A Completely Resected Paraganglioma Arising From the Right Atrioventricular Groove

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We report a case of completely resected right atrioventricular groove paraganglioma. Primary cardiac paragangliomas are extremely rare. Only about 60 cases have been reported in the literature, and most of these tumors originated from the left atrium or the base of the aortic root and pulmonary artery; 5 cases originated from the right atrium. In our patient, the paraganglioma was largely located in the right atrioventricular groove and protruded into the right atrial wall. To our knowledge, there has been no paraganglioma reported in the right atrioventricular groove. We successfully carried out complete tumor resection. Most of these tumors are locally invasive, and complete resection is often difficult.

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Paraganglioma in the chest is very rare, accounting for 1% to 2% of systemic paragangliomas, mostly located in the posterior mediastinum. Primary cardiac paraganglioma is extremely rare [1]. Paragangliomas have been described in different parts of the heart and usually occur in the left atrium or the base of the aortic root and pulmonary artery [2, 3]. There are only 5 previous cases of primary paraganglioma arising from the right atrium in the world literature [4]. In our patient, the paraganglioma was largely located in the right atrioventricular groove and protruded into the right atrial wall. We successfully carried out complete tumor resection. To our knowledge, there has been no paraganglioma reported in the right atrioventricular groove.

A 53-year-old man presenting with shortness of breath, palpitations, and sweating for 2 months was admitted to our hospital in August 2008. Despite having a medical history of hypertension for 10 years, his blood pressure was kept under fairly good control by long-term oral use of antihypertensive medication (metoprolol and nifedipine). His heart rate stood at 96 beats per minute and systolic blood pressure ranged from 110 to 120 mm Hg. Physical examination was normal except that a mild diastolic murmur could be heard in the aortic valve auscultation area. Chest radiography indicated a cardiothoracic ratio of 0.586 and an enlarged right heart shadow. The echocardiogram revealed a mass located in the right atrioventricular groove adjacent to the tricuspid valves, which had a clear-edge heterogeneous internal echo, about 47 × 40 mm (Fig 1). Contrast-enhanced resonance imaging confirmed a solid tumor in the lateral part of the right atrium above the inferior vena cava. For further identification of the spatial relationship between the tumor and the right coronary artery, computed axial tomography angiography (CTA) was also performed and showed a mass measuring 56.8 × 33.9 mm that was located in the right atrioventricular groove. The mass had a well-defined outline and was crossed by the right coronary artery, which appeared normal visually (Fig 2). In addition, segmented computed tomography (CT) of the thorax and abdomen was performed, and no further anatomic anomaly was discovered. Although the definitive diagnosis failed to be established based on the imaging evidence available (the clear boundary and no obvious signs of distant metastasis), it was suspected that the mass was benign.

The patient underwent a median sternotomy and was placed on cardiopulmonary bypass (CPB) with bicaval cannulation. The aorta was clamped and cardioplegia solution was infused. Cardiac arrest lasted 20 minutes during the operation, and the patient was weaned from CPB without difficulty. A well-defined 63 × 44 × 25 mm tumor mass was completely excised; it was largely located in the atrioventricular groove and protruded into the right atrial wall (Fig 3). The tumor had no capsule, and we dissociated the tumor from the myocardial surface and carefully avoided damage to the right coronary artery. The patient's blood pressure was maintained within a stable range during the operation. Afterward, the tumor was identified as a paraganglioma by histologic examination; it was characterized by nests of granular cells and prominent vascularity. Immunohistochemical staining

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was also used, and positive results were obtained for the endocrine markers chromogranin and synaptophysin (Fig 4).

After excision of the tumor, the patient’s blood pressure remained normal without any further treatment and he was discharged 8 days after operation. We followed this patient for about 4.5 years, during which echocardiography was performed regularly and showed no reemergence of any tumor; cardiac function was normal as well. Importantly, the patient did not present with shortness of breath, palpitation, fatigue, or any other symptoms thereafter.

Comment

Primary cardiac paragangliomas are extremely rare clinically. No more than 60 cases have been reported to date, [3, 4] and in only 5 cases were tumors found to originate from the right atrium [4]. In our patient, the paragangliomas arose from the right atrioventricular groove, which, to our knowledge, is the first case reported in the world to occur in this location.

According to the presence or absence of catecholamine secretion, paragangliomas can be divided into functional and nonfunctional types. The former usually present with clinically intermittent hypertension, sweating, and palpitations, and catecholamines or their metabolites can be detected at increased levels in the blood and urine. The nonfunctional type is often asymptomatic, but when the tumor is large, it can produce pressure symptoms. According to the clinical manifestations of our patient, the paraganglioma should have been a functional type. However, because of our lack of knowledge about primary cardiac paragangliomas, the elevated catecholamine concentrations in blood and urine were not detected before the operation. Echocardiography and CT coronary angiography are useful in locating cardiac paragangliomas, and detection of excessive plasma or urinary catecholamines or their metabolites, which feature in paragangliomas, will be particularly helpful to the diagnosis before operation [5].

Cardiac paragangliomas normally feature a nodular appearance, lack of envelope, homogeneity, and soft texture. To our knowledge, although most of them are benign, malignant paragangliomas have also been reported. Technically, it is quite difficult to differentiate benign paraganglioma from malignant paraganglioma based solely on their morphologic characteristics. In terms of biological properties, malignant cardiac paragangliomas are invasive, with 10% to 20% of them having...
the potential to metastasize or intrude into adjacent tissue, such as pericardium. They are characterized pathologically by myocardial wall infiltration, tumor necrosis, and so on. Nevertheless, the conclusive diagnosis of malignant paraganglioma cannot be established without definitive evidence of infiltrated adjacent organs or lymph node metastasis [5].

Cardiac paraganglioma is not sensitive to chemotherapy and radiotherapy; the only effective treatment is surgical excision. Once the diagnosis is established, the tumor should be resected completely as soon as possible [5, 6]. Because most of these tumors are locally invasive, complete resection is very difficult. Only a few tumors with clear boundaries can easily be completely removed [7]. To completely resect the tumor, part of the wall of the heart usually has to be excised. Even β- or β-blockers have been used preoperatively or intraoperatively. Blood pressure would rise because of extensive catecholamine secretions when the tumor is resecting during operation. Therefore, Orringer and colleagues [8] recommended that resection should be performed after cardiopulmonary bypass is established and the aorta has been clamped. Establishing CPB results in a beneficial response to cardiac accidents during the surgical procedure, such as rupture of the heart or coronary injury. In our patient, we separated the tumor using CPB and resected the tumor completely.

Cardiac paraganglioma is generally slow growing, and the prognosis is good, even if there is residual tumor after operation. Additionally, it is suggested that paraganglioma be given attention in the differential diagnosis of all cardiac masses.

References