Aortico-pulmonary Paraganglioma
Case Report and Japanese Review

Aortico-pulmonary paraganglioma (APPG) is a rare middle mediastinal tumor. We experienced a case of APPG in a 52-year-old man. Chest computed tomography and magnetic resonance imaging revealed a multi-cystic mass in the subaortic area. A left thoracotomy was performed without definitive preoperative diagnosis. The tumor was strongly adherent to the pericardium and the surrounding large vessels, but a complete resection was undertaken. Histological and immunohistochemical examination revealed that it was a malignant paraganglioma with a microscopically positive surgical margin. After radiotherapy of 50 Gy for the mediastinum, the patient almost recovered from his hoarseness by thyroplasty. We also reviewed nine Japanese cases of APPG reported previously. Though APPG is rare, we must consider that a middle mediastinal tumor may be APPG, and preoperative examination and preoperative planning are necessary to prevent massive bleeding and microscopic residual tumor.

Key words: aortico-pulmonary paraganglioma, middle mediastinal tumor, Japanese reported cases

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Aortico-pulmonary paraganglioma (APPG) is a rare neoplasm of extraadrenal paraganglion arising in the middle mediastinum. To date, only nine cases of APPG have been reported in Japan. We present a case of APPG with postoperative radiotherapy and review Japanese reported cases.

Case

A 52-year-old Japanese man consulted an otolaryngologist in our hospital suffering from hoarseness for half a month. He had a 32-year history of smoking. A left recurrent nerve paralysis was diagnosed by the otolaryngologist. He was transferred to us for further management.

There were no abnormal cardiopulmonary findings on physical examination. The patient's blood pressure was 104/68 mmHg, his pulse was regular and 60 beats/min, and his body temperature was 36.4°C.

The patient's chest X-ray showed a protrusion of the root of the left pulmonary artery (Fig. 1). The chest computed tomography (CT) revealed a mediastinal multi-cystic mass of 45 mm diameter, which was located in the aortico-pulmonary window (A-P window) along the left main pulmonary artery (Fig. 2). The magnetic...
Fig. 2. Chest CT scan shows multi-cystic ring-enhanced lesion from the subaortic area (A) to the proximal portion of the left main pulmonary artery (B).

The chest CT scan also confirmed a cystic mass in the A-P window (Fig. 3). There were no abnormal findings on abdominal echogram, upper gastrointestinal fiberscopy, or fibroptic bronchoscopy.

There were no abnormalities on urinalysis and blood cell count. The erythrocyte sedimentation rate was mildly elevated (30 mm/hr). Serum catecholamines were not evaluated preoperatively. Except for the slight elevation of C-reactive protein (1.0 mg/dl) and triglyceride (249 mg/dl), there were no abnormal values on blood biochemical examination. The patient had normal pulmonary function (VC: 3,850 ml, %VC: 102.7%, FEV1,0: 3,430 ml, and FEV1.0: 90.3%).

At thoracotomy, the tumor was found to be adherent to the ascending aorta, the proximal portion of the left main pulmonary artery, and the left recurrent laryngeal nerve in the A-P window. The frozen-section specimen, which was obtained by wedge resection of the tumor, was diagnosed as a "germ cell tumor", and tumor resection was performed. The removed tumor with pericardium was adherent to the left recurrent laryngeal nerve. The intraoperative blood loss was 310 ml.

The tumor was 45x45x28 mm in size and the weight was 27 g and it consisted of large and small cystic parts. Microscopically, the tumor cells with abundant granular cytoplasm formed nests, which were separated by vascular septa (Zellballen) (Fig. 4). Distinct granules were detected within the tumor cell cytoplasm by argyrophilic staining. Immunohistochemical staining revealed that neuron-specific enolase (NSE) and chromogranin were positive and carcinoembryonic antigen (CEA) and keratin were negative for tumor cells, and that S-100 protein was negative for sustentacular cells. The tumor was therefore diagnosed as APPG on its anatomical location. Only a small part of the surgical margin was positive, contacting the aortic surface. The tumor was diagnosed as malignant due to its histological atypism and local invasion.

In his postoperative course, the patient suffered from aspiration pneumonia several times because of the left recurrent laryngeal nerve paralysis. However, he recovered gradually with administration of antibiotics and swallowing training. Postoperative irradiation of 50 Gy was performed on his mediastinum. He was able to talk after thyroplastic surgery and was discharged. He is currently alive and well 16 months after resection.

Discussion

A paraganglion is a small cell nest belonging to the autonomic nervous system. It is divided into two groups;