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CASE REPORT

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Diaphragmatic paraganglioma protruding into the right thoracic cavity

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Abstract

Paragangliomas in the diaphragm are extremely rare. We report the case of a 27-year-old woman with a nonfunctioning paraganglioma protruding superiorly from the right diaphragm. The patient underwent an anterior thoracotomy, and a supradiaphragmatic tumor (70 mm in diameter), which compressed the inferior vena cava and the right hepatic vein, was completely resected by combined partial resection of the right diaphragm and pericardium. To our knowledge, this is the first report of a paraganglioma situated both on the diaphragm and close to the inferior vena cava and hepatic vein.

KEYWORDS

combined resection, diaphragmatic paraganglioma, hepatic vein

INTRODUCTION

Paragangliomas are rare tumors that originate from the neuroendocrine cells of extra-adrenal tissue. Although paragangliomas may arise in any paraganglia of the autonomic nervous system, most paragangliomas occur in the infra-diaphragmatic para-aortic area, and thoracic paragangliomas are extremely rare.[1] The mediastinum around the ascending aorta is a relatively common site of paraganglioma occurring in the thorax.[1–3] However, no previous study has reported diaphragmatic paragangliomas protruding into the thoracic cavity. We report a patient with a paraganglioma beyond the right diaphragm, which was completely removed by the combined resection of the pericardium and diaphragm.

CASE REPORT

A 27-year-old woman with an abnormal radiographic chest shadow was referred to our hospital. She had no symptoms, and her blood pressure and heart rate were unremarkable. There was no family history of paraganglioma. Laboratory data including catecholamines were all within the normal range. Contrast-enhanced computed tomography (CT) revealed a well-demarcated mass, which was 70 mm in diameter, located above the right diaphragm (Figure 1(a),(b)). Hyperdynamic enhancement continuing from the early to delayed phase was observed on CT. Many feeding arteries from the diaphragm and mediastinum were found around the tumor. The tumor was adjacent to the right lung and right atrium, and compressed the inferior vena cava (IVC) and the right hepatic vein (RHV) (Figure 1(b)). Enhanced magnetic resonance imaging (MRI) showed punctate areas of both low and hyperintense signal intensities on both T1- and T2-weighted images. Moreover,18F-fluorodeoxyglucose-positron emission tomography (FDG-PET) revealed a high uptake of FDG with a maximal standardized uptake value of 54.1 (Figure 1(c)). Accordingly, the tumor was suspected to be a paraganglioma, and surgery was scheduled.

The operation was performed under a half-left side-lying position with general anesthesia using one-lung ventilation. Anterior thoracotomy via the 6th intercostal space was performed. Intraoperatively, the tumor arose from the right diaphragm and densely adhered to the pericardium. Although the tumor was adjacent to the right lung and IVC, no invasions or adhesions were found. The phrenic nerve was resected at the most distal portion because of severe

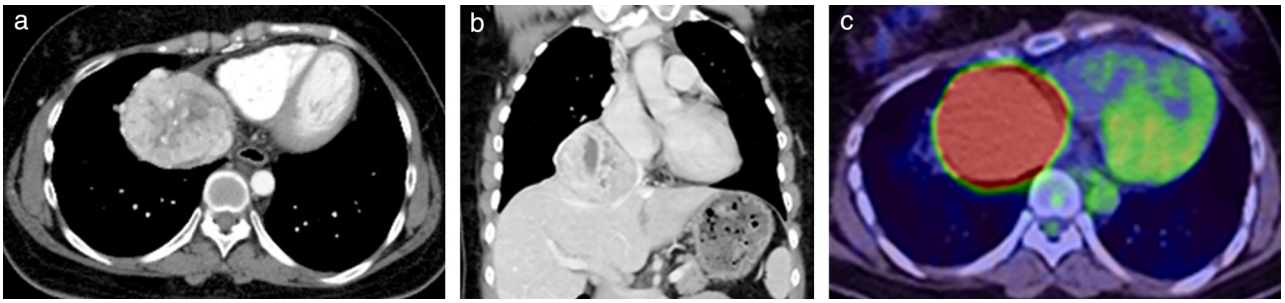

FIGURE 1 (a) Enhanced computed tomography (CT) revealed a mass 70 × 60 mm, which was a hyperdynamic enhancement, in diameter beyond the right diaphragm; (b) coronal section of the enhanced CT showed that the tumor compressed the right atrium, inferior vena cava, right lobe of the liver, and hepatic vein; and (c) 18F-fluorodeoxyglucose-positron emission tomography (FDG-PET) revealed a high uptake of FDG with a maximum standardized uptake value of 54.1 in the tumor

adhesion to the tumor. The right diaphragm was incised, and the liver was exposed. Tumor invasion into the liver was not observed. Many feeding arteries from the inferior phrenic artery were divided. Although the tumor was in broad contacted with RHV, no invasion was observed. Many veins draining to the RHV were found and divided. Furthermore, the RHV wall was partially resected and reconstructed by direct suture due to dense adhesion to the tumor after side clamping the RHV. The right diaphragm and pericardium were partially resected with the tumor, and the tumor was completely removed. Defects in the diaphragm and pericardium were repaired with Gore-Tex sheets.

