Case Report
Primary solitary extramedullary plasmacytoma of the liver mimicking hepatocellular carcinoma: case report

Yi Shao¹, Han Zhang², Qiyi Zhang¹, Sheng Yan², Weilin Wang¹,³,⁴

¹Division of Hepatobiliary and Pancreatic Surgery, ²Department of Pathology, ³Key Laboratory of Precision Diagnosis and Treatment for Hepatobiliary and Pancreatic Tumor of Zhejiang Province, ⁴State Key Laboratory and Collaborative Innovation Center for Diagnosis and Treatment of Infectious Diseases, First Affiliated Hospital, School of Medicine, Zhejiang University, Hangzhou 310003, China

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Abstract: Hepatic primary solitary extramedullary plasmacytoma (EMP) is a rare disease with few cases reported in the literature. In this case study, the patient was a 73-years-old man who exhibited a solitary hepatic mass on ultrasonography in a routine body examination when he presented to the 1st Affiliated Hospital, School of Medicine, Zhejiang University (Hangzhou, China) without any discomfort. The mass was then confirmed by magnetic resonance imaging (MRI) and computed tomography (CT) which initially suggested hepatocellular carcinoma (HCC). The patient was treated with a liver resection. Histologically, the tumor consisted of a diffused infiltration of plasma cells with slight atypia. Immunohistochemistry revealed the expression of CD138 and a Kappa light chain. Postoperatively, further examinations including bone marrow biopsy, skeletal radiography, serum and urine protein immunoelectrophoresis disclosed no abnormalities. There was no proof of anemia, hypercalcemia, or renal insufficiency. He was ultimately diagnosed with primary solitary EMP of the liver. After 13 months of close follow-up, the patient remained healthy. The imaging manifestations (ultrasound, CT and MRI) of EMP were similar to HCC, and the patient underwent the operation without chemotherapy or radiotherapy. Thus, we concluded that surgery alone gave an excellent result in this case whose resectability was good.

Keywords: Surgical resection, hepatectomy, extramedullary plasmacytoma, liver, plasma cell tumor

Background
Extramedullary plasmacytomas (EMPs) make up approximately 2.8% to 4% of all plasma cell tumors [1, 2] and occur mainly in the upper aerodigestive tract [2, 3] between the fourth and seventh decades of life at a median age of 55 to 63 years [1-3]. Besides in the upper aerodigestive tract, including the paranasal sinuses, nose, tonsils and nasopharynx, EMPs also occur in thyroid, lymph nodes, skin, brain, lung, pancreas, spleen, breast, testes, and liver [2, 3]. To the best of our knowledge, primary solitary EMP of the liver is rare, and very few cases have been reported [4-11]; however, most of these cases were treated with radiotherapy, and only one case received a surgical resection according to the literature [11]. Here, we present a second case. Written informed consent was acquired from the patient.

Case presentation
A 73-year-old man, with no history of systemic diseases, presented to the 1st Affiliated Hospital, School of Medicine, Zhejiang University (Hangzhou, China) due to a hepatic mass without any discomfort detected by ultrasonography (Figure 1A). The physical examination was unremarkable. Liver function tests, renal functions, and hematological indexes were in the normal ranges. The patient’s α-fetoprotein was not increased and was negative for hepatitis B infection. A multiphasic computed tomography (CT) scan of the liver revealed a 2.3 x 1.6 cm-sized mass in segment VII, hypovascular [7, 9-11] on non-contrasting images with a CT value of 41HU, dishomogeneously hyperenhancing in the arterial phase, with washout from the portal to the delayed phase (Figure 1B, 1C). T1-weighted magnetic resonance imaging (MRI) of the liver demonstrated a well-defined mass
Extramedullary plasmacytoma of the liver.

The ultrasound image showed a 1.7 × 1.7 cm-sized well-defined hypoechoic mass in the right lobe of the liver. The arterial phase image of CT revealed a 2.3 × 1.6 cm-sized hyperenhanced mass in segment VII, with washout in the delayed phase. The mass was of low signal intensity on a T1-weighted image of MRI, and of high signal intensity on a T2-weighted and diffusion-weighted image.

Figure 1. Ultrasonography, multiphasic computed tomography (CT), and magnetic resonance imaging (MRI) of liver.

