Sarcomatoid Hepatocellular Carcinoma With Skull Base Metastasis: A Case Report

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Case report

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Abstract

Background

Sarcomatoid hepatocellular carcinoma (SHC) is a relatively malignant tumor due to its recurrence and metastasis. Although metastatic SHC has been reported, skull base metastasis has not yet been documented.

Case presentation:

We herein report on a 51-year-old male with left throbbing headache and cranial nerve neuropathies. Magnetic resonance imaging revealed an extradural lesion of over 4 cm involving the jugular foramen, hypoglossal canal, clivus and the atlas. An endoscopic endonasal approach (EEA) was adopted for tissue biopsy and decompression, and the symptoms of the patient improved significantly after the surgery.

Conclusions

SHC invading the skull base is an extremely rare circumstance. For such extradural lesions, EEA is the most applicable and promising treatment approach.

Background

Hepatocellular carcinoma (HCC), the 5th most common type of tumor worldwide, [1] includes various type of cytological changes, of which spindle-cell like sarcomatoid hepatocellular carcinoma (SHC) is a relatively rare subtype found in only 1.8-2.0% of surgical cases and 3.9–9.4% of autopsy cases [2]. The prognosis of SHC is extremely poor due to its high risk of recurrence and metastasis; also, no efficient therapy exists for it. A few cases of SHC with pelvic bone and peritoneum metastasis have been reported; however, metastasis to the skull base has never been reported [2]. We report herein a patient harboring bilobar SHC treated with radiofrequency ablation (RFA) and transarterial chemoembolization (TACE) presenting with skull base metastasis. The hospital Institutional Review Board approved the study and the patient provided written informed consent to participate.

Case Presentation

This 51-year-old male was a Hepatitis B virus carrier complicated with Child-Pugh score A parameter liver cirrhosis whose diagnosis of bilobar SHC was confirmed through liver biopsy on January 18, 2019. The serum α-fetoprotein (AFP) level of the patient was within normal range (11.6 ng/ml) at the time of diagnosis. An F-18 fluorodeoxyglucose (FDG) whole body tumor scan performed on January 24, 2019 disclosed no focal areas of abnormal uptake of radioactivity except for Segments 3, 4 and 8 of the liver. The first episode of TACE therapy was arranged for February 10 to 19, 2019, and the patient received
portal vein embolization and lobectomy of the left lobe. RFA of the other lobe was performed on March 18, 2019.

However, in April the patient began to complain of a left throbbing headache (pain scale: 9 of 10) for several days, accompanied by hoarseness, dysphagia and left-sided deviated tongue. The patient returned to our general surgical outpatient department on April 22 and 26 for evaluation and treatment, respectively. A brain computed tomography (CT) showed no intracranial hemorrhage but did reveal an osteolytic lesion located at the skull base. Liver CT showed no abnormal enhanced lesions in the residual liver.

The patient was admitted for a Tc-99m Methylene Diphosphonate (MDP) whole body bone scan on April 30, at which time the serum AFP level was found to have increased dramatically to 1002.2 ng/mL. The Tc-99m MDP whole body bone scan showed no increased uptake of radioactivity at the skull base; nevertheless, magnetic resonance image (MRI) revealed an extensive extradural lesion of over 4 cm with jugular foramen, hypoglossal canal, clivus and C1 bony invasion (Fig. 1). The vertebral artery was encased within the lesion without significant stenosis. A second F-18 FDG whole body tumor scan was arranged on May 13, and focal areas of increased radioactivity uptake in the skull base, central and left side of C1 were compared to the previous whole body scan from January 24. A metastasis of SHC was highly suspicious due to correlation with an elevated AFP level and the image findings mentioned above.

An endoscopic endonasal approach (EEA) for decompression and tissue proof was adopted on May 1. The specimen was confirmed as a poorly differentiated carcinoma with markedly pleomorphic and hyperchromatic nuclei with frequent mitotic figures (Fig. 2). According to the histopathological result, a diagnosis of SHC with skull base metastasis was made. The patient experienced improvement in throbbing headache (pain scale: 2) and cranial nerve neuropathy after the surgery. The patient refused to receive chemotherapy or radiotherapy for the residual tumor.

Unfortunately, the patient suffered from general weakness beginning on July 15, which was suspected due to progressive pneumonia and liver cirrhosis. A CT done on July 19 disclosed no evidence of progression of the residual tumor at the skull base. The family agreed to a do-not-resuscitate order. On August 5, the patient experienced a general seizure episode, and his family members requested discharge against medical advice. The patient died on September 15, 2019.

