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Simultaneous Tricuspid and Pulmonary Involvement in Carcinoid Heart Disease

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Abstract

Introduction: Carcinoid tumors (CT) are malignant neuroendocrine tumors characterized by excessive secretion of serotonin. Cardiac involvement (CI) occurs in 40%, characterized by valvular right-sided involvement. We report a rare case of simultaneous involvement of tricuspid and pulmonary valves.

Case report: A 63-year-old man with a diagnosis of digestive endocrine carcinoma with hepatic metastases was sent to our department for cardiovascular check-up. The physical examination revealed leg edema and intense systolo-diastolic murmur. The transthoracic echocardiography showed very dilated right cavities with right/left-ventricule-ratio of 1,36 and right atrial area of 30cm2, septal flattening and preserved systolic function. Tricuspid valve appeared rigid with important restricted mobility and poor coaptation(gap=15mm) responsible for massive regurgitation. Pulmonary valve appeared also rigid with poor coaptation(gap=6mm) and severe regurgitation. The left ventricle function was preserved, and left-sided valves were normal. The patient received somatostatin analogue therapy and diuretics, but since it was insufficient; mechanical double heart valve replacement was indicated.

Keywords: Carcinoid heart disease; Carcinoid tumors; Tricuspid and pulmonary involvement

Introduction

Carcinoid tumors (CT) are malignant well-differentiated neuroendocrine tumors, rare and characterized by excessive secretion of serotonin, expressed by symptoms such as flushing, diarrhea, and bronchoconstriction [1,2]. Cardiac involvement in carcinoid tumors occurs in approximately 40% of patients. It is characterized by right-sided valvular involvement and can lead to right-sided heart failure [2]. Recently, both right- and left-sided heart disease have been reported. Carcinoid heart disease is a prognostic factor of carcinoid syndrome, however, the prognostic has been greatly improved by advances in valvular replacement surgery [3]. Medical therapy with somatostatin analogs and resection of primary tumor are also good treatment option.

Through the case of a 63-year-old man with a digestive endocrine carcinoma complicated by carcinoid heart disease, we will discuss carcinoid syndrome and the particularities of cardiac involvement. In particular, in this case, we noted simultaneous tricuspid and pulmonary involvement, which is very rare.

Case Presentation

We report the case of a 63-year-old man, chronic smoker, who was admitted initially to the department of gastro-enterology after presenting abdominal pain, alternation between constipation and diarrhea and increasing dyspnea. He has no particular personal or family pathological history. The physical examination revealed a firm hepatomegaly with smooth anterior surface, without splenomegaly, oxygen saturation at 90%, leg edema and an intense systolic murmur suggestive of tricuspid valve regurgitation. Liver status, hemogram, renal function, protidogram, and alpha-fetoprotein were all normal. Digestive endoscopy was normal.

Thoracic and abdomino-pelvic CT-scan showed multiple hepatic and mesenteric nodules. Liver biopsies showed a secondary localization of a well-differentiated endocrine carcinoma, grade 2. An immunohistochemical study showed positivity for both chromogranin and synaptophysin. The diagnosis of digestive endocrine carcinoma with hepatic metastases has been then confirmed, the patient was sent to our department for cardiovascular check-up. The transthoracic echocardiography showed very dilated right heart cavities with a right/left-ventricule-ratio of 1,36, right atrial (RA) area of 30 cm^2, with septal flattening and tricuspid ring diameter of 44 mm., with a good function in which tricuspid annular plane systolic excursion (TAPSE) was 26 mm and right ventricular (RV) velocity (S') = 14 cm/second.

Tricuspid valve appeared rigid with important restricted mobility and poor coaptation (gap=15mm) responsible for laminar massive tricuspid regurgitation (Figure 1). Pulmonary valve appeared also rigid with poor coaptation (gap=6mm) and severe pulmonary regurgitation (Figure 2). The left ventricle function was preserved, and left-sided valves were normal.

The patient received a medical treatment based on Somatostatin analogs therapy (one injection/3 weeks), associated to diuretics, but since it was insufficient; mechanical double right-heart valve replacement was indicated (Figure 3).

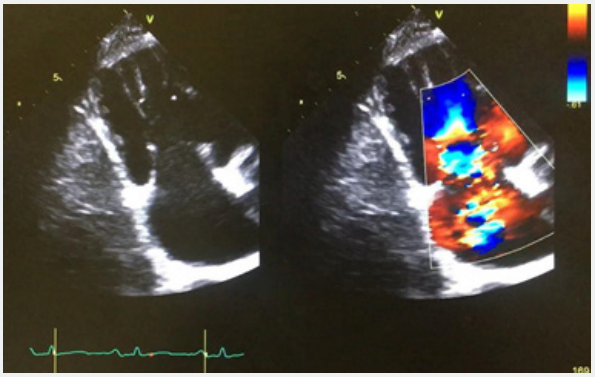

Figure 1: Transthoracic echocardiaography apical view focused on right chambers showing thickened and retracted tricuspid valve with severe regurgitation and coaptation gap.

