Clinical Medicine Insights: Cardiology

CASE REPORT

Right Ventricular Variants and Pulmonary Embolism—Association or Coincidence?

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Abstract: It has been stated that the interior of the right ventricle is as unique to each individual as one's fingerprint. This statement is backed by numerous publications which demonstrates considerable variation in the number, shape and configuration of papillary muscles inside the normal right ventricle.

It has also been shown that these variants may be the cause of cardiac rhythm disorders.

In this case report another potential complication of such right ventricular papillary muscle variants is proposed—these muscles may be the source of pulmonary emboli.

The pathogenesis may be that of local stasis around these aberrant muscular structures and/or emboli may form inside the right ventricle as a result of cardiac rhythm disorders, induced by these muscles.

It is proposed that in future the role of the right ventricle as the source of pulmonary emboli will become more apparent and an important part of the diagnostic work up in cases of idiopathic pulmonary embolism.

Keywords: Pulmonary embolism, right ventricle, papillary muscle, variants

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Introduction

According to Victor and Nayak[1] the interior of the right ventricle is as unique to each individual as one`s fingerprint. Numerous publications[2–6] has shown that inside the normal right ventricle considerable variation exists in the number of papillary muscles, the number of heads of individual papillary muscles, the number of papillae on the heads of papillary muscles and also the shape of individual papillary muscles.

Wenink[2] examined 100 normal hearts and concluded that the medial papillary muscle of the right ventricle, also known as the papillary muscle of the conus, the muscle of Luschka and the muscle of Lancisi, displays such wide morphological variations that the value of the muscle as an anatomical landmark in the right ventricle is very restricted. Due to this wide morphological variations he proposed the name the medial papillary complex.

Restivo et al[3] studied fetal hearts—81 subjects, ranging in age from 20 weeks of gestation to 13 months—and also found considerable variability in the papillary muscles of the right ventricle.

Nigri et al[4] studied the morphological characteristics of the right ventricular papillary muscles and their chordae tendineae in 79 normal human hearts—aged from 14 to 68 years— and found the following:

The anterior and posterior papillary muscles were present in all of the 79 hearts. The septal papillary muscle was absent in 21.5% of cases. The anterior papillary muscle had one head in 81% of cases and two heads in 19% of cases. A head of a papillary muscle was defined as a muscle that is directly attached to the ventricular wall. The average length of the anterior papillary muscle was 19.16 mm. The septal papillary muscle had one head in 41.7% of cases, two heads in 16.5% of cases, three heads in 12.7% of cases and four heads in 7.6% of cases and the average length of the septal papillary muscle was 5.59 mm. The posterior papillary muscle had one head in 25.4% of cases, two heads in 46.8% of cases, three heads in 21.5% of cases and four heads in 6.3% of cases. Its average length was 11.53 mm. The chordae tendineae also displayed enormous variation: From one to eleven originated from the anterior papillary muscle, from one to eight from the posterior papillary muscle and from one to five from the septal papillary muscle.

Skwarek et al[5] separates the conal papillary muscle and the papillary muscles of the posterior angle of the right ventricle from the classically described papillary muscles of the right ventricle, referred to in anatomical nomenclature—the anterior, posterior and septal papillary muscles. They found the conal papillary muscle—also described by Luschka in the seventeenth century—as the most constant of the septal papillary muscles. Furthermore, they found papillary muscles in the posterior angle of the right ventricle which could not be clearly classified as either septal or posterior muscles—thus, called the muscles of the posterior angle of the right ventricle.

Similar to Nigri et al,[4] Begum et al[6] studied fifty hearts—aged from 20 to 70 years—from the Bangladesh population and found the following:

The right ventricle had a single anterior papillary muscle in 92% of cases. The posterior papillary muscle was single in 28% of cases and double in 32%. The septal papillary muscle was single in 46% of cases and absent in 30% of cases. They found the anterior papillary muscles the longest and the septal papillary muscles the shortest.

Case report

A case report is presented where a large papillary muscle complex is present in the apical part of the lateral wall of the right ventricle in a 44 year old Caucasian woman. This aberrant papillary muscle complex is thought to be the source of recurrent pulmonary emboli.