Discussion And Conclusions

Bone metastasis often occurs in patients with breast, lung or prostate cancer, but it can also be found in most cases of advanced malignancies. Additionally, the skull and the skull base are frequent sites for bone metastasis. Bone metastasis typically involves the formation of destructive and osteolytic lesions centered in the bone on CT. It also shows hypointensly to normal bone marrow on non-contrast T1-weighted MRI [3]. The metastatic lesion at the skull base of our patient manifested as an extended infiltrative soft tissue mass with bony destruction on T1-weighted MRI, but no uptake signal at the skull base location was shown on the Tc-99m MDP whole body bone scan. The difference between the two
episodes of F-18 FDG whole body tumor scan might imply that the SHC skull base metastasis did not exist before the diagnosis of primary SHC was made. In other words, these traits indicated that the lesion was most likely a metastatic tumor rather a second primary tumor, and gradually expanded into the soft tissue mass without any bone-associated activity [2].

The serum AFP level of the patient was initially within the normal limit (11.4 ng/ml) when he was diagnosed as having SHC before TACE, left liver lobectomy and RFA of the right liver lobe. This finding is compatible with the results of an 11-patient SHC case series by Koo et al [4]. In this case series, more than 50% of the patients had low or negative serum AFP levels. SHC with distant metastasis to the skull base occurred in our patient, and serum AFP was then notably elevated (1002.2 ng/mL).

According to the literature review by Trivedi et al., [5] only 24 cases of normal-cell type HCC with skull base metastasis were reported from 1968 to 2009. An additional two SHC cases were also reported: one with pelvic skeletal metastasis and one with peritoneal dissemination [2, 6]. Based on the above, there has never been any cases reported previously as sarcomatoid cell type HCC with skull base metastasis.

By definition, a sarcomatoid cell type of carcinoma must include both epithelial and mesenchymal differentiation at the same lesion site [1]. The typical findings of SHC have been recorded to be massive expanding or multinodular confluent type with partial encapsulation; [4] more specifically, the histopathology of primary hepatic sarcoma cells often presents with spindle shaped, clear nucleolus, acidophilic cytoplasm and significant mitotic attributes [7]. To establish a diagnosis of metastatic SHC to the skull base, pathological confirmation through EEA biopsy is required. In our case, the typical appearance of the SHC subtype was also observed in the skull base metastatic biopsy. Epidural biopsy showed sheets of tumor cells with markedly pleomorphic and hyperchromatic nuclei, frequent mitotic figures and tumor necrosis.

The prognosis of SHC is extremely poor, with 3-year survival rates reported as low as 18.2% after hepatectomy, [8] and our patient had experienced a metastatic recurrence at the skull base only 34 days after a left hepatic lobectomy. Radiotherapy, surgery and palliative care have been the options for HCC with skull metastasis in the past; [9, 10] however, EEA for decompression is now among the most optimal treatments for skull base metastasis because of its minimal invasiveness. Although EEA did not considerably prolong the life expectancy of our patient, it was able to improve neurological function and help the patient maintain his quality of life [11].

SHC with skull base invasion is such a rare condition that it has not been previously reported. Definite diagnosis of it requires neurologic findings, radiographic images and biopsy proof. Tc-99m MDP whole body bone scan and serum AFP levels were not considered definitive for monitoring bone metastasis of SHC in a previous report, these tools provided insufficient findings for diagnosis in our case. EEA for tissue biopsy and decompression is regarded as the most minimally invasive and optimal diagnostic and treatment option for skull base metastasis.
List Of Abbreviations

SHC  
Sarcomatoid hepatocellular carcinoma  
EEA  
Endoscopic endonasal approach  
HCC  
Hepatocellular Carcinoma  
TACE  
Transarterial chemoembolization  
RFA  
Radiofrequency abation  
F-18 FDG  
F-18 fluorodeoxyglucose  
AFP  
Alpha fetal protein  
Tc-99m MDP  
Tc-99m methylene diphosphonate  
CT  
Computed tomography  
MRI  
Magnetic resonance image

Declarations

Ethics approval and consent to participate

Currently under process.

Consent for publication

Consent will not be provided on submission.

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

Funding
Authors' contributions

Shih-Ming Jung and Bo-An Chen both analyzed and interpreted the patient’s pathological results regarding the metastatic tumor lesion. Cheng-Chi Lee was responsible for supervising and revision of the manuscript, besides, he is also the corresponding author. Chen Lin was a major contributor in writing the manuscript. All authors read and approved the final manuscript.

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Figures

Figure 1
T1-weighted magnetic resonance images (MRI) of the patient after admission for brain metastasis survey. Soft-tissue signal-intense lesion was noticeable at the central skull base (upper arrow). The outer margin of the left vertebral artery was encased by a soft-tissue signal-intense lesion (lower arrow).

Figure 2

Hematoxylin-eosin stain of the skull base biopsy. A. Lesion tissue at the skull base (white arrow). Magnification: 100×. B. Bunchy, spindle-shaped cells revealed markedly pleomorphic hyperchromatic nuclei and significant mitotic figures (white arrow). Magnification: 400×.