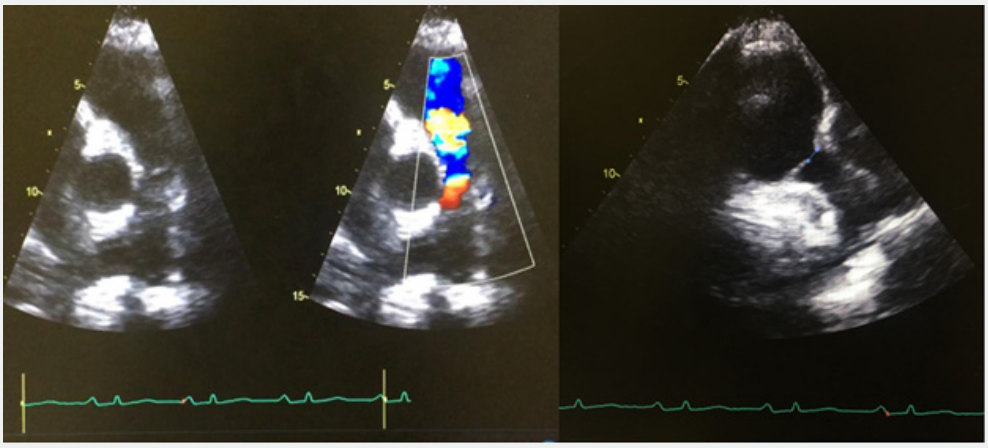

Figure 2: Parasternal short axis view showing thickened and retracted pulmonary valve with severe regurgitation and coaptation gap.

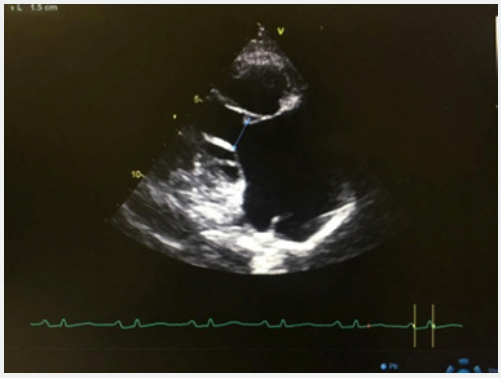

Figure 3: Modified parasternal long axis view focused on right heart showing a coaptation gap of 15 mm in the tricuspid valve.

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Discussion

Carcinoid tumors are rare. They sit in 90% of the cases in the digestive tract, particularly in the appendix or the ileum. The carcinoid tumor secretes more than 20 humoral substances, the most important of which are serotonin, histamine and kinin peptide [1]. However, these vasoactive substances are rapidly inactivated by the liver. Cardiac involvement presupposes the existence of secretory liver metastases or a carcinoid tumor located in an area not drained by the portal system.

Conventionally, the carcinoid syndrome is characterized by facial flushes, bronchospasm, diarrhea, and right-sided valvular involvement. The deposition of plaques results in pathognomonic right-sided valvular involvement of retracted and fixed valve leaflets, which result in combined regurgitation and stenosis, and, over time, right ventricular enlargement and failure. These deposits usually involve the right side of the heart (90% of cases), specifically, the downstream side of the valve leaflets, that is, the ventricular aspect of the tricuspid valve and pulmonary arterial side of the pulmonary valve [2,3]. Simultaneous involvement of the tricuspid and pulmonary valves strongly suggests carcinoid heart disease as a likely diagnosis (as it was for our patient). Left-sided valves are rarely affected because of pulmonary metabolism and desactivation of the hormonal substances [4]. However, left-sided involvement is common in patients with a patent foramen ovale [5], bronchial carcinoids due to the bypassing of inactivation of serotonin within the lung [6].

Cardiac manifestations are often silent despite severe valvular involvement. The physical examination may at a later stage show signs of right heart failure (edema, turgid jugular veins, hepatomegaly). The diagnosis is essentially based on biological examinations and echocardiography which remains the principal imaging modality in assessment of carcinoid heart disease [7-9]. Biological examinations are helpful for the diagnosis: High levels of N-terminal pro-brain natriuretic peptide (NT-proBNP), chromogranin-A (a neuroendocrine secretory protein), and urinary 5-HIAA (a metabolite of serotonin) are correlated with the progression of Carcinoid heart disease.

Echocardiography shows typical valvular involvement: the tricuspid valve is constantly thickened, rigid, and retracted with restricted mobility and poor coaptation which lead to tricuspid regurgitation (as in our case). Stenosing character is rarer (25% of cases) but may be associated. The pulmonary valve, which is more difficult to study, is reached in 30% of cases with regurgitation (as in our case) or pulmonary stenosis. Patients with the rare diagnosis of carcinoid heart disease developing in the context of metastatic neuroendocrine tumors and carcinoid syndrome are treated in specialized centers by an experienced multidisciplinary team comprising endocrinologists, oncologists, cardiologists, surgeons, and pathologists [10]. They have a decreased life expectancy compared to whom without cardiac involvement [11] and they usually die as a result of severe tricuspid regurgitation rather than carcinomatosis, and thus, surgical replacement is performed early, since it is the only effective curative treatment.

There is no optimal moment for valve replacement surgery. However, valvular surgery is proposed when patients become symptomatic or develop ventricular dysfunction [12].

It is recommended to use mechanical prostheses in the presence of carcinoid disease because of the fibrous degeneration seen in cardiac valves and the similar potential in bioprosthetic valves [13]. When the pulmonary and tricuspid valves are both involved, open pulmonary valvulotomy is a reasonable option [14].

Medical treatment is based on chemotherapy (interferon alpha, doxorubicin, 5FU), hepatic arterial chemo-embolization, eventually control of the heart failure (diuretics), and especially somatostatin analogs.

Conclusion

Carcinoid syndrome is rare and can lead to heart failure especially when it is a simultaneous tricuspid and pulmonary involvement. Echocardiography is recommended for cardiac evaluation. Surgical valve replacement can improve the clinical outcome and the prognosis of those patients.

Conflict of Interest

The authors declare that they have no conflicts of interests..

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