Right atrial papillary fibroelastoma arising from the Chiari network detected by echocardiography: a case report.

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Abstract

Right atrial papillary fibroelastomas (PFE) originated from the Chiari network (CN) are exceedingly rare. Only 2 cases have been reported in the worldwide literature. We present a case of PFE arising from the right atrial CN detected by transesophageal echocardiography (TEE) with a previous vertebral aneurysm history and underwent a complete surgical removal. In this report, we describe our clinical observations to improve our understanding of the tumor and provide evidence for its treatment and prognosis.

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Running head: Cardiac tumor detected by echocardiography

Abstract:

Right atrial papillary fibroelastomas (PFE) originated from the Chiari network (CN) are exceedingly rare. Only 2 cases have been reported in the worldwide literature. We present a case of PFE arising from the right atrial CN detected by transesophageal echocardiography (TEE) with a previous vertebral aneurysm history and underwent a complete surgical removal. In this report, we describe our clinical observations to improve our understanding of the tumor and provide evidence for its treatment and prognosis.

Key words: papillary fibroelastoma, cardiac tumor, Chiari network

Background

Papillary fibroelastoma (PFE) is the most common primary cardiac tumor surpassing myxoma [1]. PFEs typically occurred on valvular surfaces and the vast majority is seen on the left side[2]. Right atrial nonvalvular PFEs were delineated in 14 cases and only two cases were described as having located on the Chiari network (CN) [3-5]. Although PFEs were generally considered benign, PFE has a tendency leading to syncope, chest pain, stroke, myocardial infarction, pulmonary embolism [2]. Here, we report a rare case of PFE arising from the right atrial CN detected by transesophageal echocardiography (TEE) with a previous 14-day headache history.

Case presention

A 54-year-old man with a previous vertebral aneurysm history was admitted to our hospital presented with a 14-day headache. The patient denied nausea, shortness of breath, palpitations, and vomiting. Physical examination was unremarkable and the results of all routine laboratory tests were within normal limits. However, routine bedside echocardiography suggested an unidentified mass mimicking myxoma in the right atrium.

Among further cardiac examinations, an electrocardiogram showed a normal sinus rhythm. Transthoracic echocardiography revealed a mobile, sessile mass in the right atrium without obstructing the orifice of tricuspid valve. The subsequent transesophageal echocardiography confirmed the presence of a 1.56cm×1.24cm mobile, sessile, irregular mass arising from the CN (Fig.1) and showed no evidence of patent foramen ovale.

Surgical resection was considered for avoiding potential dislodgement and confirming diagnosis. The patient underwent a right atriotomy aided by cardiopulmonary bypass, the base of the stalk and the full thickness of the endocardium involved was excised. Grossly, the tumor was a cauliflower-shaped, red-colored mass attached to the fibrous reticulum between inferior vena cava and coronary sinus (Fig.2). Postoperative beside echocardiography indicated complete excision of the tumor.

Histopathologic findings showed papillary fronds with central hypocellular, avascular, hyalinized stroma covered by a single layer of endocardial cells (Fig.3). Microscopic features were compatible with PFE and the final diagnosis was a papillary fibroelastoma located on the CN of the right atrium. The patient made an uneventful postsurgical recovery and was discharged home. Three months later, the patient came back to our hospital to accept the operation for clipping vertebral aneurysm as clinicians recommended.

Discussion

Cardiac PFE is a benign tumor that predominates in adults, with a peak incidence in the 7^{th} decade of life. PFE usually develop on the cardiac valves and only about 23% originate from nonvalvular surfaces. More than 95% of PFEs arise in the left heart[2]. The CN is a fibrous reticulum resulting from incomplete regression of the right valve of sinus venous and the septum spurium, connecting different parts of the right atrium[6]. PFE attached to the CN is extremely rare with only two cases reported up to now. One case presented a PFE on the CN found incidentally in an autopsy in 1992 and another case of infective endocarditis on the CN has been reported in 2008 [4,5].

