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Case Report

A case with primary cardiac paraganglioma: imaging findings

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ABSTRACT

A 67-yeary-old middle-aged woman admitted to the hospital with chief complaints of intermittent palpitation, fatigue for more than 3 months, and bilateral lower extremity edema about 2 months. A solid mass was discovered in the right atrium by echocardiographic examination, and computerized tomography (CT) guided needle biopsy of the mass was performed and revealed a neurogenic tumor, which was a paraganglioma. She underwent surgical excision of the tumor and had uneventful recovery at a month post-operation.

Case summary

Three months prior to admission, a 67-year-old woman developed palpitation, fatigue and tachycardia without obvious predisposing causes, and exhibited bilateral lower extremity edema 2 months ago. Except a 4-year history of diabetes, she had a history of hypertension for more than 5 years with the highest blood pressure of 190/130 mm Hg, and no other specific or medical systemic diseases. A solid mass at the entrance of the inferior vena cava into the right atrium was found in the cardiac ultrasonography. The enhanced CT, Cardiac Magnetic Resonance (CMR), and positron Emission Tomography-Computed Tomography (PET-CT) all indicated the possibility of malignancy. Subsequently, CT-guided puncture biopsy was implemented for diagnosing the nature of the cardiac mass. The pathologic analysis showed short spindle-shaped and polygonal cell nests with rich cytoplasm and the possibility of epithelial tumors was considered. Immunohistochemistry showed positive of SYN, CgA and S-100, and revealed neuroendocrine tumors, which was consistent with paraganglioma (hematoxylin and eosin staining, magnification, × 200).

Following relevant pre-operative investigations completed and within normal limits, the patient was carried out surgical therapy and taken for cardiopulmonary bypass (CPB) that established with aortic and right atrial cannulation. During the surgery, we could see most of the mass were located in the left

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and right atria, and part of which invaded into the atrium wall. The size of the mass was measured about 8 × 6 × 4 cm and appeared with smooth surface, tough, yellowish and tan in color. With complete removement of the tumor along the margin of which, the bovine pericardium patch was taken to reconstruct the left and right atria. Postoperative histology and immunohistochemistry examination specimen of the patient was sent for analysis and confirmed of paraganglioma, which was corresponded to the diagnosis of CT-guided puncture biopsy.

Imaging findings and diagnosis

In this case, the imaging findings of the cardiac paraganglioma were as follows (Fig. 1). Echocardiography identified a solid mass in the right atrium without distinct features, and CT showed a mass of soft tissue density shadow in the right atrium with obvious enhancement except the center. A patchy

