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Efficacy of Transhepatic Anterior Approach to the Inferior Vena Cava for Malignant Pheochromocytoma Invading Right Posterior Lobe of the Liver: A Case Report

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Introduction: The anterior approach to the inferior vena cava (IVC) by the liver hanging maneuver is effective in resecting large retrohepatic tumors without mobilizing the right lobe.

Case presentation: A 50-year-old man was referred to our hospital with a diagnosis of pheochromocytoma. He had severe congestive heart failure and cardiac ejection fraction was 15%. Abdominal magnetic resonance imaging (MRI) and ultrasonography (US) showed an adrenal mass about 80 mm in diameter. The tumor-infiltrated posterior segment of the right hepatic lobe and tumor were widely attached to the IVC. After treatment of congestive heart failure with conservative therapy, surgery was planned. Right adrenectomy and right hepatectomy were performed, the latter using the liver hanging maneuver to avoid mobilizing the right lobe, and we were able to minimize blood pressure fluctuations and perform the operation safely. The histopathologic diagnosis was malignant pheochromocytoma.

Conclusions: We performed right hepatectomy without mobilizing the right lobe by the liver hanging maneuver and minimized stimulation of the tumor. We could perform the operation safely using the liver hanging maneuver, which seems effective in such cases.

Key words: Malignant pheochromocytoma – Liver hanging maneuver

The liver hanging maneuver is a surgical technique that facilitates an anterior approach to inferior vena cava (IVC) during hepatectomy. This anterior approach to the IVC with the liver hanging maneuver is effective for resecting large retrohepatic tumors without mobilizing the right lobe. During

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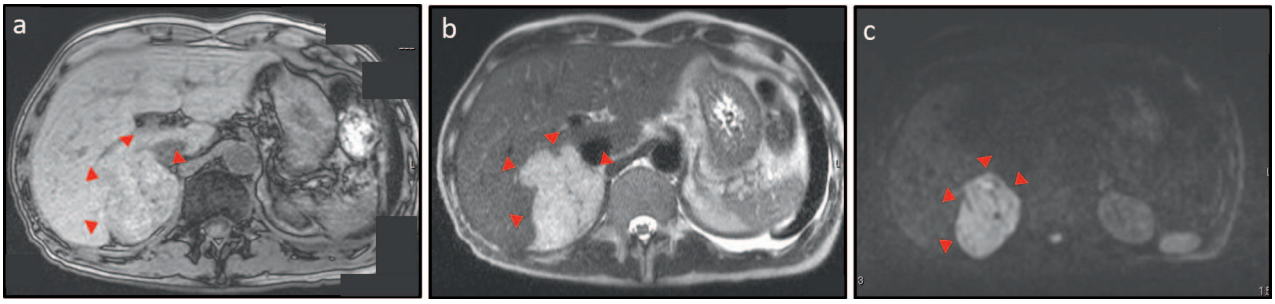

Fig. 1 Abdominal MRI shows a signal-hyperintense mass on T1- (a), T2- (b), and diffusion-weighted (c) images. Tumor has infiltrated the posterior segment of the right hepatic lobe and is close to the IVC (red arrows).

surgery for pheochromocytoma, stimulation of the tumor should be avoided to prevent additional secretion of catecholamines,[1] which can lead to uncontrollable hypertension and severe heart failure.

We report a case of malignant pheochromocytoma that infiltrated the posterior segment of the liver in a patient with severe congestive heart failure. For this case, a transhepatic anterior approach to the IVC using the liver hanging maneuver was effective to safely achieve right hepatectomy, including removal of the pheochromocytoma.

Case Presentation

A 50-year-old man presented to hospital with a chief complaint of exertional dyspnea. His past medical history included diabetes and hypertension. He was diagnosed with congestive heart failure and pneumonia. On admission, echocardiography demonstrated dyskinesis of the left ventricular anterior wall and septum; ejection fraction (EF) was 15% and the IVC was dilated to 21 mm. Severe congestive heart failure [New York Heart Association (NYHA) classification IV] was therefore diagnosed. Abdominal computed tomography (CT) revealed a right adrenal tumor about 60 mm in diameter. Pheochromocytoma was suspected, and the patient was referred to our department.

