

## Primary Ovarian Carcinoid Heart Disease Curatively Treated with a Two-Stage Procedure

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### Abstract



Primary ovarian carcinoid disease with cardiac involvement is a very rare syndrome, which requires interdisciplinary management. We report on a patient with primary ovarian carcinoid tumor with severe tricuspid valve insufficiency who was curatively treated after successful tumor resection and tricuspid valve replacement.

### Key words

Heart disease · heart valve surgery · carcinoid

### Introduction



Carcinoid tumors are rare, with an incidence of 1.2 to 2.1 per 100 000 persons per year. Primary ovarian carcinoid tumors account for less than 0.5% of all carcinoid tumors. The characteristic feature of primary ovarian carcinoid tumor which distinguishes it from the more common carcinoid tumors of the gastrointestinal tract is the development of carcinoid heart disease in the absence of liver metastases, as the venous drainage of the ovaries bypasses the portal circulation. The distinctive carcinoid cardiac lesions consist of fibrous tissue deposits devoid of elastic fibers on the ventricular aspect of the pulmonic valve cusps [1]. In this report, we present the case of a patient with a primary carcinoid tumor of the left ovary with severe tricuspid valve insufficiency where the primary diagnosis was based on typical echocardiographic findings.

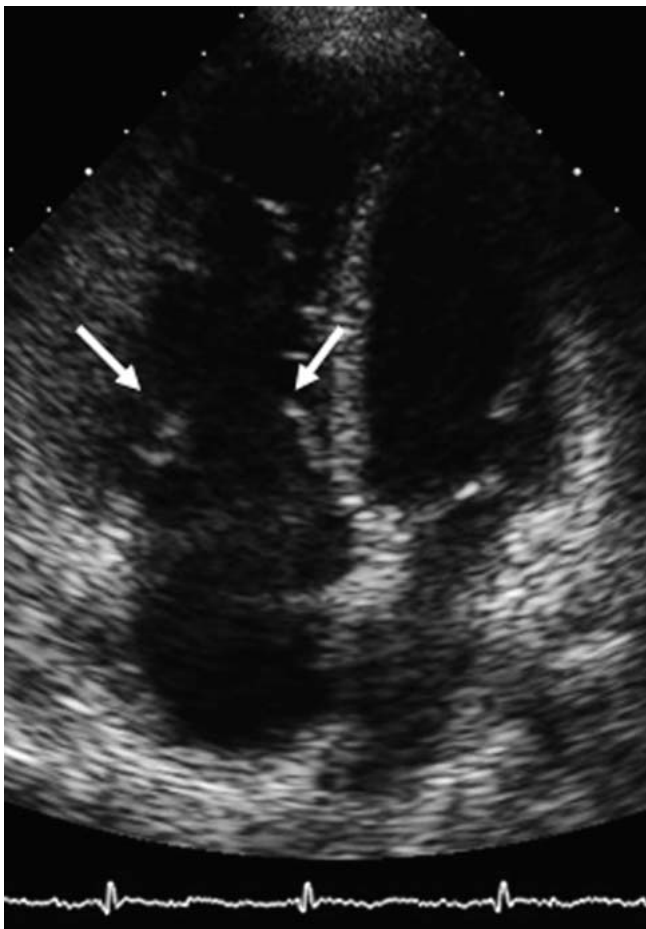
### Case Report



A 73-year-old woman was admitted to the cardiological outpatient clinic with increasing dyspnea and flushing over the last 6 months. Physical examination revealed distension of both jugular veins and lip cyanosis. A grade 2/6 systolic ejection murmur was noted along the left sternal border without irradiation. Neither cyanosis nor edema of the lower extremities could be detected.

Two-dimensional and Doppler echocardiography showed normal left ventricular size and systolic function but a marked enlargement of the right heart chambers (► Fig. 1). The finding of severe tricuspid valve regurgitation due to a thickened tricuspid valve with akinetic leaflets was pathognomonic for a carcinoid cardiac lesion. Carcinoid syndrome with cardiac involvement was corroborated by means of blood chromogranin A concentration measurement (> 420 U/L) and 24-hour urinary excretion of 5-hydroxyindoleacetic acid (5-HIAA). The patient underwent abdominal computerized imaging to identify the primary tumor. An 8.2 × 6 × 6-cm cystic and solid mass extending from the left ovary without evidence of metastatic disease was detected (► Fig. 2). A large accumulation of isotope was detected above the bladder on octreotide scan. There was no evidence of metastatic disease either in repeated MR scans or in contrast agent sonography of the liver. Classical nested and trabecular carcinoid structures with uniform, middle sized tumor cells with round nuclei with coarse chromatin were detected histologically in a percutaneous core biopsy from the lesion.

The patient underwent laparotomy, during which an 8 × 6 × 6-cm firm, relatively mobile mass occupying the entire left ovary was resected (total salpingo-oophorectomy). Histological examination revealed an ovarian carcinoid tumor which had developed



**Fig. 1** Systolic apical four-chamber view showing a markedly dilated right atrium (RA) and right ventricle (RV) with thickened and retracted leaflets of the tricuspid valve (arrows). Note that the tricuspid valve is not capable of closing during the systole.

into a mature teratoma (● **Fig. 3**). Immunohistochemically, the tumor was pancytokeratin AE1/AE3, synaptophysin, CD56, and chromogranin A positive. The postoperative period was uneventful and the patient was discharged home.

