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Case Report

Functional Intracardiac Paraganglioma

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A 39-year-old man presented to the department of emergency medicine in Seoul National University Hospital complaining of chest pain, heart palpitation, and headache. Upon arrival, a computed tomography scan showed a 7.0 cm×6.2 cm lesion with the typical features of a paraganglioma. The patient was treated with an alpha-blocker and a beta-blocker prior to surgical intervention. We removed the tumor successfully, and histopathologic findings indicated that the tumor was indeed a paraganglioma. Since intracardiac paraganglioma is a rare disease, we present this case together with a literature review.

Keywords: Heart neoplasms, Paraganglioma, Hypertension

Case report

Paragangliomas are rare tumors originating from chromaffin cells of the sympathetic ganglia. Intracardiac paragangliomas are extremely rare, and the paucity of research on these tumors makes them challenging to treat [1]. Some paragangliomas are metabolically functional masses and may present with symptoms [2]. In this case report, we discuss a case of intracardiac paraganglioma that was managed by surgical intervention.

A 39-year-old man presented to the department of emergency medicine in Seoul National University Hospital complaining of chest pain, heart palpitation, and headache. Upon arrival, his blood pressure was 142/95 mm Hg, and his pulse rate was 106 with sinus rhythm. He had previously been prescribed and was taking medications for hypertension and diabetes mellitus. Echocardiography was ordered, and showed a large mass measuring 5.1 cm×4.9 cm in the left atrium (Fig. 1). The mass displayed heterogeneous echogenicity, with a clearly defined border. A computed tomography (CT) scan showed that the lesion was 7.0 cm×6.2 cm, and it had the typical appearance of a paraganglioma. Elevated serum normetanephrine, urinary norepinephrine, and urinary normetanephrine levels were observed, indicating that the tumor was a functional cardiac paraganglioma. Coronary catheterization revealed a tumor-feeding vessel originating from the left main coronary artery, not from the left anterior descending artery and left circumflex artery. The patient was referred to cardiac surgery, and alpha-blocker and beta-blocker medications were started as preoperative management.

During cardiopulmonary bypass, we approached the left atrium posterior wall via left atriotomy, and performed extended septotomy and aortotomy. The entire mass, measuring 7.5 cm×6.0 cm×3.5 cm, was resected en bloc along with the left atrial posterior wall (Fig. 2). After the mass was resected, the empty left atrial posterior wall was reconstructed using bovine pericardium. Cardiopulmonary bypass weaning was unremarkable. After the operation, a histopathologic examination confirmed the tumor to be a paraganglioma (Fig. 3).

The patient was discharged on postoperative day 13, and no longer required medication for hypertension. Postoperative echocardiography showed normal cardiac function and a 2-year follow-up CT examination showed no recurrence. At his regular follow-up, the patient still did not require medication for hypertension.

The patient provided written informed consent for the publication of his clinical details and images.

Discussion

Paragangliomas are rare neuroendocrine tumors closely related to pheochromocytomas, which originate in the ad-

renal glands. The incidence of paragangliomas is between 1.5 and 9 cases per million people. Approximately 2% of paragangliomas occur in the thorax [3]. Most thoracic paragangliomas exist in the posterior mediastinum; thus, cardiac paragangliomas are very rare. Cardiac paragangliomas can originate from the branchiomeric or visceral autonomic paraganglia, resulting in tumors in the aortic body and left atrium [3,4]. Although most paragangliomas occur sporadically, recent evidence suggests that up to a third may be due to heritable factors [1].

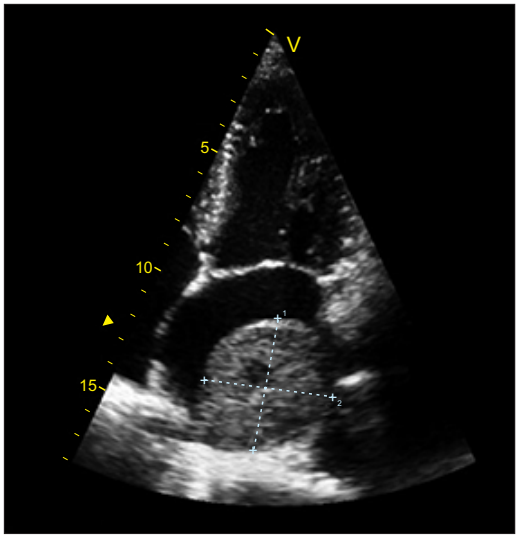

Fig. 1. Preoperative echocardiography showing a left atrial mass. Written informed consent for publication of this image was obtained from the patient.