Blood pressure was 164/100 mmHg, and heart rate was 112 beats/min. Blood examination showed an N-terminal pro-B-type natriuretic peptide (NT-proBNP) concentration of 1101 pg/mL and a noradrenaline concentration of 12 ng/mL. Urine examination showed a vanillylmandelic acid (VMA) concentration of 32.9 mg/d and a normetanephrine concentration of 7.8 mg/d. Abdominal magnetic resonance imaging (MRI) revealed a signal-hyperintense mass measuring about 80 mm in diameter on T1-, T2- and diffusion-weighted imaging, and tumor size had increased from a month earlier. Moreover, the tumor seemed to have infiltrated the posterior segment of the right hepatic lobe and was widely attached to the IVC (Fig. 1).

Abdominal ultrasonography (AUS) showed a right adrenal tumor about 80 mm in diameter and clear infiltration of the liver. The tumor was widely attached to and compressing the IVC over a region of 4 cm, and surrounded one-third of the IVC (Fig. 2). Moreover, imaging visualized disruption of the IVC wall at the point of tumor compression, with tumor protruding into the tunica intima of the IVC.

Accumulation in the tumor was identified on ^{123}I-meta-iodobenzylguanidine (MIBG) scintigraphy, but no metastatic or heterotopic lesions were found (Fig. 3).

Using continuous hemodiafiltration to remove catecholamines, we controlled blood pressure with furosemide and α-blockers (doxazosin and fentolamine mesilate). Blood pressure was successfully controlled, but EF had slightly increased to only 24% after the initial 2 weeks of treatment. Treatment for a further 2 weeks achieved no improvement of EF. As excess secretion of catecholamines from the tumor was continuing, we considered that continuing conservative therapy would not improve cardiac function. Surgical resection was thus considered to represent the only potential method of rescue for the patient. Surgery was planned for 1 month after admission.

Preparation for operation

We considered that stimulation of the tumor would risk uncontrollable hypertension and fatal heart failure caused by excess secretion of catecholamines. Under such a situation, we should prepare total vascular clamping with a bypass from the IVC and portal vein to the superior vena cava (SVC), and also

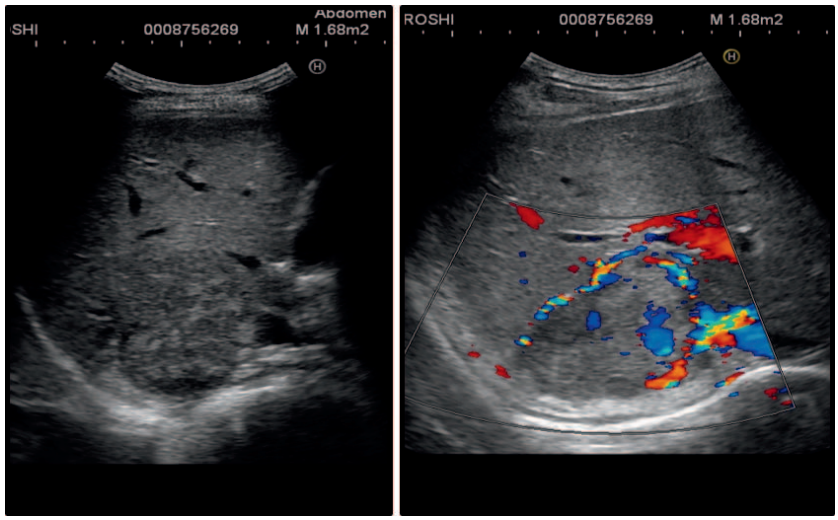

Fig. 2 Abdominal ultrasonography shows a right adrenal tumor about 80 mm in diameter infiltrating to the liver. Tumor appears close to the IVC.

prepare percutaneous cardiopulmonary support (PCPS). According to the degree of tumor infiltration into the liver, right adrenalectomy with posterior subsegmentectomy of the liver was considered the appropriate operative method, but tumor infiltration to the IVC was suspected, and we therefore planned right hepatectomy to allow broad expansion to the anterior side of the IVC.

