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CLINICAL SPOTLIGHT

Carcinoid Heart Disease without Liver Involvement Caused by a Primary Ovarian Carcinoid Tumour

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Carcinoid heart disease, caused by primary ovarian carcinoid tumour, is a rare form of valvular heart disease. This form of heart disease usually presents with symptoms of right-sided valvular dysfunction, ultimately leading to right-sided heart failure. This entity is unique as it develops in the absence of liver metastasis. We report a case of 75 year-old woman with primary ovarian carcinoid tumour who presented with symptoms of severe right-sided heart failure and successfully underwent pulmonic and tricuspid valve replacement along with a right ventricular (RV) outflow patch enlargement. This patient later underwent uneventful resection of the primary ovarian carcinoid tumour, with complete resolution of her symptoms.

Keywords Ovarian carcinoid • Carcinoid heart disease • Serotonin • Right side heart failure • Echocardiogram.

Introduction

Carcinoid tumours are rare neuroendocrine tumours (NET) with an incidence of 1-2/100,000 per year [1]. The majority of carcinoid tumours originate from enterochromaffin cells of the small intestine (41.8%), rectum (27.4%), stomach (8.7%) and bronchopulmonary system (25%) and secrete a large amount of vasoactive peptides such as serotonin, which the liver ordinarily metabolises before it enters into the systemic circulation [2]. Primary ovarian carcinoid tumours are very rare, accounting for less than 0.1% of ovarian cancers and only 1% of all carcinoid tumours [3]. The venous drainage of the ovary bypasses the liver in its course to the inferior vena cava and therefore, ovarian carcinoids can produce the carcinoid syndrome without hepatic involvement [4]. Here, we present such a case of an ovarian carcinoid tumour primary with resultant carcinoid heart disease in the absence of an hepatic carcinoid tumour.

Case Summary

A 75 year-old female with a past medical history of hypertension, atrial fibrillation and hypothyroidism was admitted to our hospital for planned multi-valvular heart surgery. The patient complained of persistent diarrhea and anorexia five years prior to the hospital admission and recently started experiencing dyspnoea on exertion (DOE) which had progressed from walking half a block to one block to even a few steps. The patient had developed recurrent ascites in past requiring repeated paracentesis and had a CT scan of the abdomen eight months prior to presentation, which revealed a pelvic tumour (dermoid). Patient was planned for tumour resection and had a pre-operative echocardiogram which

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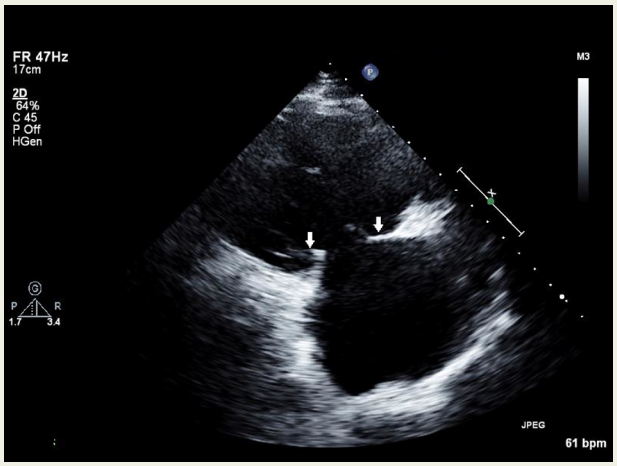

Figure 1 Right ventricular inflow tract view showing diffusely thickened immobile, retracted and non-coapting tricuspid valve (marked with white arrow) with enlarged right atrium and ventricle.