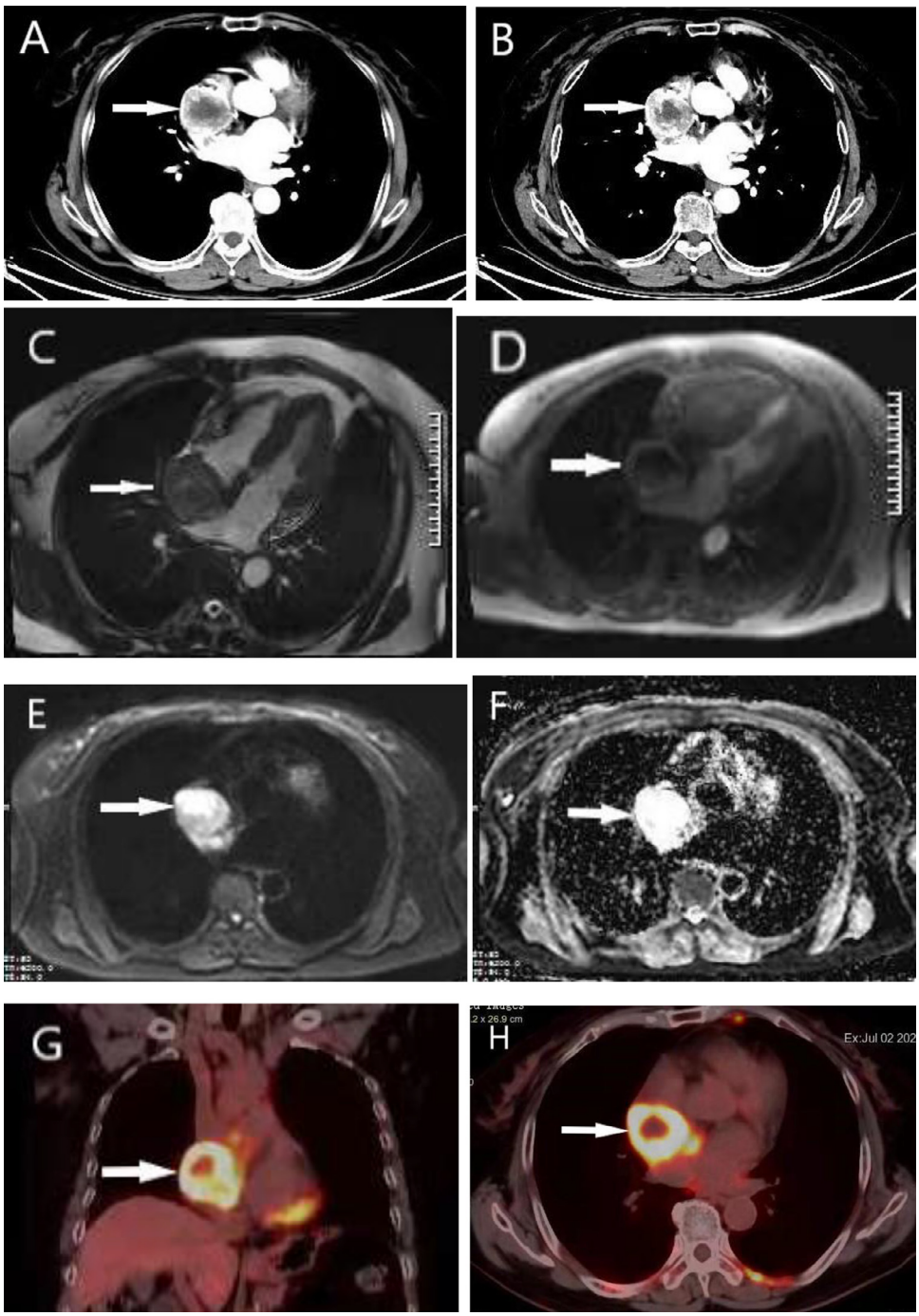

Fig. 1 – (A-B) Contrast-enhanced CT imaging. (C-F) CMR images. (G-I) PET-CT images. (J) Echocardiographic image. (K-L) Pathologic images. The arrows indicate lesions.