Operation

We performed right adrenectomy and right hepatectomy as one lump. Operation time was 355 minutes, blood loss was 400 mL, and blood transfusion was 840 mL because of continuing anemia since admission. To start the operation, we began monitoring cardiac function with transesophageal echocardiography and inserted a catheter into the internal jugular vein and femoral vein to prepare for total vascular clamping. Intraoperatively, we used α- and β-blockers to avoid hypertension due to stimulation of tumor, and noradrenalin to prevent hypotension after ligation of the drainage vein of the tumor. Tumor had deeply infiltrated to the posterior segment of the liver. We first exposed the IVC and bilateral renal veins on the inferior side of the liver, then exposed the IVC on the cranial side of the liver to prepare for total vascular clamping if uncontrollable hypertension and unexpected hemorrhea resulted from catecholamine release (Fig. 4a and 4b). Right hepatectomy was achieved without mobilizing the right lobe using the liver hanging maneuver, and we were able to minimize blood pressure fluctuations (Fig. 4c and 4d). We carefully dissected the tumor from the IVC. No direct invasion to the IVC was apparent, but invasion to the root of the inferior right hepatic vein (IRHV) was found. We resected the root of the IRHV under side clamping of the IVC and achieved closure with running sutures (Fig. 4e). Intraoperatively, we controlled blood pressure to under 200 mmHg and cardiac function was monitored using transesophageal echocardiography, allowing the operation to be completed without clamping the IVC or using PCPS.

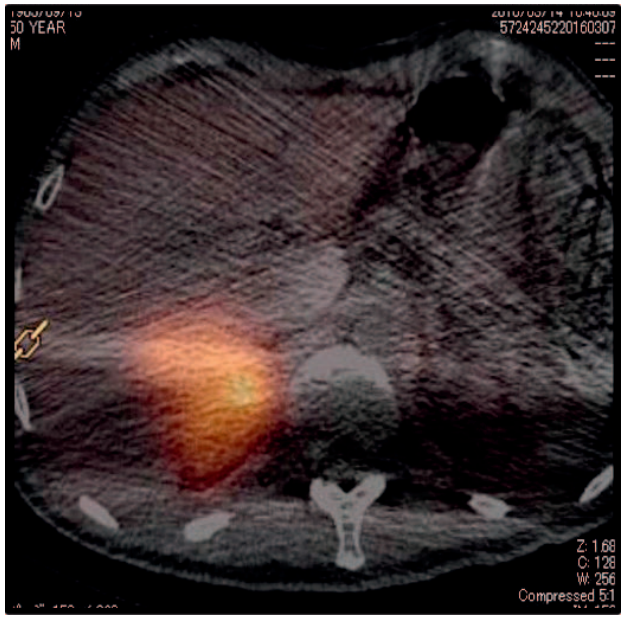

Fig. 3 MIBG imaging reveals accumulation in the tumor, but no metastatic or heterotopic lesions.

