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The Link between the Ovaries and the Heart, an Untold Mystery of Ovarian Carcinoid with Cardiac Involvement!

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Abstract

Carcinoid Heart Diseases (CHD) generally are considered a rare occurrence and CHD caused by Primary Ovarian Carcinoid Tumor (POCT) form a rare form of acquired valvular heart disease. CHD usually presents with right-sided valvular dysfunctional picture, which subsequently lead to right-sided heart failure. However, in rare instances the left sided-valvular involvement may occur. The cardiac involvement due to POCT is a unique entity compared to other forms of cardiac involvement as this develops without hepatic metastasis, which has clear clinical implications in terms of management principles and long-term outcomes.

Keywords: Carcinoid; Ovaries; Carcinoid heart diseases; Octreotide; Serotonin

Abbreviations

CVS: Cardiovascular; CHD: Carcinoid Heart Diseases; OC: Ovarian Carcinoid; POCT: Primary Ovarian Carcinoid Tumor

Introduction

Primary cardiac tumors are rare, however metastatic tumors to the heart should be sort considerably which mostly include lung and breast cancers, hematological malignancies including leukemia and lymphomas, and malignant melanoma, which are all regarded to form majority of secondary cardiac tumors. Cardiac carcinoids, although considered to be rare are important causes of valvular heart diseases, characterized by plaque-like deposits of fibrous tissue on endocardial surface of valve cusps and leaflets commonly involving the right heart valvular architecture. In rare instances, the left side of the heart can be involved particularly in those with primary bronchial carcinoma, intracardial right to left shunt and those with overwhelmingly high level of circulating cytokines [1-4]. Resumption of patent foramen ovale or atrial septal defects is some of the causes of the left-sided valvular involvement. Left-sided lesions occur in less than 10% of the patients because the substances are mostly inactivated in the lung [1-4]. The left-sided CHD can cause hemodynamic instability that necessitates surgical intervention. As a result, we opted to take this opportunity to review POCT and CHD in a broader sense.

Discussion

History

Carcinoid Heart Disease (CHD) can lead to heart failure and remains a major cause of morbidity and mortality in patients with carcinoid syndrome. POCT is a rare neuroendocrine neoplasm in elderly female patients, accounting for only 0.3% to 1.0% of all carcinoid tumors [5-10].

In POCT, direct venous drainage of vasoreactive substances into the systemic circulation results in cardiac involvement without hepatic metastasis The vasoreactive substances from the carcinoid tumor cause valvular lesion development, however it must be taken with cognisant that these substances may involve other cardiac structures including myocardium, pericardium, vena caval system, out-flow tracts and vasculature [11]. There are few published reports about POCT-related carcinoid heart disease [12]. Further few case reports have been published regarding ovarian carcinoid tumors or mass lesion with cardiac manifestations involving predominantly the tricuspid and pulmonary valves [11]. One report demonstrated an interesting association in a 75-year-old woman with POCT who was admitted with features of severe right-sided heart failure and successfully underwent pulmonic and tricuspid valve replacement, and a Right Ventricular (RV) outflow patch enlargement. In the same report, authors further indicated that the patient

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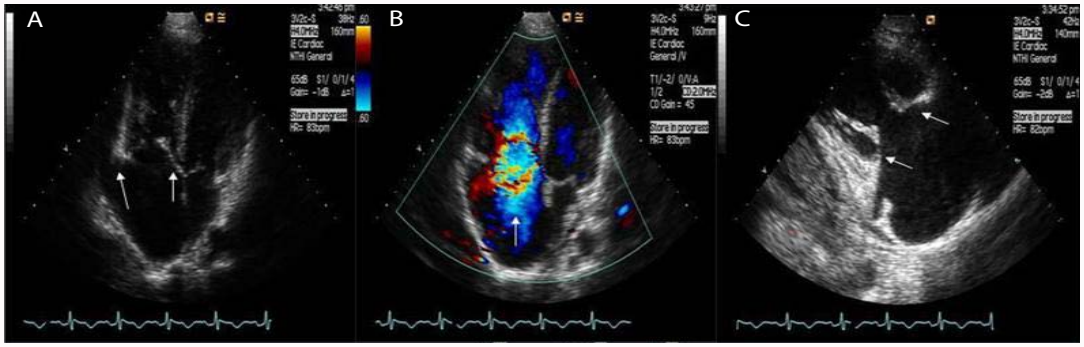

