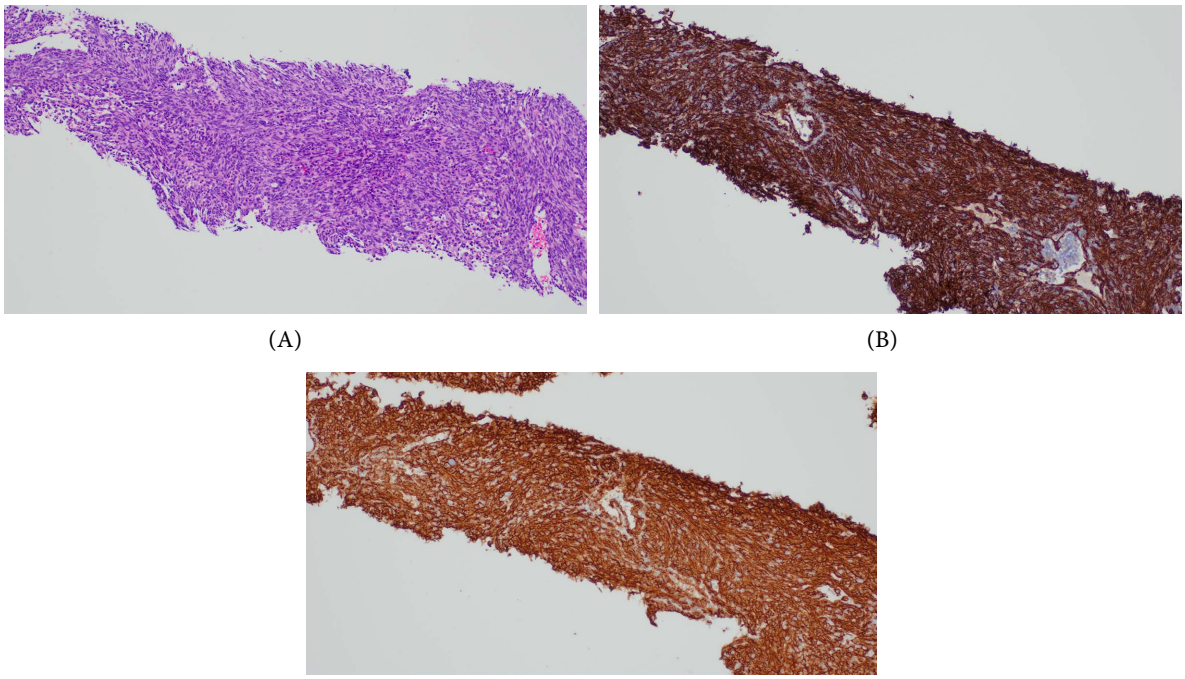
Abbreviations: AFP: alpha-fetoprotein; AST: aspartate aminotransferase; ALT: alanine aminotransferase; ALP: alkaline phosphatase; BUN: blood urea nitrogen; CA19-9: carbohydrate antigen 19-9; CEA: carcinoembryonic antigen; CRP: C-reactive protein; HBs ab: hepatitis virus B surface antibody; HBs ag: hepatitis virus B surface antigen; HVC ab: hepatitis virus C antibody.



(A) (B)

(C) (D)

Figure 1. Computed tomography examination image of the diagnosis. (A) numerous tumors with contrast effects were observed in the liver. (B) an 8-cm cystic mass was observed in the left kidney. The margins of the cystic mass were marked with an irregular contrast effect. (C) multiple nodules were observed in both lungs. (D) multiple bone metastases were observed in the spine and pelvis.



(A) (B)

(C)

Figure 2. Liver biopsy of diagnosis. (A) hematoxylin and eosin-stained liver biopsy demonstrated dense proliferation of spindle-shaped tumor cells. (B) immunohistochemical staining demonstrated positive for CD31. (C) immunohistochemical staining demonstrated positive for CD34.

On the fourth day of the visit, 1 mg fentanyl citrate per day and 10 mg morphine hydrochloride hydrate suppositories were started for pain and respiratory distress. Home oxygen therapy (HOT) was simultaneously initiated. Respiratory distress

improved (oxygen saturation: 95% at 1.0 L/m, nasal cannula), the NSR decreased from 6 to approximately 3. Thereafter, his level of consciousness gradually declined, making it impossible for him to eat or drink (GCS 7, E2V2M3). The patient was treated with fentanyl citrate (1 mg/day) without any obvious pain complaints. However, the respiratory distress remained mild and did not improve with the use of HOT and morphine hydrochloride hydrate suppositories. On the ninth day of the visit, the patient died at home (36 days after visiting the previous hospital).

