An Uncommon Case of Tricuspid Regurgitation

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Introduction

Carcinoid heart disease is a sequel of fibrotic reaction incited by vasoactive substances released from hepatic metastasis of carcinoid tumours. The fibrous reaction affects valve leaflets and subvalvular apparatus. It predominantly involves right sided valves and subvalvular apparatus since vasoactive substances are deactivated in the lung. Carcinoid heart disease is seen in 50% of patients with carcinoid syndrome\(^5\).

Case report

A 63 year old man who was diagnosed to have carcinoid syndrome in 2007 with normal echocardiogram presented with right hypochondrial pain, hematemesis and fatigue. On examination he had elevated jugular venous pressure with 'c-v' waves and pulsatile liver. A short systolic murmur was presented in left sternal border. His echocardiogram revealed dilated right atrium and right ventricle. Tricuspid valve leaflets and subvalvular apparatus were thickened, fibrosed and retracted. The valve was immobile in a semi-open position, leading to severe regurgitation (Image1). Pulmonary valves and left sided valves were normal. Continuous wave Doppler shows a dagger shaped pattern of tricuspid regurgitation because of rapid equalization of right ventricle and right atrial pressures.

![Image1](image1)

Modified apical view in systole demonstrating poor mobility and non coaptation of tricuspid valve and severe Tricuspid regurgitation

Discussion

Carcinoid tumours are rare neuroendocrine tumours arise from enterochromaffin cells in the gastrointestinal tract\(^6\). Carcinoid tumours of midgut origin secrete large amounts of vasoactive substances

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like 5-hydroxytryptamine, tachykinins and prostaglandins\(^2\). These substances will be transported by portal circulation to liver and will be inactivated. However when tumour cells metastasize to liver, the vasoactive substances produced can reach systemic circulation leading to carcinoid syndrome. This is characterized by flushing, diarrhea and bronchospasm. Once the carcinoid syndrome has developed, approximately 50% of these patients develop carcinoid heart disease. Since lung can inactivate these vasoactive agents, carcinoid heart disease mainly affects right sided valves. However PFO and septal defects can lead to left sided valve involvement. Carcinoid heart valves demonstrate accumulation of transforming growth factor- beta latency associated peptide and latent binding protein. 5-hydroxy tryptamine (5-HT) receptors are found in human heart valves. 5-HT has been shown to increase synthesis and up-regulate TGF-8by interstitial cells of heart valve\(^2\). Somatostatin analogues reduced the incidence of carcinoid heart disease by inactivating vasoactive substances. Surgical replacement of tricuspid valve gives marked symptomatic relief and survival benefit. The choice of prosthesis is still a debate. Earlier series shows mechanical prosthesis is durable when compared to bioprosthetic valves. But with advent of somatostatin analogues and other antitumour drugs durability of bioprosthetic valve has been increased along with low risk of bleeding\(^2\).

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References.