The tumor was well-circumscribed and microscopically composed of cells arranged in a characteristic nesting pattern, named Zellballen (Figure 2), which is not found in neurologic tumors other than paragangliomas. No infiltrations to the pericardium and peritoneum were observed. The margin of the specimen was negative for tumor cells. On immunohistochemical examination, the tumor tested positive for CD 56, chromogranin, synaptophysin, and adrenomedullin. S100 was positive for only sustentacular cells. The Ki-67 labeling index was <2%. Based on these pathological findings, the tumor was diagnosed as a paraganglioma.

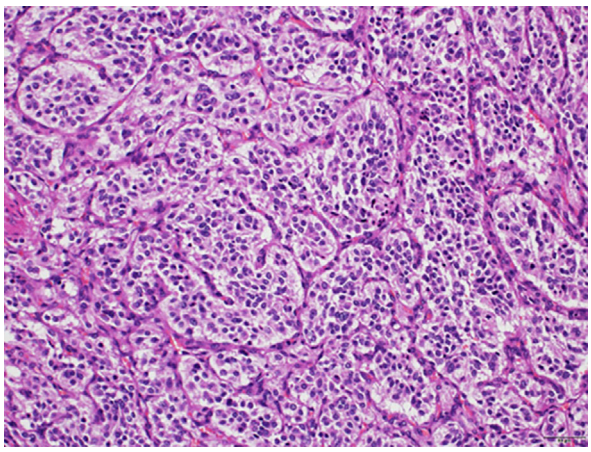

FIGURE 2 Hematoxylin and eosin staining of the resected specimen

The tracheal tube was removed at the operating room postoperatively, and the patient left the intensive care unit on postoperative day 1. The postoperative course was uneventful, and the patient was discharged after 12 days. No postoperative adjuvant therapy was performed. At the 1-year postoperative follow-up, no evidence of disease recurrence was observed. The diaphragm was elevated as noted on the chest radiograph, but no increase in pleural and pericardial effusion was observed during follow-up. Although postoperative lung function was not evaluated, the patient has not complained of dyspnea.

DISCUSSION

To our knowledge, only two cases of primary diaphragmatic paragangliomas have been reported.[4,5] These two tumors were located below the diaphragm, and no studies have reported diaphragmatic paragangliomas developing superiorly. Contrary to prior cases, our patient showed a nonfunctional paraganglioma.

Surgery is the only curative treatment for paraganglioma. Although complete resection is necessary to obtain long-term survival,[6] it is sometimes difficult to perform in cases of intrathoracic paraganglioma because of the proximity to important vascular structure.[3] In our case, since the tumor was close to the IVC, RHV, and liver, we consulted both cardiovascular and liver surgeons preoperatively. If the tumor invasion into the right lobe of the liver or the RHV had been found intraoperatively, the right branch of the portal vein would have been ligated to induce left lobe hypertrophy for the right hepatic lobe resection without tumor resection. In that case, a second surgery to remove the tumor would have been performed by the combined resection of the IVC and the right hepatic lobe using a cardiopulmonary bypass after left lobe hypertrophy. However, no infiltrations to the liver and RHV were observed intraoperatively, and the tumor was resected without combined resection of the IVC and liver. Preoperative embolization, which was not performed in our case, has been reported to be useful to reduce perioperative

hemorrhagic complications.[7] This technique might be considered especially in patients with a bulky paraganglioma with large feeding arteries.

Since the biopsy of a paraganglioma using mediastinoscopy or video-assisted thoracoscopy can cause massive hemorrhage because of the hypervascularity of the tumor,[3,6] radiologic diagnosis should be favored. Radiologic findings of CT and MRI are beneficial for diagnosing paragangliomas, distinguishing it from other tumors. Enhanced CT usually reveals a heterogeneous mass with enhancement and sometimes a low attenuation of central areas, which represent tumor degeneration or necrosis.[8] Contrast- enhanced MRI showed a "salt-and-pepper" appearance on both T1- and T2-weighted images. This was due to the presence of punctate areas of low-signal intensity corresponding to the flow voids from tumor vascularity as well as the presence of hyperintense signals corresponding to areas of hemorrhage in the tumor.[8] On FDG-PET, a FDG high uptake is usually observed.[7] Since these characteristic radiologic findings were observed in our case, the tumor was strongly suspected to be a paraganglioma.

A previous report demonstrated that 60% of patients with paragangliomas had metastatic diseases, and metastatic sites include lymph nodes, liver, bones, etc.[1] It was also reported that increased tumor size was associated with metastasis and shorter overall survival.[1] Although the protocol of follow-ups postoperatively for paragangliomas regarding modality and frequency has not been determined, careful follow-up is necessary in our case since the tumor size was relatively large.

We report a patient who underwent the complete resection of a paraganglioma occurring in the diaphragm. Owing to the location of the boundary area, a collaboration between cardiovascular and liver surgeons was necessary.

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DISCLOSURE

None.

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