Case Report

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대한영상의학회지

Cardiac Papillary Fibroelastoma in Left Ventricular Trabeculation as a Potential Cause of Cerebral Infarction: A Case Report

뇌경색의 원인으로 생각되는 좌심실 섬유주에 생긴 심장 섬유탄력종: 증례 보고

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Cardiac papillary fibroelastoma (CPF) is the second or third most common primary cardiac tumor. Although histologically benign, it can cause serious symptoms depending on its location of occurrence, size, and motility. Herein, we report CPF in the left ventricular trabeculation as a potential cause of cerebral infarction.

Index terms Cardiac Papillary Fibroelastoma; Left Ventricle; Cerebral Infarction; Echocardiography; Contrast Echocardiography; Computed Tomography, X-Ray

INTRODUCTION

Primary cardiac tumors are rare, but more frequently diagnosed with the development of cardiac imaging techniques. Cardiac papillary fibroelastoma (CPF) is the second or third most common primary cardiac tumor (1, 2). It is histologically benign, but can cause serious symptoms depending on the location, size, and motility (1). It mainly affects the valve, but can occur anywhere in the heart. We report CPF in left ventricular trabeculation as potential cause of cerebral infarction.

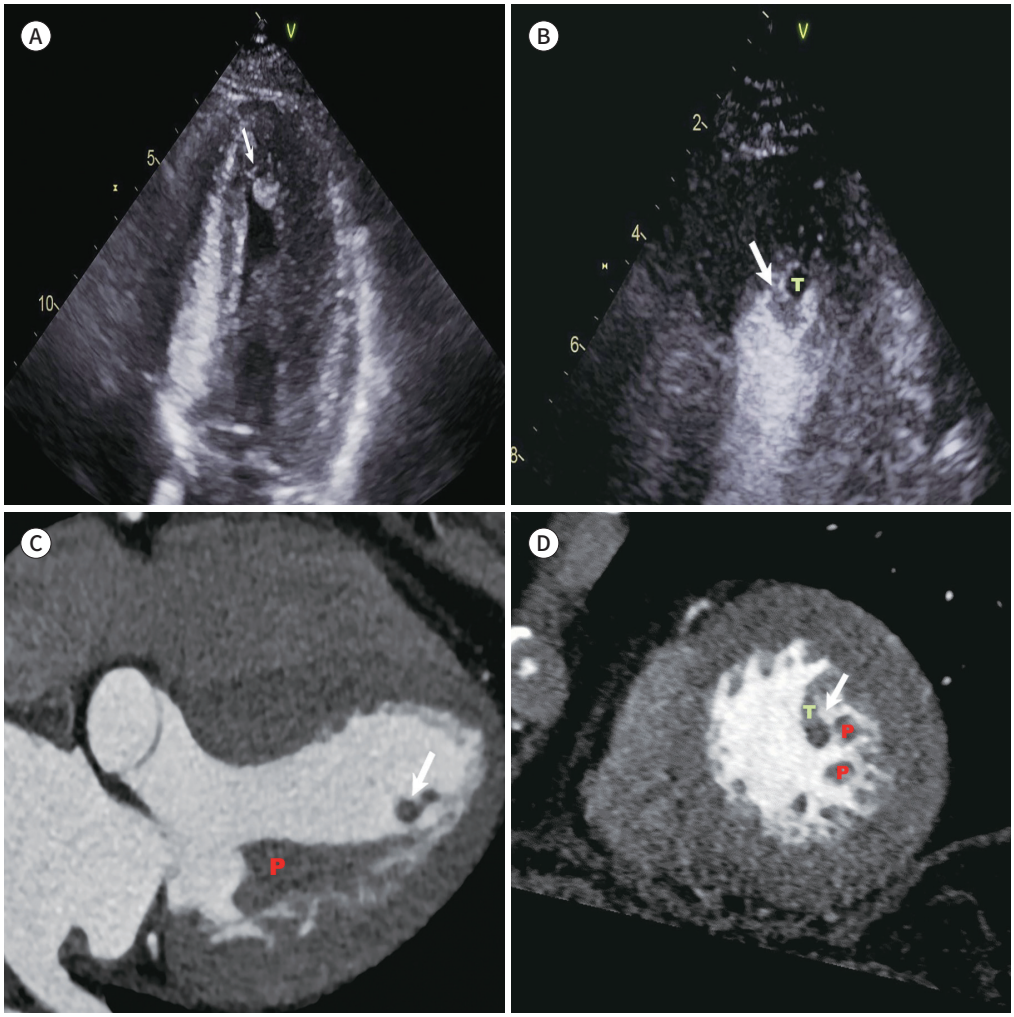
CASE REPORT

A 62-year-old male with heavy alcoholics came to the hospital with intermittent dizzi-

