

Primary Extraskkeletal Osteosarcoma of Liver: Case of Report

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Case report

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Abstract

Background

Extraskelatal osteosarcoma (ESOS) is a highly malignant osteosarcoma that occurs in extraskelatal tissues. It often affects the soft tissues of the limbs. ESOS is classified as primary or secondary ESOS.

Case presentation

we report a case of primary hepatic osteosarcoma in a 76-year-old male patient. The patient had a giant cystic-solid mass in the right liver that was evident on ultrasound and computed tomography. Postoperative pathology and immunohistochemistry of the mass, which was surgically removed, suggested fibroblastic osteosarcoma. No other abnormal lesions were found. Therefore, the patient was diagnosed with primary hepatic osteosarcoma. The hepatic osteosarcoma reoccurred 48 days after surgery, resulting in significant compression and narrowing of the hepatic segment of the inferior vena cava. Consequently, the patient underwent stent implantation in the inferior vena cava and transcatheter arterial chemoembolization. Unfortunately, the patient died of postoperative multiple organ failure.

Conclusions

Hepatic osteosarcoma is a rare mesenchymal tumor with a short course and a high likelihood of metastasis and recurrence. If a biopsy were to return osteoid in a large liver tumor, ESOS would be suspected. However, there is no evidence-based treatment plan to date. Surgical resection combined with adjuvant chemoradiotherapy seems to be the best treatment option.

Background

Extraskelatal osteosarcoma (ESOS) is a highly malignant osteosarcoma that occurs in extraosseous tissues. It is characterized by low incidence, invasive growth, high proneness to metastasis and recurrence, and a poor prognosis [16]. ESOS often involves the soft tissues of the limbs. There are few reports of ESOS occurring in organs, and those are mostly case reports [3, 4]. The pathogenesis of ESOS is still unclear. ESOS is classified as primary or secondary ESOS according to its origin. Primary ESOS occurs in the extraskelatal organs and soft tissues and does not attach to bone or periosteum. No primary ESOSs are of bone origin. In contrast, secondary ESOSs are most often metastases of osteosarcomas of bone origin to the extraskelatal organs and soft tissues or are secondary to certain primary diseases [13]. The imaging manifestations of hepatic osteosarcoma are not specific, and its diagnosis depends on pathology and immunochemistry.

Case Report

A 76-year-old male patient was admitted to the hospital on July 20, 2020, due to the aggravation of existing abdominal pain and discomfort for 1 day. Prior to admission, he had had the abdominal pain

and discomfort for more than half a month. In terms of laboratory tests, alpha-fetoprotein was 7.86 ng/ml, a hepatitis B virus surface antigen test was negative, the patient was negative for hepatitis C antibodies, and HBV-DNA was $<1.00E+02$ IU/ml. Abdominal color Doppler ultrasound suggested a giant mixed echogenic mass in the right liver, and color Doppler flow imaging showed a small number of blood flow signals in and around the mixed echogenic mass (Figure 1). Computed tomography (CT) showed liver enlargement and a giant cystic-solid mass in the right liver, and the enhanced scan showed mild to moderate enhancement of the solid component of the mass (Figure 2). On July 28, 2020, the patient underwent liver mass resection at our hospital. During the operation, a cystic-solid mass (120×70×70 mm) was observed in the section of the liver next to the liver capsule. The cystic fluid was already lost, and the grayish-red and grayish-yellow solid area of the tumor was soft with a cut-fish-like surface. A rapid intraoperative pathology examination suggested mesenchymal sarcoma. Immunohistochemistry showed the following results: CK (pan) (-), EMA (-), CD34 (-), S-100 (-), SMA (scattered -), STAT6 (-), Ki67 (+, 30%), SATB2 (partially weak +), p16 (-), CD163 (scattered +), CD68 (scattered +), CD56 (-), desmin (-), and H-Cald (-). Postoperative pathology and immunohistochemistry suggested fibroblastic osteosarcoma (Figures 3 and 4). The patient received capecitabine monotherapy and was discharged 34 days after surgery. On September 14, 2020, he was readmitted to the hospital due to abdominal distension and pain. Whole-abdomen nonenhanced and contrast-enhanced CT scan examinations showed that the residual liver parenchyma had a patchy lesion with mixed attenuation and apparently uneven enhancement near the inferior vena cava and that the hepatic segment of the inferior vena cava was significantly compressed and narrowed (Figures 5), suggesting tumor recurrence. Laboratory tests showed poor liver and coagulation function. Considering that the patient had inferior vena cava compression, stenosis, and a large amount of ascites, stent implantation in the inferior vena cava and transcatheter arterial chemoembolization were performed on September 22, 2020. Seven days after surgery, the patient died of multiple organ failure.