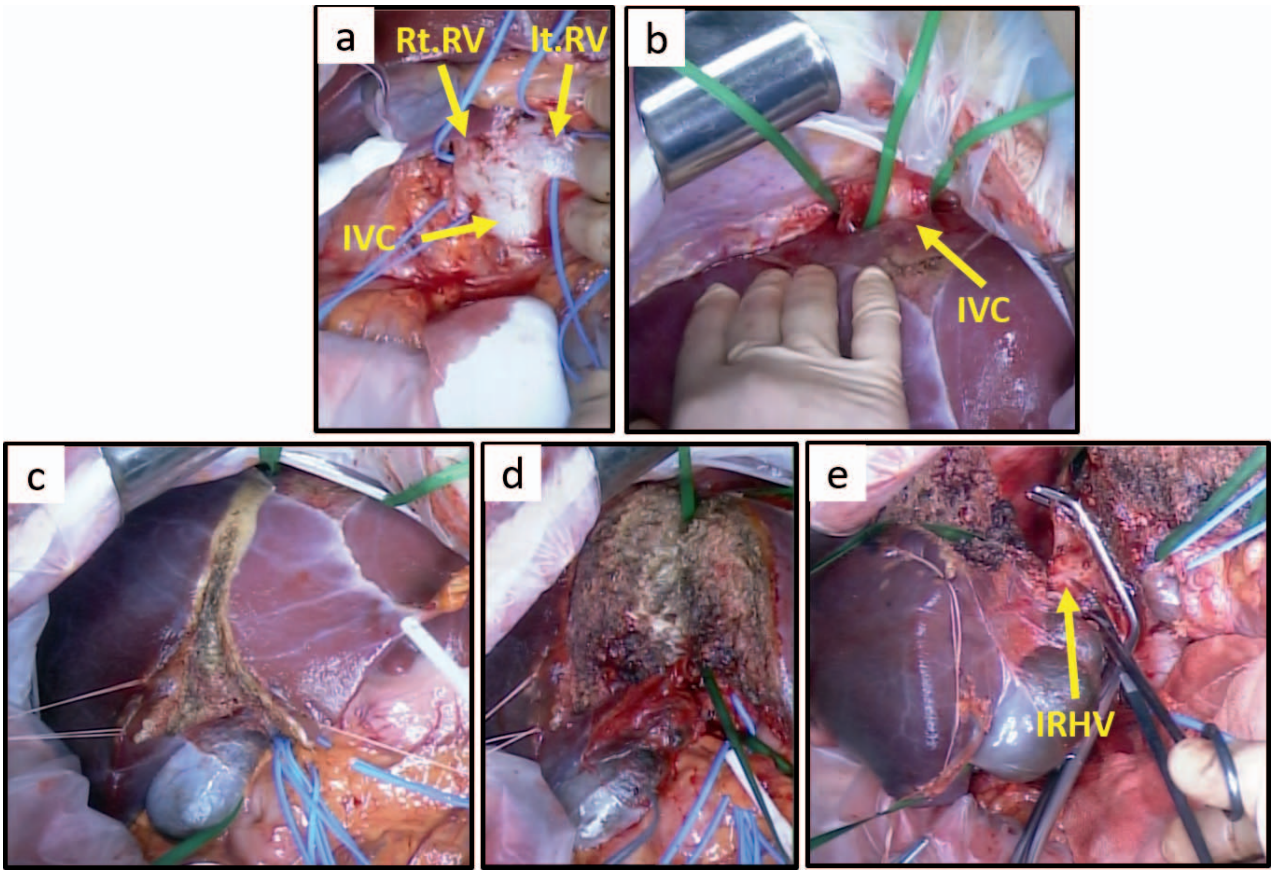

Fig. 4 Intraoperative findings. We exposed the IVC and bilateral renal veins (rt. RV, lt. RV) on the posterior surface of the liver (a) and the IVC on the cranial surface of the liver (b). Right hepatectomy was performed using the liver hanging maneuver (c, d). We dissected and closed the inferior right hepatic vein (IRHV) by side clamping of the IVC (e).

Gross appearance revealed tumor invasion into the liver (Fig. 5a and 5b). Histopathologic examination showed diffuse proliferation of dyskaryotic cells (Fig. 6a and 6b). Ki-67 labeling index was less than 5%, but tumor had infiltrated the liver, so malignant pheochromocytoma was diagnosed (Fig. 6c).

Just after surgical resection, hypotension appeared. Otherwise, circulation dynamics gradually improved, and blood pressure was maintained without catecholamines on postoperative day 2. On follow-up at 12 months postoperatively, the patient showed a good prognosis with no signs of relapse.

