Successful outcome after resection of liver metastasis arising from an extraadrenal retroperitoneal paraganglioma that appeared 9 years after surgical excision of the primary lesion

Abstract A 54-year-old man underwent surgery for excision of a retroperitoneal tumor that measured 12 cm in diameter, and histopathological examination revealed the tumor was an extraadrenal retroperitoneal paraganglioma. He presented 9 years later with epigastric discomfort. Abdominal ultrasound showed a solitary liver tumor. The diagnosis, based on radiological workup, was metastatic paraganglioma. The tumor was surgically resected and the histological findings resembled those of the primary tumor. The patient has been followed up for 3 years and remains recurrence-free. Surgical resection is an effective treatment approach for primary and secondary paragangliomas, but the resectability of liver metastatic lesions is usually low, although complete resection with a wide surgical margin was possible in this patient. This case suggests that a good prognosis after the resection of hepatic metastasis depends not only on the curative resection of the metastatic lesion but also on the tumor characteristics, such as slow growth or low aggressiveness.

Key words Extraadrenal paraganglioma · Liver metastasis · Retroperitoneum · Operation

Introduction

Extraadrenal paragangliomas are unusual neoplasms that have been described in many anatomic sites. Extraadrenal locations include the head and neck area, mediastinum, retroperitoneum, gallbladder, extrahepatic bile ducts, urinary bladder, and spinal region. The behavior of extraadrenal retroperitoneal paragangliomas is known to be more malignant than the behavior of those found elsewhere. Malignant paragangliomas exhibit hematogenous spread, and some patients show distant metastases at presentation, with the most common sites being bone, liver, and lungs. Once distant metastases occur, the median survival is reported to be 16–34 months. As for the treatment of paragangliomas, surgical resection is the only effective treatment modality for primary and secondary tumors.

Here, we present a case of a liver metastatic tumor originating from extraadrenal retroperitoneal paraganglioma; the metastatic tumor was diagnosed 9 years after resection of the primary lesion, and was successfully treated by surgical resection.

Case report

A 54-year-old man underwent surgical resection of a retroperitoneal tumor at Osaka University Hospital in 1995. Preoperative abdominal computed tomography (CT) showed a large and hypervascular tumor in the left upper abdominal cavity (Fig. 1a). The tumor size was 12 cm in diameter and its blood supply originated from the splenic and celiac arteries. The operation was performed safely and no hypertensive crisis was experienced perioperatively. Histopathologically, the mass was entirely covered with a fibrous capsule (Fig. 1b), and its architectural pattern resembled “zellballen”. The size of the tumor cells varied and the nuclei of the tumor cells were round or oval; some tumor cells had large nuclei or multiple nuclei (Fig. 1c). The cytoplasm was clear. Immunohistochemical examination showed positive reaction for chromogranin A. Based on the above findings, the tumor was considered to be an extraadrenal retroperitoneal paraganglioma without malignant changes (such as mitosis, infiltration of the fibrous capsule, or
vascular invasion). The patient’s clinical course was uneventful for 9 years after the surgery.

In June 2004, the patient presented with epigastric discomfort. Abdominal ultrasound examination revealed a hepatic tumor. He was admitted to our hospital for further evaluation of the tumor. Physical examination was normal. Laboratory tests showed normal results for biochemical tests with negative viral markers. Serum catecholamine concentrations were normal, although 24-h urine catecholamine was not examined. The level of neuron-specific enolase (NSE), a tumor marker, was slightly elevated, at 10.2 ng/ml. Abdominal CT showed a solitary and hypervascular tumor with central necrosis in segments 8 and 4 of the liver (Fig. 2a). Magnetic resonance imaging (MRI) showed low-intensity signal in T1 weighted images and high-intensity signal in T2 weighted images (Fig. 2b, c). Angiography showed that the tumor was supplied by branches from the right and left hepatic arteries. No other lesions were detected. 131I-Metaiodobenzylguanidine (MIBG) scintigraphy showed positive uptake of MIBG in the tumor in segments 8 and 4 (Fig. 2d). 18F-Fluorodeoxyglucose (FDG) positron emission tomography showed FDG uptake in the tumor, but no uptake in any other areas of the liver or other organs.

Based on these findings, the diagnosis was a solitary hepatic metastatic tumor of extraadrenal paraganglioma. Accordingly, the patient underwent tumor resection, with a sufficient surgical margin. No hypertensive crisis developed before or during surgery. The hepatic mass was visible from the liver surface, and was encapsulated by a thin layer of the serosa. It measured 8.2 × 6.0 × 6.0 cm. The cross-section showed a mass consisting of peripherally solid areas together with pockets of cystic degeneration (Fig. 3a). The histological findings were consistent with a metastatic lesion from the extraadrenal paraganglioma resected 9 years earlier (Fig. 3b, c; H&E staining). Infiltration of the capsule was evident, and the nontumorous tissue was normal liver tissue. Immunohistochemical examination showed positive immunoreactivity for chromogranin A (Fig. 3d) and synaptophysin. The final diagnosis was metastatic paraganglioma.

The postoperative course was uneventful. At the last follow-up examination, 3 years after the second surgery, the patient was asymptomatic and free from any sign of recurrence.

**Discussion**

Extraadrenal paragangliomas typically exhibit variable clinical features. The majority of these tumors are functional and actively produce catecholamines. The clinical signs and symptoms are most likely caused by excess catecholamine secretion, and such excess secretion has been reported in 36% to 65% of patients. Moreover, extraadrenal paragangliomas are associated with a high rate of malignancy, ranging from 14% to 50%. Malignant paragangliomas demonstrate both hematogenous and lymphatic spread, with the most common sites of distant metastases being bone, liver, and lungs. Sclafani et al. reported a series of 22 cases of extraadrenal paragangliomas. In their report, 11 of the 22 (50%) patients developed distant metastases. The most frequent sites of metastasis were bone (7/11), liver (3/11), and peritoneum (2/11). The median time to first metastasis after excision of the primary tumor was 2 months, but the time to the first metastasis in 2 patients was more than 7 years. Morikawa et al. reported a case of adrenal...