Discussion

ESOS is a highly malignant osteosarcoma that occurs in extraosseous tissues. This tumor was first reported in 1941 by Wilsom [1]. Its incidence is low, and it occurs primarily in elderly adults: The average age of patients with ESOS is 47.5 to 61 years old. ESOS accounts for 1% of all soft tissue sarcomas and 4% of osteogenic osteosarcomas [15]. ESOS is characterized by invasive growth, a high likelihood of metastasis and recurrence, and poor prognosis [16]. ESOS often involves the soft tissues of the limbs. There are few reports of ESOS occurring in organs, and most of these are case reports [3, 4]. The pathogenesis of ESOS is still unclear. There are two theories on the pathogenesis of ESOS [5]: (1) The tissue residual theory: Residual mesenchymal components from the embryonic development stage form bone and osteosarcomas. (2) The metaplasia theory: Interstitial fibroblasts in muscle tissues are converted into osteoblasts and chondroblasts in response to internal or external stimuli and then evolve into osteosarcoma. Currently, most scholars support the metaplasia theory. According to its origin, ESOSs are classified as primary or secondary ESOSs [13]. Primary ESOS occurs in extraskelatal organs and soft tissues and does not attach to the bone or periosteum. No primary ESOS is of bone origin. In contrast,

secondary ESOS is mostly the metastasis of an osteosarcoma of bone origin to the extraskelatal organs and soft tissues or is secondary to certain primary diseases, such as myositis ossificans. In the case reported in this study, except for the cystic-solid mass in the liver, no evidence of primary tumors or primary bone lesions was found. Therefore, the patient was diagnosed with primary hepatic osteosarcoma. Primary hepatic osteosarcoma is rare and has only been reported in individual cases [3, 4]. Although imaging examinations can help to identify lesions, the imaging findings of hepatic osteosarcoma are nonspecific and are not different from those of a variety of tumor-like lesions; consequently, it is difficult to accurately diagnose hepatic osteosarcoma preoperatively. In the case reported in this study, the hepatic osteosarcoma manifested as a cystic-solid mass. The histology of hepatic osteosarcoma is similar to that of skeletal osteosarcoma. Although the direct production of osteoid components by osteosarcoma cells has significant diagnostic value, it has no specificity [7]. Therefore, the diagnosis of hepatic osteosarcoma still relies on pathology and immunochemistry.

ESOS has a short course, rapid progression, a high local recurrence rate, and a high risk of distant metastasis [3]. Lee et al. [5] reported that the 5-year survival rate of a group of patients diagnosed with ESOS was only 37% and that most of them died within 2 to 3 years after the initial diagnosis. Studies [2, 9, 14] have shown that distant metastasis, large tumors (≥ 10 cm), tumors of the axial skeleton, and advanced age are poor prognostic factors for ESOS, while radiotherapy and chemotherapy have no significant correlation with mortality. The patient reported in this study was 76 years old. He had a large intrahepatic tumor of 17-18 cm. After surgical resection, he underwent chemotherapy. However, local recurrence occurred within a short time after surgery, and the disease progressed rapidly. The patient died within 3 months after the first occurrence of symptoms. At present, the treatment methods for hepatic osteosarcoma are similar to those used for other soft tissue sarcomas. Because this disease is rare, there is no evidence-based treatment plan to date. Surgical resection combined with adjuvant chemoradiotherapy seems to be the best treatment option [8-12].

Conclusion

Hepatic osteosarcoma is a rare mesenchymal tumor with a short course and a high likelihood of metastasis and recurrence. It is difficult to distinguish it from other tumors by imaging. Its diagnosis still relies on pathological and immunochemical examinations.

Declarations

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

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Contributions

Qiuyi Di and Qiongli Wen performed the manuscript writing and the literature collecting; Zhihong Cheng and Gang Zhong were involved in the operation; Xiangdang Long, Zhiqun Mao, and Gang Zhong conceived, designed, and supervised all studies and the drafting and editing of the manuscript. All the authors have read and approved the final manuscript.