3. Discussion

In this case, we administered palliative treatment for a rare hepatic angiosarcoma at home. Primary hepatic angiomas account for 2% of all primary hepatic malignancies [7]. Hepatic angiosarcoma progresses rapidly, and many patients develop distant metastases. This could be attributed to the fact that hepatic angiosarcomas do not merely enlarge in a full-blown manner, as in hepatocellular carcinoma, but also develop in a spongy fashion or infiltrate through blood vessels without destroying the hepatic structure [7]. Immunohistochemistry plays an important role in diagnosis, with CD31 expressed in 79.2% and CD34 in 87.5% of hepatic angiosarcomas [8]. In this case, CD31 and CD34 were also positive, and the histological findings confirmed the diagnosis of hepatic angiosarcoma.

Previous studies have demonstrated that distant metastasis is detected in 28.3% of cases at the time of diagnosis [3], and the prognosis is expected to be markedly short, with a median of 6 months [2]. Surgical resection is the recommended treatment; however, resectable cases are limited because multiple lesions are often found in the liver [9]. Chemotherapy and transcatheter arterial chemo-embolization have been reported as other treatment options; no established standard of palliative treatment exists [10] [11]. This case also showed multiple tumors in the liver and distant metastases. Due to the patient's age and dementia, the patient and family preferred palliative treatment at home. Prognostic factors associated with hepatic angiosarcoma include unresected and distant metastases [2]; however, detailed prognostic prediction in patients with hepatic angiosarcoma remains difficult owing to a lack of adequate literature. Although the Prognostic index (PPI) is a commonly used prognostic indicator for cancer patients [12], its application in rapidly progressing cases, such as the present case, is challenging. Rapid progression of illness can lead to premature end-of-life care and inadequate decision-making support for patients and their families [13]. Therefore, home care physicians should be aware of the characteristics of such rare diseases. When managing a rapidly progressing disease like this, it is essential to communicate effectively with the family and medical providers by holding sufficient meetings in advance.

Palliative treatment performed at home faced challenges in controlling respiratory distress in this case, despite the use of opioid preparations and HOT. Multiple possible causes of respiratory distress exist, including airway obstruction, bronchospasms, anxiety, and anemia [14]. As the patient had hepatic and renal

dysfunction, the opioid dose was carefully increased while checking for side effects. This may explain why opioids alone do not produce sufficient therapeutic effects.

HOT is useful for symptomatic relief in terminally ill cancer patients who present with hypoxemia, and home healthcare settings are commonly used. A previous study suggested that patients perceived more advantages than disadvantages of HOT, including increased functional capacity and symptom management, despite limitations such as decreased mobility and equipment-related discomfort [15]. HOT was also performed in this case; however, the improvement in respiratory distress was limited. The usefulness of NPPV in patients with terminal cancer has recently been reported. Nava *et al.* compared NPPV and oxygen therapy in patients with end-stage cancer presenting with acute respiratory failure; the NPPV group showed predominant improvement in respiratory distress [16]. However, in Japan, the indications for NPPV are limited to conditions such as acute respiratory failure and hypercarbonemia. Benzodiazepines were considered if the respiratory distress did not improve with the use of opioids and HOT. In this case, benzodiazepines were not administered because of the rapid progression of impaired consciousness. In palliative care at home, patients often struggle to control respiratory distress, and the home physician must modify the treatment accurately.

There are limitations to this case report. It is a retrospective study of a single case. Because of the rarity of the disease, the challenges in managing cases treated at home are not well known. It is important to accumulate more reports on cases of rare diseases in the future.

We report a case of rapidly progressing hepatic angiosarcoma treated at home. Hepatic angiosarcoma is rarely encountered at home, and physicians should be well-informed about its characteristics and poor prognosis. Notably, symptom control is made more difficult by the rapid progression of hepatic angiosarcoma.

Authors' Contributions Statement

Hiroaki Yamane: conceptualization (lead); writing—original draft (lead); formal analysis (lead); writing—review and editing (equal).

Aki Yoshimitsu: review and editing (equal).

Tomoko Itagaki: review and editing (equal).

Motoi Yamane: review and editing (equal).

All the authors have read and approved the final manuscript.

Statement of Ethics

Ethical approval was not required in accordance with the local guidelines for this retrospective, unplanned study. Written informed consent was obtained from all participants.

Data Availability Statement

All data generated or analyzed in this study are included in the published article.

Further inquiries can be directed to the corresponding authors.

Conflicts of Interest

The authors declare no conflicts of interest.

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