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A Case of Carcinoid Syndrome with Carcinoid Valvular Heart Disease Treated Successfully

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Abstract

A 39-year-old lady presented in early 2011 with Carcinoid syndrome due to Ileal Carcinoid with liver metastases and Carcinoid valvular heart disease with Right heart failure. Liver resection combined with ethanol injection of Liver metastases was carried out after reduction of Central Venous Pressure by an initial Tricuspid valve replacement (TVR), in a staged manner, with resolution of symptoms. This case highlights the necessity of TVR prior to major Liver resection for Liver metastases in Carcinoid heart disease with Tricuspid regurgitation as well as the value of treating Carcinoid heart disease aggressively.

Key words: Carcinoid heart disease; Tricuspid valve replacement; Liver resection.

Clinical Report

A 39-year-old lady presented in early 2011 with history of postprandial flushing attacks of 6 years duration, pedal oedema and puffiness of face with persistent dry cough of 2 months duration. Elevated Jugular Venous Pressure (JVP), bilateral pitting pedal oedema and nontender hepatomegaly were noted on clinical examination. Ultrasound abdomen showed an 8 × 8 cm mass lesion with mixed echogenicity in the Left lobe, liver. Contrast Enhanced Computerized Tomography (CECT) of abdomen revealed a mixed density mass lesion with irregular contrast enhancement in the Left lobe of liver with multiple enhancing lesions in the right lobe, liver as well as an enhancing lesion in the terminal ileum. Carcinoid syndrome due to Ileal carcinoid with liver metastases and carcinoid valvular heart disease with right heart failure was diagnosed. 24 hours urinary 5 hydroxy indole acetic acid (24 hr urinary 5HIAA) estimation showed a very high value of 153 mg/day. Echocardiography revealed tricuspid stenosis with

regurgitation, mild pulmonary artery hypertension and Right heart failure (Figure 1).

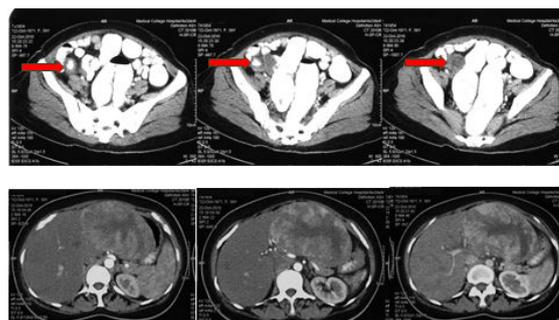


Figure 1 Contrast enhanced CT scan abdomen showing large mixed density liver metastasis.

At the multidisciplinary meeting, further evaluation by Cardiology and Cardiothoracic Surgery regarding the status of pulmonary artery hypertension and the feasibility of tricuspid valve replacement (TVR) was discussed as TVR is a rarely performed procedure with operative mortality in the range of 14% to 17% [1,2]. Further, the risk of TVR in the presence of Carcinoid syndrome was discussed. After detailed evaluation, it was decided to suggest TVR considering the favorable tumor biology and the fact that the resultant drop in Central Venous Pressure (CVP) would make way for a safe liver resection. It is well known that liver resection in the presence of high CVP carries higher morbidity and mortality [3].

However, the relatives wanted time to think over, arrange finances and take a decision on the treatment plan. She was maintained on monthly Depot Octreotide. Meanwhile as she developed persistent pain in the lower abdomen with features of subacute bowel obstruction within a couple of months, a palliative resection of the primary in the ileum was carried out under regional anesthesia.

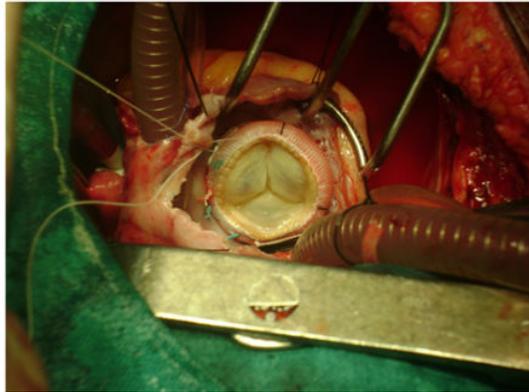


Figure 2 Intraoperative photograph showing replaced bio prosthetic valve.

The intraoperative and postoperative period was stormy with fluctuations of blood pressure and needed short acting Octreotide in addition to other measures. Tricuspid valve replacement with a bio prosthetic valve was carried out in May 2011, the patient being on octreotide and anticoagulants in the perioperative period. She made an uneventful recovery and was discharged (**Figure 2**).

She was taken off oral anticoagulants at 3 months but continued to be on Depot Octreotide. Repeat echocardiography showed normal functioning of the replaced tricuspid valve with no evidence of tricuspid regurgitation. Under cover of Depot Octreotide and infective endocarditis prophylaxis, she underwent left lateral sectionectomy plus multiple metastasectomies of superficial metastasis in right lobe in September 2011. Ethanol injection under intraoperative ultrasound guidance was carried out for deep seated liver metastases, the resection of which was not considered feasible in view of the inadequate future liver remnant it would leave behind. The patient made an uneventful postoperative recovery except for mild atelectasis and the symptoms of carcinoid syndrome abated promptly. The 24 hr urinary 5 HIAA normalized to 3.1 mg/per day at 1 month after surgery and could be weaned off Octreotide. At 50 months follow up in late 2015, although symptom free, she

underwent Peptide Receptor Radionuclide Therapy (PRRT) as she was found to have progressively increasing levels of 24 hour 5 HIAA with increase in size of the residual liver lesions. Following this, her 24 hours 5 HIAA levels have normalized and the residual liver lesions remain stable to date.

It has been demonstrated previously that liver resection should be preceded by TVR in carcinoid heart disease with right heart failure and that Liver resection carried out after TVR does not carry a higher mortality [4,5]. This case highlights the necessity of TVR prior to major Liver resection for Liver metastases in Carcinoid heart disease with Tricuspid regurgitation and the need to offer detailed evaluation and aggressive surgical treatment for carcinoid heart disease by a multidisciplinary approach.

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References

1. Leviner DB, Medalion B, Baruch I, Sagie A, Sharoni E, et al. (2014) Tricuspid valve replacement: The effect of gender on operative results. *J Heart Valve Diseases* 23: 209-215.
2. Songur CM, Simsek E, Ozen A, Kocabeyoglu S, Donmez TA (2014) Long term results comparing mechanical and biological prostheses in the tricuspid valve position: Which valve types are better mechanical or biological prostheses? 23: 1175-1178.
3. McNally, Revie EJ, Massie LJ, McKeown DW, Parks RW, et al. (2012) Factors in perioperative care that determine blood loss in liver surgery. *HPB (Oxford)* 14: 236-241.
4. Lillegard JB, Fisher JE, McKenzie TJ, Que FG, Farnell MB, et al. (2011) Hepatic resection for the carcinoid syndrome in patients with severe carcinoid heart disease: Does valve replacement permit safe hepatic resection? *J Am Coll Surg* 213: 130-136.
5. Bhattacharyya S, Raja SG, Toumpanakis C, Caplin ME, Dreyfus GD, et al. (2011) Outcomes, risks and complications of cardiac surgery for carcinoid heart disease. *Eur J Cardiothorac Surg* 40: 168-172.