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Ethics declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

Competing interests

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Figures

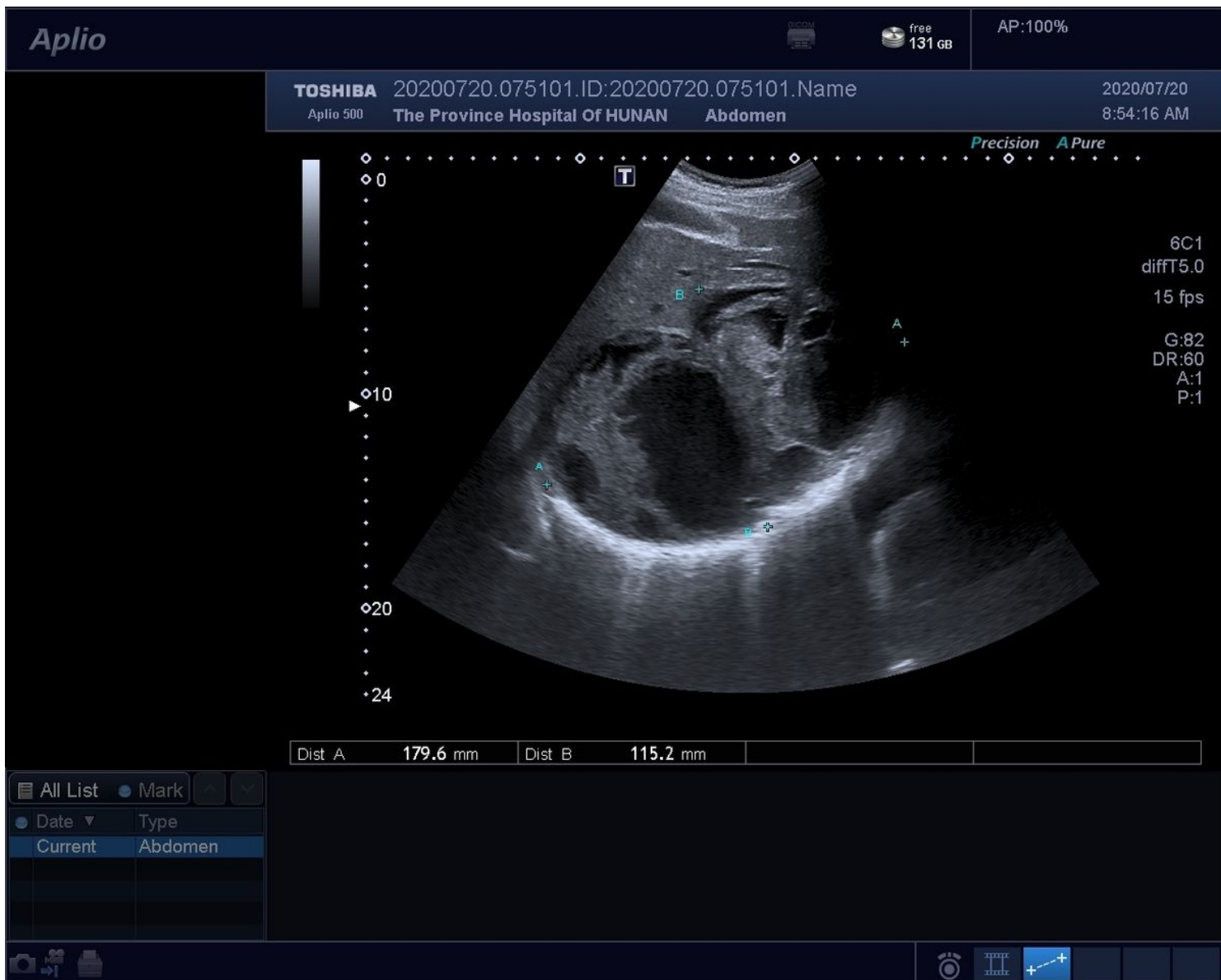


Figure 1

Ultrasound examination: A giant mixed echogenic mass (approximately 180×115×120 mm) was observed in the right lobe of the liver with a poorly defined boundary, irregular morphology, and uneven internal echoes. The mass was mainly cystic, with multiple solid parts inside. CDFI: A small number of blood flow signals were visible in and around the mixed echogenic mass, and the intrahepatic and right hepatic veins were compressed and offset.

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Figure 2

Computed tomography (CT) examination showed liver enlargement and a large cystic-solid mass (approximately 171 mm×127 mm×124 mm) in the right liver with irregular morphology, a poorly defined boundary, uneven attenuation, and CT values ranging from 11 to 62 HU. Contrast-enhanced computed tomography (CT): The solid component of the mass was mildly to moderately enhanced. The arterial branches were observed in the mass. The intrahepatic and right hepatic veins and the right anterior branch of the portal vein were occluded.