ness. Blood pressure on presentation was 170/100 mm Hg. The physical examination to check the vestibular nervous system was normal. So, we thought about the possibility of brain abnormalities.

He underwent brain computed tomography (CT) and magnetic resonance image (MRI), which revealed tiny focal infarctions in right parietal lobe and left cerebellum. With the possibility of embolic infarction, transthoracic echocardiography (TTE) was performed with VIVID E9 (GE Healthcare Co., Milwaukee, WI, USA). It showed that about 1.2 cm × 0.6 cm-sized, round, mobile, heterogeneously echogenic mass attached to left ventricular trabeculation (Fig. 1A). On TTE with contrast (SonoVue; Bracco, Milan, Italy), the mass also was detected (Fig. 1B). To examine the mass in detail, coronary CT angiogram with 128 channel multidetector CT (Somatom Definition AS+; Siemens, Erlangen, Germany) was obtained. On

Fig. 1. A 62-year-old male with cardiac papillary fibroelastoma in left ventricle.
A, B. Transthoracic echocardiography shows a hypermobile mass (arrows) in the left ventricle (A). After contrast administration, it shows similar echogenicity to the myocardium, with more apparent changes in shape (B).
C, D. Computed tomography images reveal a small, round mass (arrows) attached to the left ventricular trabeculation on 4-chamber (C) and short-axis views (D).
P = anterior papillary muscle, T = trabeculation