Discussion

The liver hanging maneuver is a surgical technique that facilitates anterior approaches to the IVC during hepatectomy, using tape passed in front of the anterior wall of the IVC and pulled upward during parenchymal transection.[2,3] Using the liver hanging maneuver, we were able to expose the avascular space along the anterior surface of the retrohepatic IVC[4] without needing to mobilize the right lobe. This approach requires no compression of the remnant liver[3] and improved visualization for resection.[5] Blood loss related to the anterior approach is controllable, because the blood supply comes from the liver and the retroperitoneal tumor.[5] We can thus avoid the risk of troublesome bleeding. This anterior approach to the IVC with the liver hanging maneuver is reportedly effective in resecting large retrohepatic tumors as one lump with the right hemiliver.[5,6]

Pheochromocytoma, a neuroendocrine tumor of chromaffin cells from the adrenal medulla,[7] usually presents with the clinical triad of paroxysmal headache, profuse sweating and palpitations, associated with labile hypertension.[8] A key characteristic of malignant pheochromocytoma is the presence of

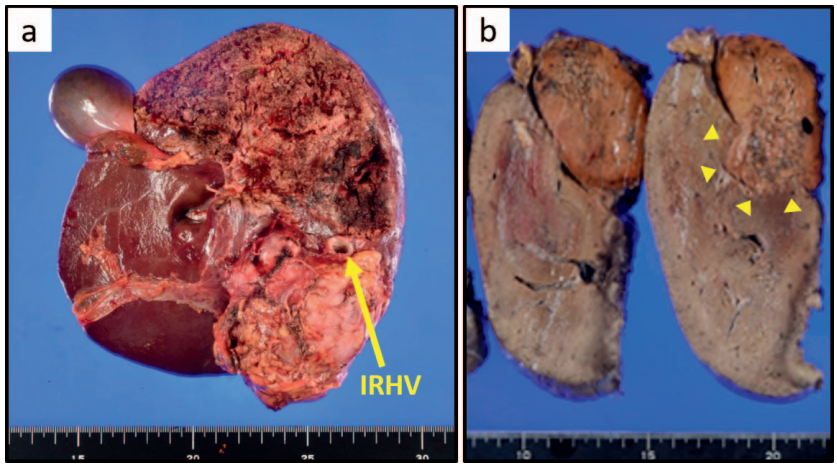

Fig. 5 Gross appearance. Gross examination reveals tumor invasion to the liver (yellow arrowheads) (a, b).

metastases that spread to organs lacking chromaffin cells.[9] Ki-67 labeling index is at least helpful in identifying neoplasms with an increased risk of recurrence,[10] but reported cut-off values for Ki-67 expression vary widely, ranging from 2% to over 5%.[11,12]

The literature on adrenal malignant pheochromocytoma invading the liver remains limited. If