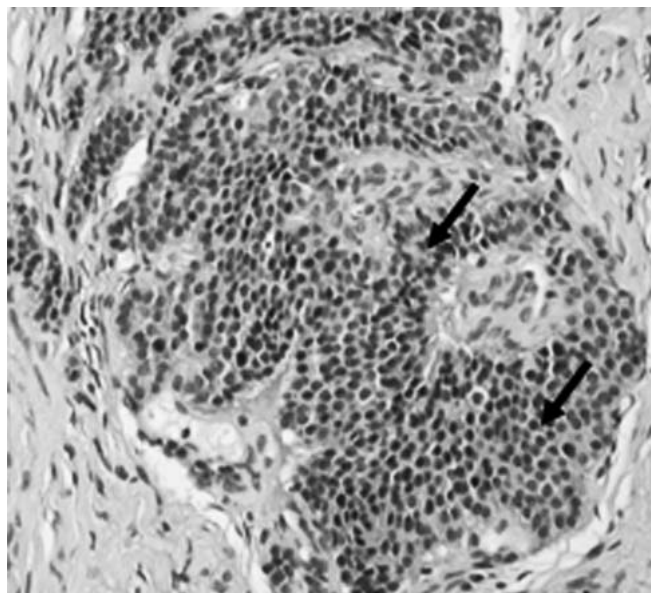
In the 10-month follow-up, tachycardia, flushing or diarrhea no longer occurred and 5-HIAA urine levels and serotonin serum levels were normal; however, the patient had clinical symptoms of right heart congestion, including dyspnea (NYHA Class III to IV) and edema of the lower extremities. The patient underwent tricuspid valve replacement with native leaflet preservation using a 31-mm St. Jude Medical Biocor bioprosthesis following an unsuccessful attempt to perform valve reconstruction due to the restrictive pattern of the tricuspid valve disease (● **Fig. 4**). The postoperative period was uneventful and the patient was discharged home on the seventh postoperative day.

## Discussion

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The course of the present case shows that primary ovarian carcinoid disease with cardiac involvement can be curatively treated by surgery. The improved performance status after tricuspid valve replacement and complete resolution of the carcinoid-associated symptoms after tumor resection clinically support this hypothesis. Due to the pathognomonic malformation of the tricuspid valve, the primary diagnosis of carcinoid heart disease



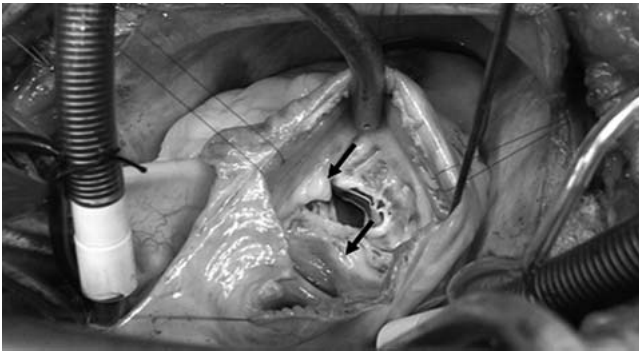
**Fig. 2** Axial contrast-enhanced CT showing an 8.2 × 5.6 × 5.8-cm contrast-enhancing mass of left ovarian origin that proved to be the primary tumor (arrows). The lesion has multiple cystic components and is located anteriorly to the uterus.



**Fig. 3** Trabecular structures of uniform, middle sized tumor cells with round nuclei with coarse granular chromatin surrounded by fibroblast-rich stroma in ovarian carcinoid; H&E stain, original magnification 100 ×.

can be easily made by echocardiography performed by experienced hands. The presence of carcinoid syndrome with cardiac involvement has significant implications for disease prognosis, since it is related to significantly reduced survival rates [2]. The role of echocardiography gains additional importance as the presence of carcinoid tumor with cardiac involvement cannot be excluded on the basis of negative symptomatology and clinical examination [3].

Although medical treatment for carcinoid tumors has become increasingly effective in terms of the improvement of quality of life and mid-term survival, the presence of cardiac involvement is a significant limiting factor. Surgical treatment of carcinoid heart disease has proven beneficial even in patients with unresectable tumor metastases [4]. The present case emphasizes that carcinoid syndrome consisting of a non-metastatic extra-intestinal carcinoid tumor with cardiac involvement can be curatively



**Fig. 4** Intraoperative findings of the tricuspid valve. Note fibrotic thickening of the posterior and the septal leaflet (arrows).

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#### Bibliography

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treated by means of a two-stage surgery which includes tumor resection and treatment of the carcinoid heart disease.

There are few data which address the question of which procedure should be performed first. The advantages of primary tumor resection before treatment of the heart disease include relief of carcinoid-associated symptoms, more convenient anesthetic management during cardiac surgery and time to reconsider the need for heart valve replacement or reconstruction after depletion of vasoactive substances from the blood circulation [5]. On the other hand, addressing valve surgery first may be advantageous in terms of preventing a rapid progression of carcinoid heart disease to severe right heart failure, thus reducing the anesthesia risk.

The timing of cardiac surgery plays an important role for the selection of the operative strategy. Both reconstruction and replacement of the tricuspid valve using a biological prosthesis prior to tumor resection has the disadvantage of a potential mid-term failure due to progression of valve disease due to the ongoing secretion of vasoactive tumor substances. In this case, adjuvant treatment using synthetic somatostatin agonists and/or embolization of the hepatic artery should be previously considered. Previous reports recommending tricuspid valve replacement with a mechanical prosthesis seem to be outdated and are only based on single case observation studies. We suggest that the choice of prosthesis should be individualized according to the needs of the patient [6].

In summary, a very rare primary ovarian carcinoid tumor with cardiac involvement can be curatively treated by means of a two-stage surgical procedure. The ideal timing for tumor resection and cardiac surgery should be defined on an individual basis for each patient according to the respective risk-benefit stratification.

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