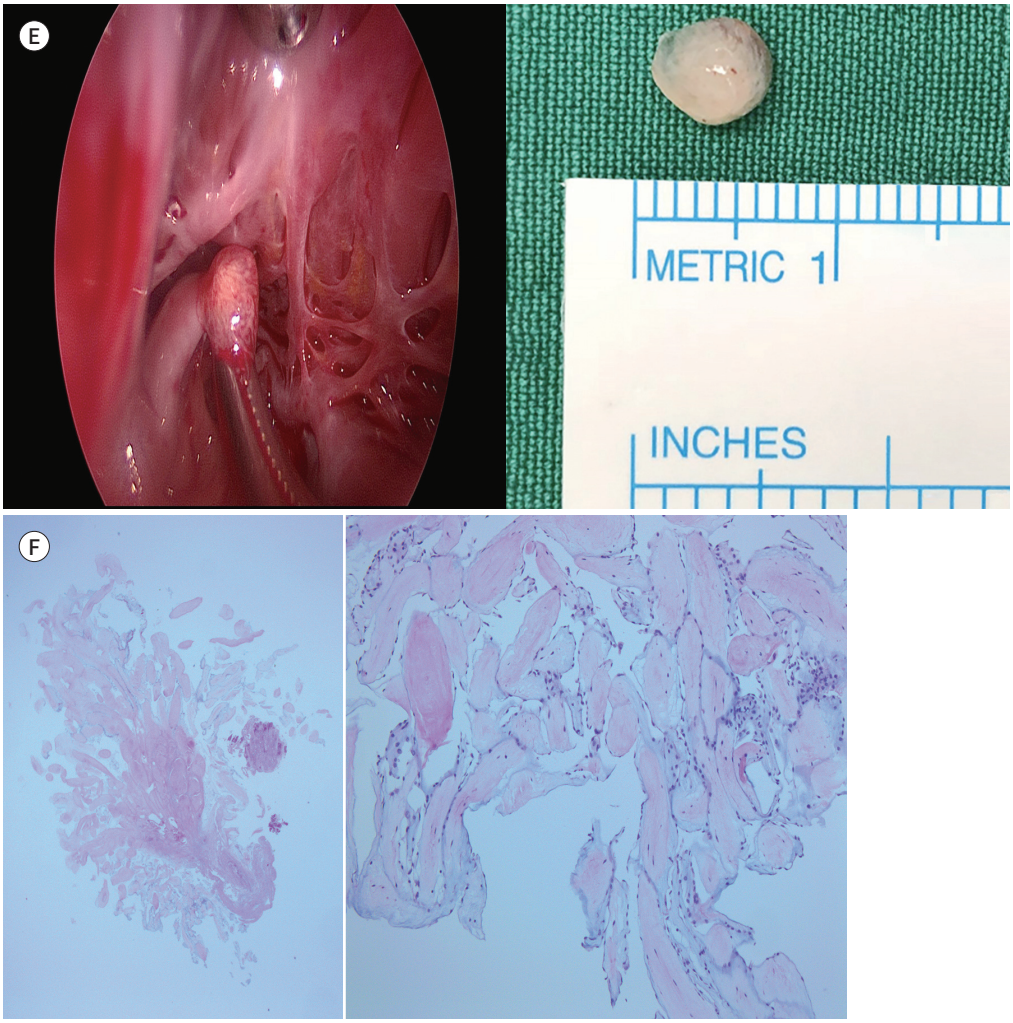
non-contrast CT, mass was indistinguishable from unopacified blood. It was about 1.0 cm × 0.6 cm-sized, small round mass on contrast CT and had intermediate density similar to that of myocardium, measured at 98 Hounsfield unit (HU) when region of interest was 5 mm^2 (Fig. 1C, D). Electrocardiogram was normal sinus rhythm.

Considered as a potential cause of cerebral infarction, the patient was referred to the Department of Thoracic Surgery for surgical removal. The resected mass was gelatinous (Fig. 1E). Intraoperative transesophageal echocardiography was performed, which demonstrated no residual mass. Histologic examination confirmed CPF (Fig. 1F).

DISCUSSION

CPF is the second or third most common primary cardiac tumor. In analysis of 725 case (1),

Fig. 1. A 62-year-old male with cardiac papillary fibroelastoma in left ventricle.
E. Intraoperative photographs show the mass firmly attached to the left ventricular trabeculation and covered with gelatinous materials.
F. Hematoxylin and eosin staining demonstrates numerous arborizing fronds arising from the central stalk (left; × 25) and avascular fronds, covered with bland endothelium (right; × 100).



CPF can occur at any age, including neonate. It is more common between the 4th and 8th decades and mean age that it is detected is about 60 years. It occurs more often in male. It is mainly acquired, but exact etiology is unknown (1).

The tumors vary in size from 2 mm to 70 mm (1). In large analysis with available data, 84% of the cases were located valvular surface. It is located predominantly on aortic valve, followed by mitral valve, tricuspid valve and pulmonary valve. Left ventricle is the most common non-valvular location of the tumor (1).

Grossly, CPF resembles sea anemone when it is put in normal saline solution. It is covered by endothelium that surrounds connective tissue matrix with variable proportion of collagen, muscle cells, and elastic fibers (1, 3).

Most of patients with CPF is asymptomatic (1). Clinical manifestation of CPF vary according to size, mobility, and location of tumors and the most common clinical presentation is transient ischemic attack and stroke (1). Other clinical manifestations are as follows: angina, myocardial infarction, sudden death, heart failure, pulmonary embolism, blindness, mesenteric ischemia, peripheral emboli and renal infarction. It is not clear whether the cause of embolus is the tumor itself or the thrombus caused by the tumor (1).