A 44 year old Caucasian woman presented with a 3 month history of pleuritic chest pain. During and after all three her pregnancies she also experienced the same pleuritic chest pain for variable periods, ranging from one to four months.

She was taking levothyroxine for hypothyroidism, which had been diagnosed two years earlier. No other medical problems were present.

The clinical examination was normal. A 12-lead electrocardiogram and chest radiograph were also normal. A ventilation-perfusion (V/Q) scan demonstrated multiple pulmonary emboli in both lungs.

A comprehensive search did not reveal any known cause for these multiple pulmonary emboli: No deep venous thromboses of the lower and upper limbs or

pelvic veins were present, no malignant process were present and the following biochemical measurements were all normal: Antithrombin and proteins C and S levels were within normal limits, factor IX levels were within normal limits and factor V Leiden was negative, lipoprotein(a) and homocyteine levels were normal, anti-nuclear factor was absent, antiphospholipid antibodies were absent, and the prothrombin mutation PT G20210A was absent.

Echocardiography revealed the presence of a large, papillary muscle complex in the apical part of the lateral wall of the right ventricle (Fig. 1). Figures 2 and 3 demonstrate that this papillary muscle complex consists of a base with a single, tall head flanked by a smaller head on either side. In supplementary Figure 1, it is clear that this muscular complex is papillary muscle, as chordae tendineae extends from the papillary muscle complex.

Discussion

What might be the potential clinical complications of these various variants of papillary muscles in the right ventricle?

They may be the source of cardiac rhythm disorders.[7] Lazzari et al[7] examined the short-term behavior of extrasystoles, arising from the anterior papillary muscle of the right ventricle. 20 subjects were studied with Holter recordings and exercise data. According to the authors a right ventricular anterior papillary muscle extrasystole has a left bundle branch block morphology with a downward oriented QRS axis in the frontal plane, a slurred r wave in lead V1 and an R/S ratio less that 1 in lead V1. They concluded that this entity can be found in otherwise normal hearts with an uncomplicated outcome in the short-term.

Aktas et al[8] investigated papillary muscle variants around the tricuspid valve in cases of sudden death. 400 hearts were studied and a great variability in the number of papillary muscles in the right ventricle was found, ranging from two to nine. The authors also observed a striking incidence of conical and flat topped configurations of the posterior papillary muscle in these deaths. The authors concluded that these pap-

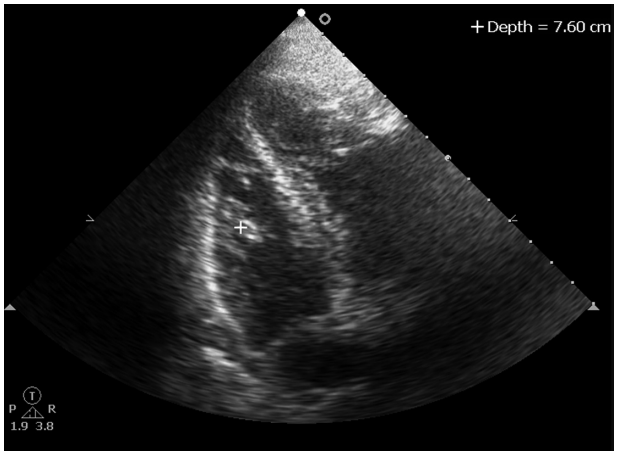

Figure 2. Papillary muscle complex.
Note: Another view of the aberrant papillary muscle complex.

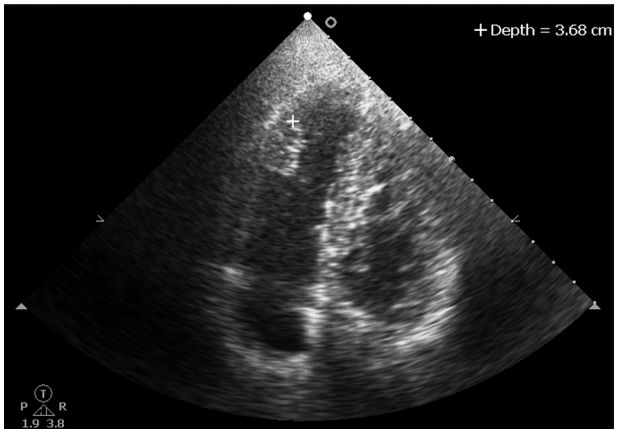

Figure 1. Aberrant papillary muscle complex in the lateral wall of the right ventricle.
Note: This is an apical, four chamber view demonstrating a large papillary muscle complex, with its origin from the lateral wall of the right ventricle.