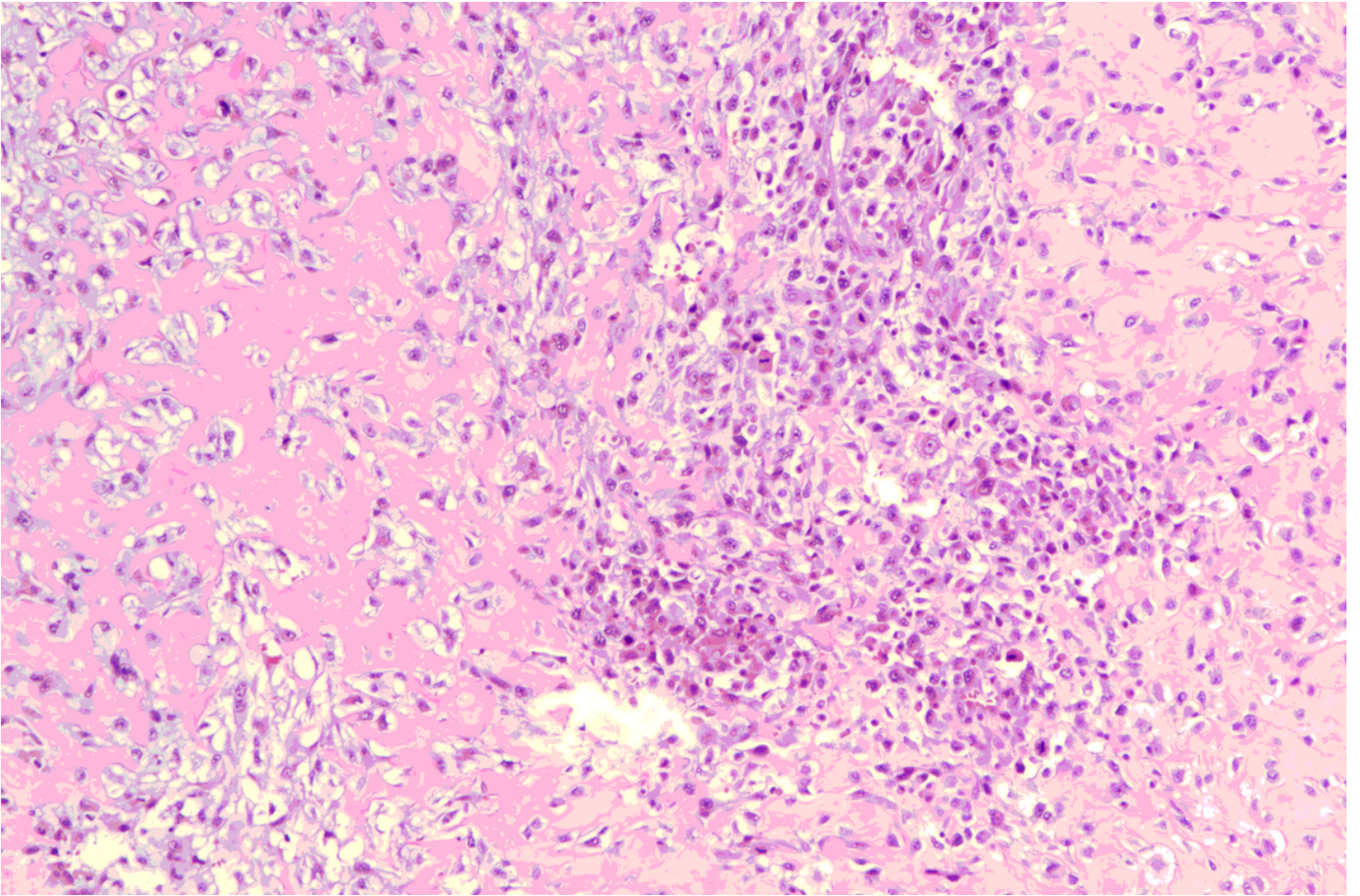


Figure 3

Pathological microscopy: The short spindle-shaped tumor cells had medium density, an increased nucleus-to-plasma ratio, atypical nuclei, and pathological mitosis. Some tumor cells were converted to osteoblasts. There was osteoid matrix between tumor cells and osteoclast-like giant cells.