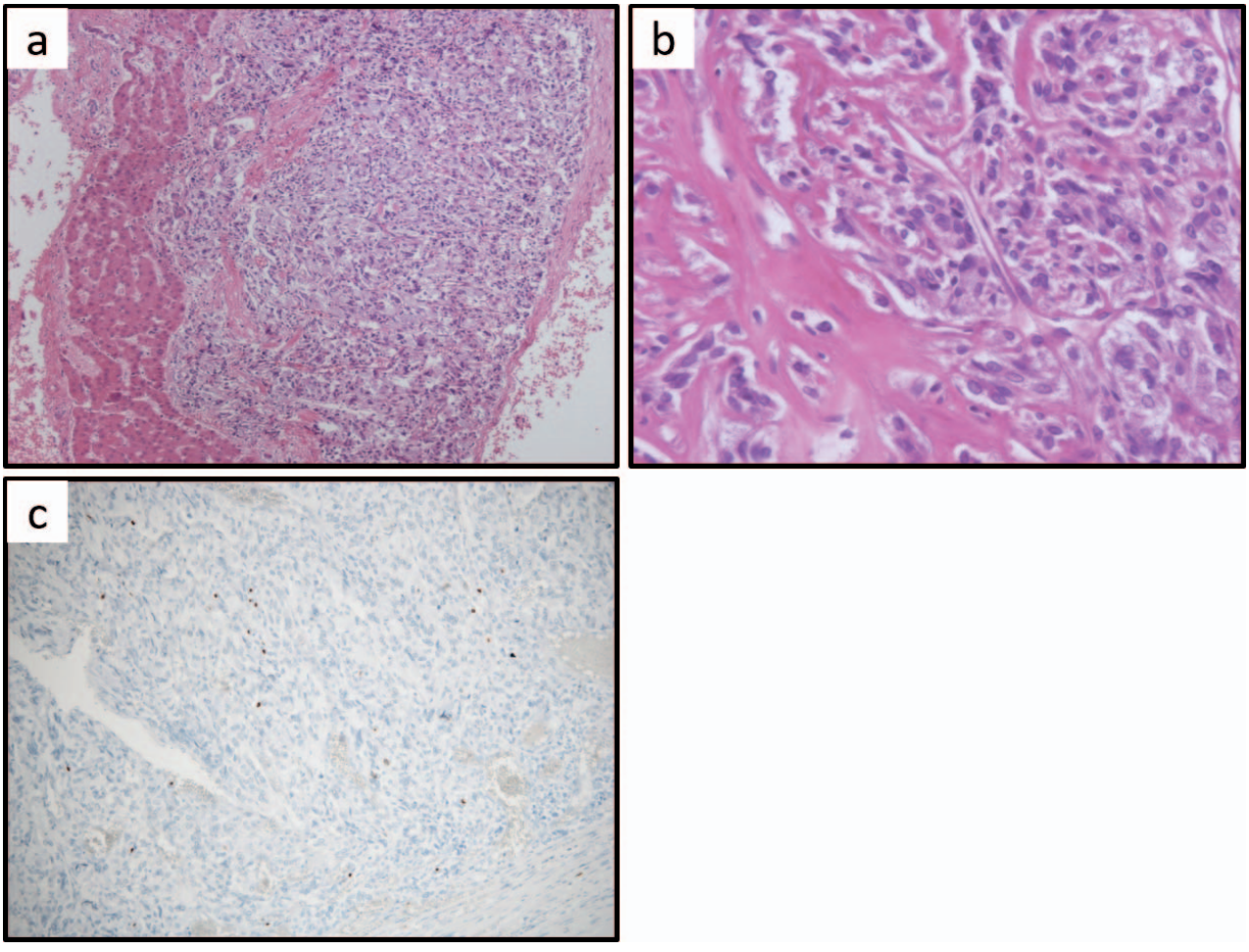

Fig. 6 Histopathological examination shows diffuse proliferation of dyskaryotic cells (a, b). Ki-67 labeling index is <5% (c).

adrenal tumor invades or adheres to adjacent organs such as the liver, IVC, kidney, spleen, and pancreas, margin-free resection represents a strong predictor of long-term survival. In such cases, en bloc resection of surrounding organs is often required to achieve R0 resection and prevent tumor spillage.[13]

During surgery for pheochromocytoma, stimulation of the tumor should be avoided to prevent excess secretion of catecholamines.

In the current case, the patient experienced severe heart failure, so hypertension during the operation could well have proven fatal. If we had performed right hepatectomy under the usual method, we would have mobilized the right lobe of the liver, risking excess secretion of catecholamines. We thus performed right hepatectomy using a transhepatic anterior approach and minimized stimulation of the tumor. We were thus able to finish the operation safely without clamping the IVC or using PCPS. In addition, we were able to resect the tumor as one lump with the right hemiliver and achieved R0 resection.

Large adrenal or retroperitoneal tumors, especially arising from the right adrenal gland, tend to invade the IVC because of the anatomical position.[14,15] For large retroperitoneal tumors behind the liver, including adrenal tumor with suspected invasion to the IVC, right hepatectomy using an anterior approach is safe to perform and widely exposes the anterior surface of the IVC. Of course, few reports have described retroperitoneal tumor suspected to have invaded the IVC but showing no invasion to the IVC intraoperatively or histologically.[15]

In the current case, tumor infiltration to the IVC was suspected preoperatively, but intraoperative findings showed tumor invasion was limited to the root of the IRHV and we were able to separate the tumor from the IVC and thus preserved the IVC.

Conclusions

We encountered a case of malignant pheochromocytoma invading the right posterior lobe of the liver with severe cardiac failure. We safely applied an anterior transhepatic approach to the IVC using the liver hanging maneuver, which seems effective in such cases.

Acknowledgments

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