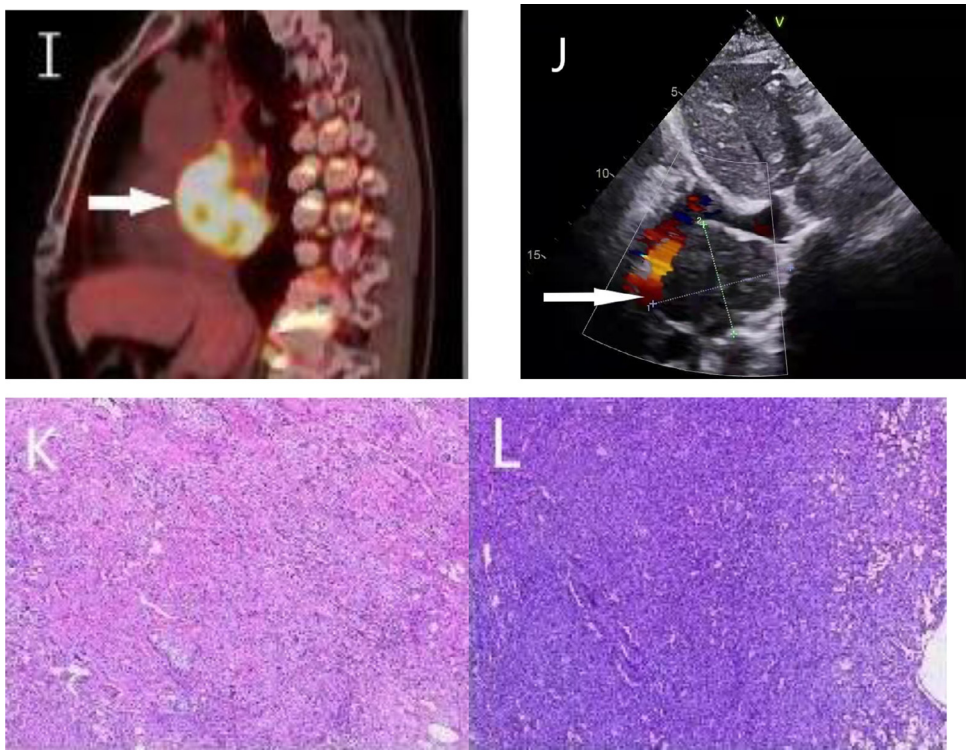

Fig. 1 – Continued

mixed long T2 signal shadow can be seen in the right atrium in the CMR, whose inferior border reached the entrance of the inferior vena cava. After intravenous gadolinium contrast administration, the right atrium lesions presented mixed hyperintense in the first pass perfusion, and heterogeneous signals in the delayed enhancement. ^{18}F-Fluorodopamine (^{18}F-FDA) PET-CT revealed a soft tissue mass in the pericardium with increased metabolic uptake and the maximum standardized uptake value (SUV max) was 15.2.

Discussion

Paraganglioma, or extra-adrenal pheochromocytoma, is a type of neuroendocrine tumor that originates from nerve spinal cells [1]. It usually occurs in the posterior peritoneum beside the spine showing similar incidence between the sexes among the ages of 40-60 years, rarely in internal organs, and even less involving in the heart. The tumor can be divided into 2 types. One is derived from pheochromocytoma and has the function of secreting catecholamine transmitters, causing high metabolic clinical manifestations similar to pheochromocytoma such as hyperglycemia, hypertension, headache, hyperhidrosis, etc [2]. The other is originated from chemoreceptor cells and does not secrete catecholamine transmitters. There are no presenting clinical signs and symptoms in the early stage, but as the enlargement of the tumor volume and compression of surrounding tissue, the patient can develop corresponding clinical symptoms, and be diagnosed during physical examination.

In this case, the patient with symptoms of palpitation, fatigue, rapid heart rate and histories of hypertension and diabetes presented to the hospital and the disease was discovered by echocardiography. Specific ultrasonographic finding are lacking in paraganglioma. The tumor is generally large and accompanied by necrosis, hemorrhage, and cystic degeneration, manifesting as solid mixed echo mass with clear boundaries in the 2-dimensional ultrasound, and detectable blood flow signals inside it in the color Doppler. The CT plain scan of the tumor usually manifests as single or multiple soft tissue density nodules or masses with clear boundaries and internal scattered plaque-like calcification inside, which are round, round-like, fusiform or irregular. Contrast-enhanced CT scan images can show obvious enhancement and rich blood supply in the smaller tumors with homogeneous density, but heterogeneous enhancement was showed in the larger ones which prone to hemorrhage, necrosis and cystic degeneration [3]. On magnetic resonance imaging (MRI), the lesion shows equisignal or low signal on T1WI, equi-signal or high signal on T2WI, and high signal on DWI (diffusion-weighted imaging) [4]. Part of the tumor parenchyma is enhanced significantly following the intravenous administration of gadolinium on the contrast-enhanced T1WI. Pathology is the gold standard for diagnosis of paraganglioma, but it is the difficult to judge the benign and malignant by histopathology alone, and the only reliable malignant standard is the detection of distant metastasis.

Finally, surgical resection was performed, and the histopathological diagnosis was cardiac paraganglioma. Surgery remains the main treatment of paraganglioma, meanwhile, the effects of radiotherapy, and chemotherapy

have also been confirmed. Long-term follow-up of the patient is recommended with regard to the recurrence rates of up to 25% as reported, which should include CT scan regularly.

Conclusion

Paraganglioma is rare, and that occurs in the heart is even unusual. The imaging manifestations of cardiac paraganglioma are specific, and the diagnosis of the disease can be considered in combination with clinical manifestations.

Author's contributions

Shaohua Hua designed the case report. Mengmeng Liu and Pengge Li drafting the paper. Yonggao Zhang and Xinxin Sun provided the images.

Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Data availability

Publicly available datasets were used in this study.

Fund program

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Patient consent

The written and informed consent for publication of the case was obtained from the patient .

Declaration of Competing Interest

The authors declare that they have no conflicts of interests.

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