The initial impressions of the CT and MRI suggested a diagnosis of high-grade hepatocellular carcinoma (HCC). The resectability of this patient was good with a reasonable risk, so he received a hepatectomy as therapy. The postoperative pathology exhibited an EMP. A histological examination of the resected specimen exposed a diffused infiltration of plasma cells, invading and destroying the liver parenchyma without any eosinophils, fibroblasts, or lymphoid cells. Occasional binucleated and heterotypic forms were noted. No amyloid accumulation was detected.
Extramedullary plasmacytoma of the liver

Histochemical analysis, the neoplastic cells were positive for Kappa-chain (Figure 2C), CD138 (Figure 2D), CD38, CD43, MUM1, Bcl-2, and CyclinD1, but Lambda-chain, CK, CD3, CD5, CD10, CD20, CD21, CD23, CD79a, Bcl-6, SOX11, and PAX-5 were all negative. Additionally, further examinations including a bone marrow biopsy, a skeletal radiography, and serum and urine protein immunoelectrophoresis revealed no abnormalities. Consequently, a diagnosis of truly localized, primary solitary EMP of the liver was finally established. After 13 months of close follow-up, the patient remained healthy.

Discussion and conclusions

EMPs represent about 2.8% to 4% of all plasma cell tumors [1, 2], and most commonly affect the upper aerodigestive tract up to about 82.2% [3]. Other locations include the thyroid, lymph nodes, skin, brain, lung, spleen, pancreas, testes, breasts, and liver [2, 3]. In most series, EMPs have been reported to be distinct entities. The reason for the various incidences of EMPs between the presenting sites is still unclear.

The diagnosis of EMP was based on three aspects: (1) having a solitary soft-tissue mass which was pathologically proven to be a plasmacytoma, (2) plasma cell infiltration less than 5% in the bone marrow at diagnosis, (3) the absence of other tissue involvement or osteolytic bone lesions without evidence of myeloma, anemia, hypercalcemia, or renal insufficiency, and low serum or urine M-protein concentration, if it exists [1, 2]. Primary solitary EMP of the liver is so rare that only a few cases have been reported so far [4-11], so the diagnosis of hepatic EMP is very difficult. The case we describe here is a male patient who suffered from a solitary plasma cell neoplasia in the liver without any other proof of myeloma, including no M-protein in the urine or serum. However,
his CT and MRI manifestations demonstrated an enhancement pattern identical with that expected of HCC. Hence without pathology it can be confused with HCC, but the therapeutic strategy is different.

As EMPs are highly radiosensitive, the most common treatment is radiotherapy. About 4700-6500 cGy of radiation doses could achieve local control in almost all EMP patients [1], and 5,000 cGy of radiotherapy was used most for the treatment of hepatic EMP, resulting in an encouraging outcome [8, 9]. Other approaches include chemotherapy [6, 7] and surgery. The first case of EMP of the liver that underwent a successful radical resection was reported in 2016 [11]. To our knowledge, the case we presented here is the second one. Limited information is available about the surgical therapy of hepatic EMP. Synthesizing earlier studies reporting on EMP at other sites, we argue that surgery alone might yield the best results when resectability is good and negative surgical margins can be obtained [3].

The prognosis of EMP is excellent, with a 10-years overall survival rate of 70%-78% [2, 3]. After treatment, 17.5%-21% of the patients experienced a recurrence, and 14%-36% of the cases developed multiple myeloma [2, 3, 12, 13]. Moreover, adjuvant chemotherapy did not influence the incidence of conversion [1, 13]. Also, adjuvant radiotherapy was not suggested for patients who had undergone a radical resection and would only be required in the event of inadequate surgical margins [3].

The published papers on this disease are restricted to case reports. With few cases and limited information, many problems remain unclear. Based on existing information, we suggest that upon the detection of a liver mass, if it is a large-sized lesion, with a great complexity of the anatomical location, high surgical risks, or surgical unresectability, and the diagnosis of EMP is suspected, a biopsy should be performed. Under these conditions, radiotherapy is the optimal choice; otherwise if the resectability is good, surgery can be recommended first.

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Written informed consent was acquired from the patient.

Disclosure of conflict of interest

None.

Abbreviations

EMP, extramedullary plasmacytoma; MRI, magnetic resonance imaging; CT, computed tomography; HCC, hepatocellular carcinoma.

Address correspondence to: Dr. Weilin Wang, Division of Hepatobiliary and Pancreatic Surgery, First Affiliated Hospital, School of Medicine, Zhejiang University, No. 79 Qingchun Road, Hangzhou 310003, China. E-mail: wam@zju.edu.cn

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