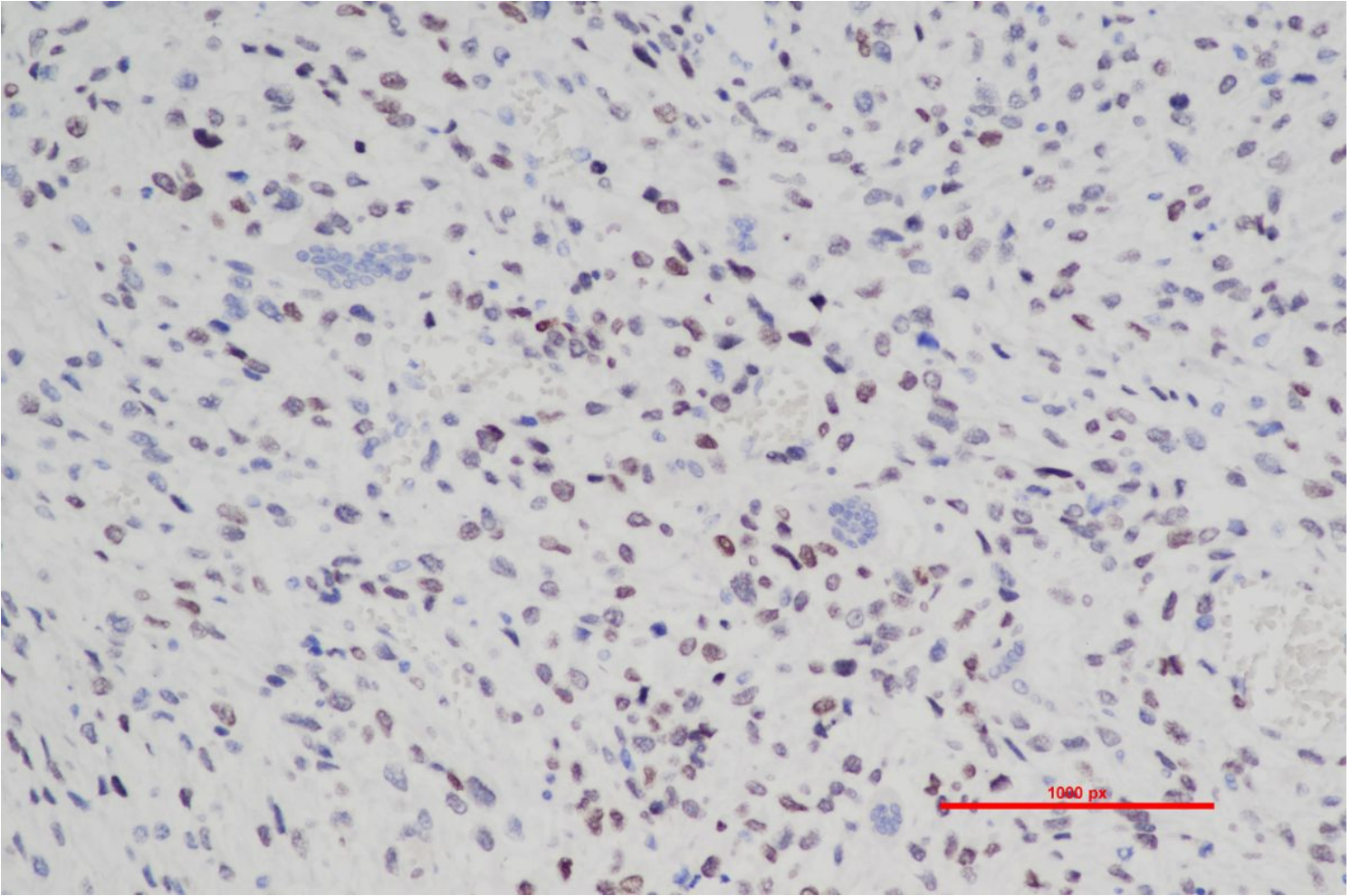


Figure 4

Immunohistochemistry: SATB2 was partially weakly positive.

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Figure 5

Noncontrast-enhanced CT scan: The residual liver parenchyma showed a patchy lesion (110×80×100 mm) with mixed attenuation and a clear boundary near the inferior vena cava. The lesion grew out of the liver contour and had multiply small, patchy hypoattenuating areas. Contrast-enhanced CT: The lesion showed obviously uneven enhancement, while the hypoattenuating areas of the lesion had no obvious enhancement. The hepatic segment of the inferior vena cava was significantly compressed and narrowed.