In this case, we occasionally found a PFE located on the CN by echocardiography with a 14-day headache history. With the increased use of echocardiography and enhanced awareness of PFE, PFEs are diagnosed

more often in recent years and become the most common primary cardiac tumor in adults [1]. Echocardiography is a non-invasive and convenient diagnostic technique and should be the first choice for detecting suspected PFEs. TEE is extremely useful to preoperative evaluation and decision-making of treatment, providing specific delineation of the size, shape, location, mobility, and the presence of stalk of neoplasms with high-resolution imaging [7]. In addition, TEE is capable of guiding the surgery intraoperatively and evaluating the cardiac function postoperatively.

PFEs are associated with different embolic symptoms according to their location in the heart. Tumors located in the left heart are usually relevant to obstructive and embolic symptoms, and the most common sites of embolization are cerebral, coronary, and systemic circulation. Right-sided tumors mostly remain asymptomatic. However, the cases of embolization to the pulmonary vessels leading to subsequent pulmonary hypertension have been reported. The exact etiology behind the symptoms remains unclear, whereas some researchers view that the embolization may originate from the tumor fragments or a thrombus formed on the surface of the tumor. In Vandergoten's report, congenital patent foramen ovale with right-to-left shunts may attribute to paradoxical embolisms from the right-sided PFEs [2]. Consequently, the right atrial mass in our case seems to be irrelevant to the headache symptom without any evidence of congenital anomalies.

Differential diagnosis of PFE encompasses myxoma, thrombi, vegetations, strands, and giant Lambl's excrescences [8]. Clinical data, blood cultures, laboratory tests together with echocardiographic features may be useful to differentiate these lesions. But the ultimate diagnosis depends on the characteristic histopathological features.

In our report, the patient was treated surgically and made an uneventful postsurgical recovery. One study has shown that the tumor mobility is the independent predictor of death [2]. Surgery is curative with an excellent long-term prognosis and a careful follow-up echocardiogram is recommended for potential recurrence. If the patient is not a surgical candidate, chronic anticoagulation with antiplatelet agents or aspirin is usually suggested [2,8].

References

1. Tamin SS, Maleszewski JJ, Scott CG, et al. Prognostic and Bioepidemiologic Implications of Papillary Fibroelastomas. J Am Coll Cardiol 2015;65:2420-9.

2. Gowda RM, Khan IA, Nair CK, Mehta NJ, Vasavada BC, Sacchi TJ. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. Am Heart J 2003; 146:404–11.

3. Hakemi EU, Bero J, Sekosan M, Ansari A. Nonvalvular right atrial papillary fibroelastoma. J Thorac Cardiovasc Surg 2013;145:e71-3.

4. Latif F, Peyton M, Laszik Z, Sivaram CA. Infective endocarditis of a papillary fibroelastoma on Chiari network of right atrium: a case report. J Am Soc Echocardiogr 2008;21:188.e3-4.

5. Wasdahl DA, Wasdahl WA, Edwards WD. Fibroelastic papilloma arising in a Chiari network. Clin Cardiol 1992;15:45-7.

6. Loukas M, Sullivan A, Tubbs RS, Weinhaus AJ, Derderian T, Hanna M. Chiari's network: review of the literature. Surg Radiol Anat 2010;32:895-901.

7. Klarich KW, Enriquez-Sarano M, Gura GM, Edwards WD, Tajik AJ, Seward JB. Papillary fibroelastoma: echocardiographic characteristics for diagnosis and pathologic correlation. J Am Coll Cardiol 1997;30:784–90.

8. Sun JP, Asher CR, Yang XS, et al. Clinical and echocardiographic characteristics of papillary fibroelastomas: a retrospective and prospective study in 162 patients. Circulation 2001;103:2687-93.

Figure legends

Fig.1.Transesophageal echocardiography illustrating a 1.56cm×1.24cm mobile, sessile, irregular mass (solid arrow) attached to the Chiari network. IVC = inferior vena cava,RA = right atrium, RV = right ventricle.

Fig.2. Intraoperative findings: tumor adhering to the Chiari network between inferior vena cava and coronary sinus. CN = Chiari network, CS = coronary sinus, IVC = inferior vena cava, PFE = papillary fibroelastomas.

Fig.3. High-power (100) magnification of papillary fronds: consisting of a central avascular core rich in elastic fibers and lined by superficial endothelial cells.