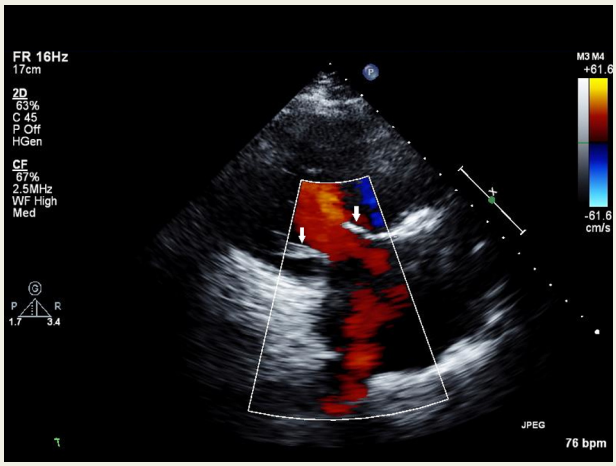

Figure 2 Right ventricular inflow tract colour Doppler view showing severe tricuspid regurgitation with retracted wide open tricuspid valve (marked with white arrow).

revealed severe right ventricle (RV) dilatation and right atrial (RA) dilatation; tricuspid stenosis (TS), thickened tricuspid valve leaflets, with limited mobility, severe central tricuspid regurgitation (TR) [Figures 1, 2], pulmonic stenosis (PS) and severe pulmonary regurgitation (PR) with leaflets appearing thickened and fused in an open position. Based on the echocardiographic findings, the patient was considered a poor surgical candidate and was referred to a carcinoid centre for possible carcinoid heart disease.

The patient presented to Carcinoid Heart Center in New York City and had laboratory tests including 24 hour urine 5-Hydroxyindoleacetic acid (5-HIAA) -60 mg (2-6 mg/24 hour), Serotonin- 1618ng/ml (50-220 ng/ml), chromogranin A- 178 ng/ml (0-95 ng/ml), pancreastatin >1920 pg/ml (0-88 pg/ml) and Neuron-specific enolase -63.3 ng/ml (15ng/ml). Patient also underwent an Octreoscan which revealed a large Octreotide-avid right anterior pelvic mass. The patient's diagnosis was confirmed as right anterior pelvic Neuro endocrine tumour (NET) with carcinoid heart disease. She was admitted to our hospital for multi-valve cardiac surgery prior to the tumour-reductive surgery because of the excessive risk of abdominal surgery in the presence of significant hepatic congestion resulting from the right heart failure. The patient underwent pulmonic valve replacement (PVR) and tricuspid valve replacement (TVR) with tissue valves, and an RV outflow tract patch enlargement to enable placement of a larger valve [Figure 3]. The patient tolerated the procedure well and was discharged from the hospital with a planned pelvic carcinoid resection in three months.

On follow-up visit to the carcinoid centre two weeks after cardiac surgery, the patient reported marked symptomatic improvement and had decreased levels of serotonin,

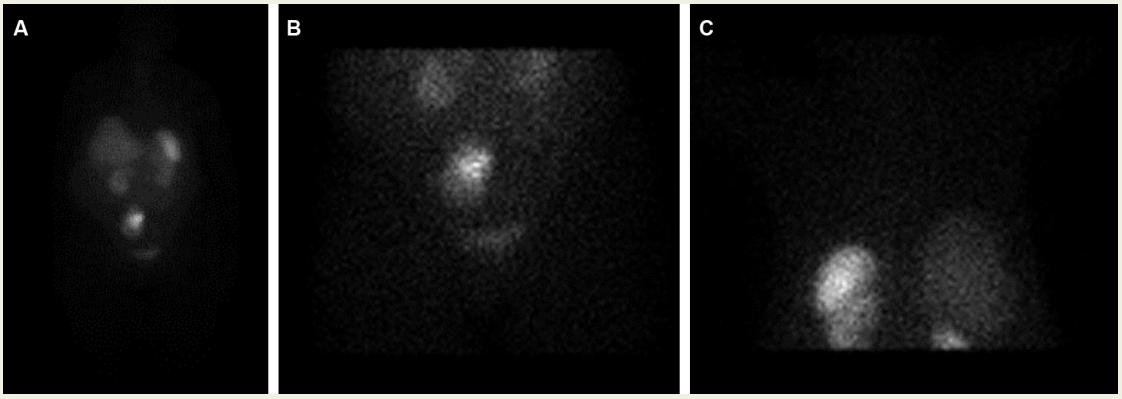

Figure 3 Scintigraphic imaging with Indium-111-labelled Octreoscan showing a large right anterior pelvic mass with enhanced uptake of the radioisotope within the mass.