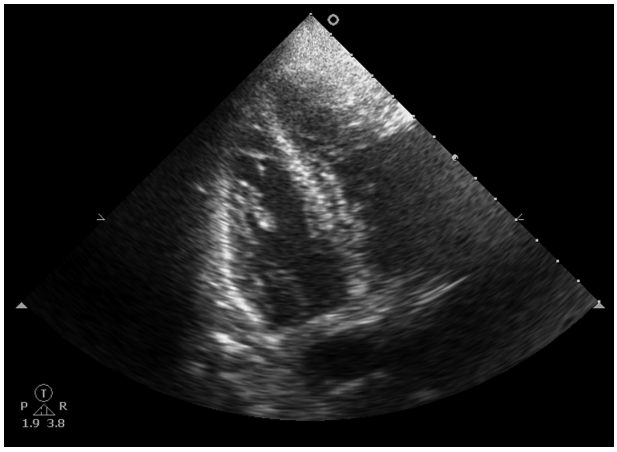

Figure 3. Three headed papillary muscle complex.
Note: This is an apical view which demonstrates that the papillary muscle complex consists of a base with a single, tall head flanked by a smaller head on either side.

illary muscle variants may be the cause of sudden cardiac death. However, there is a paucity of data on the existence and possible mechanisms of arrhythmia in patients with papillary muscle variants.

In this case report another potential complication of such papillary muscle variants in the right ventricle is proposed—these variants may be the source of pulmonary emboli.

The physiological basis of this may be due to stasis of blood around these structures with resultant emboli or these muscles may be the source of brief episodes of cardiac rhythm disorders in the right ventricle, as previously described,[7] with resultant episodes of stasis and consequent emboli.

The right side of the heart as the source of pulmonary emboli is not a new concept: Various primary and secondary cardiac tumors have been shown to be a potential source of pulmonary emboli.[9–12] The primary tumors include myxomas[9] and papillary fibroelastomas,[10] whereas testicular embryonal carcinoma[11] and squamous cervical carcinoma[12] are examples of metastatic right ventricular tumors with resultant pulmonary emboli.

Possible mimics of such a papillary muscle variant in the right ventricle include: valvular vegetations, a papillary fibroelastoma, thrombus and metastatic tumors. A valvular vegetation was not considered due to the fact that there is no continuity between the mass and the tricuspid valve. The unique echocardiographic features of papillary fibroelastomas include small size, attachment to the endocardium via a stalk or pedicle that is highly mobile, a refractive appearance and areas of echolucency within the tumor itself.[10] No evidence of any primary malignancy could be found and therefore the possibility of a cardiac metastasis was not considered. Endocardial metastases are extremely rare and this has been attributed to the strong kneading action of the heart, the metabolic peculiarities of the myocardium, the rapidity of coronary blood flow and the lymphatic connections that drain afferently from the heart.[13]

Lastly, Maron et al[14] have shown that there may be a "spillover" of the primary left ventricular hypertrophic process from the septum into adjacent segments of the right ventricular wall in patients with hypertrophic cardiomyopathy. However, in this case the muscular complex is seen in the lateral wall and no echocardiographic stigmata of hypertrophic cardiomyopathy were present in the left ventricle. However, it is possible that pulmonary embolism may arise from this right ventricular apical muscular complex by the same mechanism as that seen in patients with left ventricular apical hypertrophic cardiomyopathy.

It is hoped that this report will lead to more focus on the right ventricle as the possible source in cases of unexplained pulmonary emboli. It is proposed that in the future of clinical cardiology the right ventricle will become an important area of investigation in cases of unexplained pulmonary embolism.

Disclosure

This manuscript has been read and approved by the author. This paper is unique and is not under consideration by any other publication and has not been published elsewhere. The author and peer reviewers of this paper report no conflicts of interest. The author confirms that they have permission to reproduce any copyrighted material. Written consent was obtained from the patient for publication of this study.

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