The standard tool for diagnosis is echocardiography. Typical findings on echocardiography are that they can be single or multiple, small (< 2 cm), varied in shape (oval, round, irregular), well-circumscribed, and mobile. Nearly half of the tumor have stalk and they are often associated with cardiac valvular disease (3). In this study, stalk was confirmed by histological examination even though it was not delineated on CT and TTE. On contrast echocardiography, they can appear vascularized because a contrast agent can disperse between the frond-like appendages, although CPFs are avascular tumor. This can lead to misunderstanding it as a vascular tumor (4).

It has been reported to show 52–156 HU on the contrast CT (5, 6), but the characteristic features have not been established. In this study, it was estimated at 98 HU. When cardiac tumors are found clinically, MRI is mainly performed. It is known that there are no literatures on the standard CT protocol for cardiac tumors, but several CT phases are helpful in identifying cardiac tumor characteristics. On non-contrast image, it is possible to know the composition of mass according to the difference in attenuation (fat, calcification, and myxoid) (7). On enhanced and delayed images, it is helpful to distinguish thrombus and mass according to the presence of enhancement, and in the case of delayed images, it may be helpful to distinguish thrombus and blood stasis (7, 8).

A few literatures about MR features of CPF have been reported (9, 10). CPF appears as isointensity on T1-weighted images and hyperintensity on T2-weighted images compared to myocardium. After contrast administration and delayed contrast images, it appears as hyperintense mass (9). Turbulent flow can be seen in the cine MR around the mass (9).

Differential diagnosis includes other cardiac tumor, thrombus, metastasis and vegetation (especially when it occurs in the valve). Most of myxomas are found in the left atrium, typically, in the interatrial septum in the region of the fossa ovalis; rare cases are found in the ventricles. Tumor attenuation is lower than that of the unopacified blood (2) Thrombi can be differentiated by laminated appearance, irregular or lobulated border, microcavitations, and absence of pedicle on echocardiography (1). It can also be distinguished from tumor by ab-

sence of late MR enhancement. Metastasis typically manifests as multiple masses or nodules, but can be seen as diffuse infiltration. The pericardium, epicardium, and myocardium are most often affected. Endocardial involvement is less common and most often affects the right heart chambers (10). The vegetations can be similar to CPF due to its valvular location and mobility. However, these vegetations are usually associated with clinical signs of endocarditis and valvular destruction, and may resolve or change in appearance over time with treatment (1).

For symptomatic CPF, surgical resection is the treatment of choice. If it is motility or grows in size at follow up, surgical treatment is recommended, even if there are no symptoms. Surgical treatment is curative and postoperative recurrence has not been reported (1). For patients with CPF who do not have symptoms and mobility, close follow up or anticoagulation is recommended, but there is no exact guideline based on randomized controlled data (1).

In conclusion, we report CPF in left ventricular trabeculation as potential cause of cerebral infarction.

Author Contributions

Conceptualization, K.D.S.; investigation, I.J.Y.; methodology, K.D.S.; project administration, K.D.S.; supervision, K.D.S.; visualization, K.D.S.; writing—original draft, all authors; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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뇌경색의 원인으로 생각되는 좌심실 섬유주에 생긴 심장 섬유탄력종: 증례 보고

임준용 · 김동수*

심장섬유탄력종(cardiac papillary fibroelastoma; 이하 CPF)은 두 번째 혹은 세 번째로 흔한 원발성 심장종양이다. 그것은 조직학적으로 양성이지만, 생긴 위치, 크기, 운동성에 따라 심각한 증상을 초래할 수 있다. 우리는 뇌경색을 일으킨 원인으로 생각되는 좌심실 섬유주에 생긴 CPF를 보고하고자 한다.

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