cases and ulcerative in three of ten cases. It seems that esophageal collision tumor grows more easily into the cavity, which will cause the symptom of dysphagia. Seven of ten cases occurred in the lower third the esophagus, unlike the common esophageal cancer, which most commonly affects the middle third part: 72% (1946/2704) in our department from 2005 to 2012. Collision tumor often occurs in the gastroesophageal junction. Five of the 7 patients whose tumors were in the lower third of the esophagus had both Barrett’s esophagus and adenocarcinoma, so the presence of Barrett’s metaplasia may predispose to the development of esophageal collision tumor composed of adenocarcinoma. Biopsy of the tumor by preoperative endoscopy did not show good diagnostic results. In fact, only two of ten cases were given the correct diagnosis of collision tumor by biopsy at that time, whereas in the other cases another type of collision tumor was missed. Definitive diagnosis was made by histologic examination. Spagnolo and colleagues [8] have proposed these guidelines for diagnosing collision tumors: (1) Two distinct topographically separate sites of origin for the two components must be present. (2) There must be at least some separation of the two components so that, despite intimate mixing at points of juxtaposition, dual origin can still be recognized. (3) At the areas of collision, in addition to intimate mixing of the two components, some transitional patterns may be seen. The most common components of esophageal collision tumors are squamous cell carcinoma and adenocarcinoma, and they also are the most common tumors in esophageal cancer. However, four cases included the component of small cell carcinoma, whereas the incidence of small cell carcinoma in esophageal cancer is 0.9% in our department. The reason why small cell carcinoma is so common in esophageal collision tumor is unclear. Furthermore, adenoid cystic carcinoma was present in one of our cases, which is uncommon in esophageal cancer; it occurred in 0.07% of the tumors of esophagus in our department. The depth of invasion is to the lamina propria in one case, the submucosa in four cases, the muscularis propria in two cases, and the adventitia in one case. The different components of the other two cases had different depths of invasion: the submucosa and the muscularis propria, respectively. It seems that small cell carcinoma more easily invades deeply. Because of the rarity of esophageal collision tumor, we cannot give an exact prognosis, but collision tumor usually has an unfavorable prognosis. The prognosis appears to depend on the stage of the tumor at the time of initial diagnosis and the type of components. Collision tumors are clinically relevant in that the individual tumors may require different treatments. Therefore, physicians should be aware of collision tumor and not overlook the second primary tumor.

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References


Asymptomatic but Functional Paraganglioma of the Posterior Mediastinum

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A 72-year-old woman was referred to our hospital because of a posterior mediastinal tumor. On the basis of detailed imaging tests, including 123I-metaiodobenzylguanidine single photon emission computed tomography-computed tomography, and elevated values of catecholamines in the plasma and urine, the tumor was diagnosed as a functional mediastinal paraganglioma even in the absence of symptoms. After preoperative blood pressure control, surgical resection was performed. During the operation, the systemic blood pressure increased transiently as a result of surgical manipulation of the tumor. Soon after the tumor was removed, the patient conversely experienced hypotension. The postoperative course was uneventful, and pathologic diagnosis revealed a paraganglioma.


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pheochromocytomas located outside the adrenal glands are referred to as paragangliomas. Mediastinal paragangliomas are rare, slow-growing neurogenic tumors. Twenty percent of patients with paraganglioma have symptoms referable to catecholamine-secreting tumor (palpitation, headache, or sweating), and most patients present with mass effect symptoms or an incidental finding [1]. The management of functional paragangliomas is difficult because of complications such as hypertension, tachycardia, arrhythmia, myocardial dysfunction, hyperglycemia, and sudden hypotension after tumor removal caused by intravascular volume depletion [2]. Therefore, correct pharmacologic perioperative management is indispensable. Regardless of the functional tumor, the patients with paraganglioma are sometimes asymptomatic [3]. In such cases, unexpected intraoperative elevation of the blood pressure may cause a critical situation. Thus, preoperative diagnosis of the functional paraganglioma is preferable, although the asymptomatic cases that are diagnosed preoperatively are rare. Here we report a case of functional paraganglioma arising from the posterior mediastinum, which manifested no symptoms related to the tumor but was diagnosed by the detailed diagnostic imaging tests.