Figure 1: A) is an apical four chamber view: demonstrating thickened; immobile and retracted tricuspid leaflets and minimally thickened mitral valve leaflets (arrows). Also dilated right atrium and ventricle, B) illustrates torrential tricuspid regurgitation (the arrow), C) shows marked failure of coaptation (the arrows) of the tricuspid valve leaflets (Image courtesy of Matshela MR [11]).

subsequently underwent a successful resection of the primary tumor, with complete resolution of her symptoms (REF).

Investigations and diagnosis

Echocardiography is a gold standard and an important modality to utilize at first hand in any suspected case of CHD, to demonstrate retraction and/or fixation of the valvular architecture, and features that are pathognomonic for the specific disease entity, Figure 1 [11,13-15]. Although newer non-invasive imaging modalities has recently been in evolution, 3-dimensional echocardiography could provide an incremental value to better evaluate and characterize heart valvular lesions, right ventricular parameters and function [13-15]. Preoperative 3-dimensional echocardiogram and Cardiac Magnetic Resonance (CMR) imaging could provide detailed anatomic information and additional prognostic indices related to its long-term degenerative changes [13-15]. In fully resourced centers, CMR could provide an accurate anatomic and functional information of cardiac chambers, heart valves and other related structures [16,17]. CMR has an advantage over CT to demonstrate tricuspid valve motion and valvular dysfunction, which could be precisely assessed and quantified using the cine CMR in combination with velocity-encoded cine CMR [16,17]. Despite these diagnostic pros and cons regarding POCT and CHD, a combination of echocardiographic imaging and circulating biomarkers should provide useful insights as well, leading accurately and undoubtedly to the diagnosis [18]. Where applicable, availability of most lesions should be subsequently verified by histological study. To this point, an integrated approach utilizing multimodality imaging, in order to assess the severity of disease, is perhaps the most suitable management strategy.

Treatment

Medical treatment: Strategic management principles of POCT with or without CHD constitutes mainly of somatostatin analogues, which inhibit the release of various biogenic amines and peptides, including serotonin, and resulting in marked symptoms alleviation.

Surgical management: Cardiac surgery should be considered early for patients with symptomatic carcinoid valve disease and those with controlled carcinoid symptoms [19]. The progression of POCT and CHD disease may have an unfavorable impact on surgery and survival outcomes, so early interventional strategic approach is crucial. However, despite these premises there are controversies regarding the choice of surgical management approach and the role or introduction of anticoagulants. Although the optimal timing of surgical intervention has not been fully established, the onset of symptoms may be one of the best timings considering the poor prognostic implications in symptomatic CHD. Therefore, early diagnosis and subsequent follow-up are indispensable.

Prognosis: The follow-up of heart disease is more important especially when a primary tumor is inoperable because there is still a possibility of disease progression. Unexpectedly long-term progression could be seen mainly related to the inoperable state of the undiagnosed POCT. POCT-related CHD should be considered as an important differential diagnosis in any elderly female with isolated right-sided valve disease and ovarian tumor or mass. In general, the mean life expectancy of CHD is estimated to be less 2 years.

Conclusion

POCT is still regarded as a rare occurrence despite published case reports, due to the unusual origin of the primary tumor and atypical valve involvement. POCT are reported to represent almost 0.1% of all ovarian malignancies and less than 5% of carcinoid tumors. In POCT, the tumor's usually affects the right cardiac chambers leading to thickening and retraction of the affected valves, with the tricuspid valve being the most commonly affected, however in rare instances the left side can be affected as well. Therefore, as evidenced in most recent reports, both tricuspid and pulmonary valve are involved, consists a minority of carcinoid valve disease cases. Echocardiography is the fundamental modality of choice for initial evaluation of CHD, with real-time three-dimensional echocardiography providing additional unique views useful to thoroughly dissect the complex anatomy of cardiac valves and valvular apparatus. Where tricuspid and pulmonary valves lesions are severe and patients are unsuitable for valvular intervention, supportive care and heart transplantation should be considered.

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