Because paragangliomas secrete catecholamines, most patients with paragangliomas present with hypertension, palpitations, or headache. Therefore, 24-hour urinary vanillylmandelic acid and metanephrine testing are standard, and serum catecholamine levels are also a useful diagnostic tool. In many cases, complete resection is the first treatment option. Proper preoperative preparations are vital to ensure a successful resection. To avoid a hypertensive crisis, it is important to administer an alpha-blocker with phenoxybenzamine and prazosin. The prevention of hypovolemia is also important. A beta-blocker for residual tachycardia may be added only after an adequate alpha-blocker has been implemented to prevent unopposed alpha-adrenergic activity.

In cases of cardiac paraganglioma, complete surgical resection is challenging [5]. In this case, we resected almost the entire left atrial posterior wall and performed reconstruction with bovine pericardium. However, in some cas-

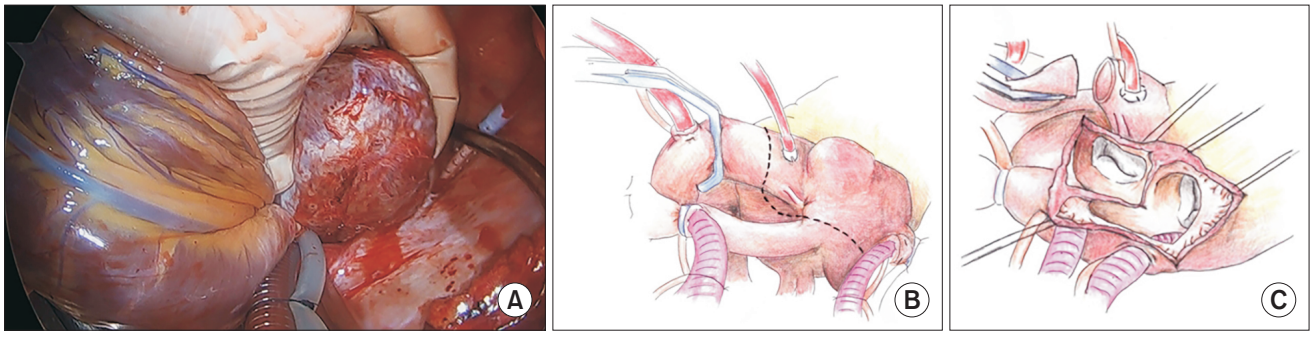

Fig. 2. Operative findings. (A) A mass measuring 7.5 cm×6.0 cm×3.5 cm on the left atrial posterior wall. (B) Diagram showing the surgical resection. (C) The aorta was divided to obtain better access to the left atrium. Written informed consent for publication of this image was obtained from the patient.

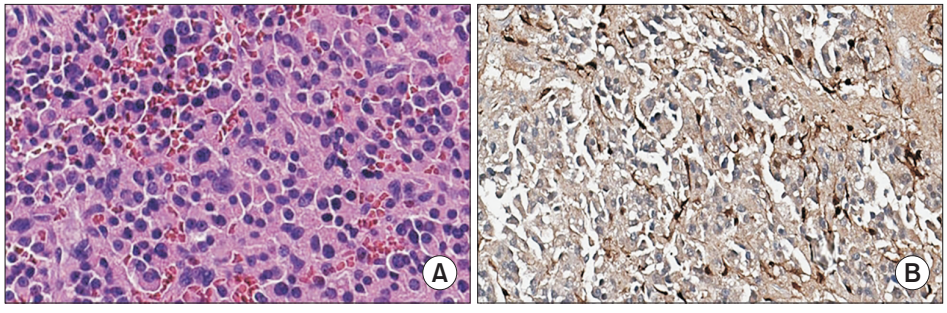

Fig. 3. Microscopic slides showing classic histoarchitecture with prominent uniform cell nests, known as the Zellballen pattern. (A) Hematoxylin and eosin staining (×400). (B) S100 staining (×400).

es, if the tumor is located deep in the left atrial cavity, auto-transplantation is a good option. Thus, preoperative CT and/or magnetic resonance imaging is important.

In conclusion, cardiac paragangliomas are rare and their surgical treatment is challenging. Adequate preoperative imaging and appropriate preoperative medication are essential. Complete surgical resection is currently the standard treatment.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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