A 72-year-old woman was referred to our hospital for surgical treatment of a posterior mediastinal tumor, which had gradually enlarged after a follow-up time of 7 years. She had no history of hypertension or other specific signs or symptoms, including tachycardia. Contrast medium–enhanced computed tomography (CT) of the chest showed a left posterior mediastinal tumor with hyper-vascularity (Fig 1A). Because this finding suggested the possibility of paraganglioma as the differential diagnosis, further diagnostic imaging analyses were performed. High uptake was shown only in the posterior mediastinal tumor by 123I-metaiodobenzylguanidine (MIBG) single photon emission computed tomography (SPECT)-CT (Fig 1B). Positron emission tomography (PET)-CT demonstrated the tumor with strong fluorine [F-18]-fluorodeoxy-D-glucose (FDG) activity, with a maximum standardized uptake value (SUV$_{\text{max}}$) of 38.03 (Fig 1C). Moreover, blood and urine examination revealed elevated levels of the following catecholamines: serum norepinephrine of 1.77 ng/mL (normal range, 0.1–0.5 ng/mL), and urinary norepinephrine of 671 μg/day (normal range, 31–160 μg/day), vanillyl mandelic acid of 9.0 mg/day (normal range, 1.5–4.3 mg/day), metanephrine of 0.39 mg/day (normal range, 0.04–0.18 mg/day), and normetanephrine of 0.81 mg/day (normal range, 0.1–0.28 mg/day). On the basis of these findings, the tumor was diagnosed as functional mediastinal paraganglioma, although the patient was asymptomatic.

Preoperative medical management included blood pressure control with an α-adrenergic blocker and intravascular volume expansion. She started medication of doxazosin with 1 mg daily, titrated up to 8 mg daily in 3 weeks before the operation.

Complete surgical resection was undertaken by video-assisted thoracic surgery (VATS) through four ports by use of a vessel-sealing device. Although transient elevation of the systolic blood pressure to 190 mm Hg was observed when the tumor was being dissected from the deepest portion of the chest wall near the intercostal vessels, the hypertension status was improved soon after the intercostal vessels were sealed. As a consequence, surgical resection was completed without any critical situation arising. Of note, particular vessels that were connected to the tumor were not identified. After the tumor was completely resected, the patient immediately experienced hypotension, with a systolic blood pressure of 75 mm Hg, requiring norepinephrine to maintain adequate blood pressure during the operation and on the first day of her stay in the intensive care unit. Her postoperative course was uneventful, and the urine norepinephrine levels normalized on the seventh postoperative day.
The excised tumor was encapsulated and of firm consistency, with a maximum diameter of 2.2 cm, and had a yellow-brownish color. Histopathologic examination of the tumor revealed that the tumor cells were arranged in nests (Zellballen). Necrosis was not seen, nor was mitosis in high-power field or invasion into the fibrous capsule (Fig 2A). Immunohistochemical staining was positive for chromogranin (Fig 2B) and synaptophysin (Fig 2C) and focal positive for protein S-100 (Fig 2D).

Comment

Paragangliomas are rare tumors arising from the paraganglia associated with the autonomic nervous system. Mediastinal paragangliomas are uncommon (2%) and are most frequently located in the anterior mediastinum but less commonly in the posterior mediastinum. About 50% of paragangliomas of the posterior mediastinum are functional [4]. Functional mediastinal paragangliomas are often discovered during the surveillance of hypertension and other symptoms secondary to catecholamine secretion, such as headache, palpitation, sweats, and tremor [1]. Nonfunctional mediastinal paragangliomas are asymptomatic and are usually found incidentally. However, in the present case, the patient was asymptomatic, although the paraganglioma was later proved to be functional. Thus, it is important to consider the possibility of paraganglioma from the findings of the imaging tests. Paragangliomas show marked contrast agent enhancement on computed tomography or magnetic resonance imaging. Additionally, $^{123}$I-MIBG SPECT/CT or FDG-PET imaging is useful for localization and staging. It is reported that $^{123}$I-MIBG SPECT/CT is an important nuclear imaging method in paragangliomas, and the specificity and the sensitivity are 99% and 90%, respectively [5].

Complete surgical resection remains the standard therapy for paraganglioma because of the malignant potential of the tumor and its resistance to chemotherapy or radiation. Although surgical treatment is recommended, it should be noted that paragangliomas are hypervascular tumors, adhere to adjacent structures, and are located close to the great vessels. Therefore, the operation can lead to serious bleeding. Some authors recommend preoperative embolization to reduce intraoperative bleeding [6]. On the contrary, there is a possibility that tumor necrosis may induce an uncontrollable hypertensive crisis through catecholamine release by preoperative embolization [7]. In our case, we could complete the tumor removal by the VATS approach using a vessel-sealing system, although conversion to thoracotomy should not be delayed in case of massive bleeding.

Furthermore, in cases of catecholamine-secreting tumors from the findings of blood and urine examination, preoperative management such as antihypertensive therapy with adrenergic blockers ($\alpha$-blocker or $\beta$-blocker), calcium channel blockers, and volume loading is important to prevent hypertensive crises caused by manipulation of the tumor and hypotensive crises after the complete interruption of venous drainage from the tumor.

In conclusion, we treated a case of asymptomatic but functional paraganglioma arising from the posterior mediastinum. Although a mediastinal paraganglioma, especially a functional one, is uncommon, it should be included in the differential diagnosis of a mediastinal tumor. Preoperative precise diagnosis of paraganglioma...
and the following appropriate preoperative management led to safe performance of the operation.

References