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Metastatic Carcinoid Tumor to the Heart, A Dreadful Complication of Neuroendocrine Tumors Yet Under Estimated: About an Observation

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ABSTRACT

Cardiac metastases are uncommon in neuroendocrine tumors (NET). Features of patients with metastatic carcinoid tumor involving the heart have not been well described. Although carcinoid tumors have been described in almost every organ, few reported cases of confirmed cardiac involvement. The overall incidence of myocardial carcinoid metastases among patients with metastatic carcinoid disease is about 4%. There is limited knowledge on the prevalence, clinical presentation, and management of heart metastasis. The availability and increasing use of modern imaging techniques leads to more frequent discovery of rare metastatic sites. Our aim is to further clarify the clinical manifestations of heart metastasis, to increase the knowledge of rare localizations of NET metastasis and insist on the systematic screening of metastatic carcinoid heart disease as an integral part of the echocardiographic evaluation in patients with carcinoid syndrome. We report the case of a 67-year-old male with history of a low differentiated neuroendocrine tumor, presenting with palpitations and dyspnea and which investigations have led to diagnose myocardial metastasis signing the progression of the NETs.

KEYWORDS: Carcinoid Syndrome, Myocardial Metastasis, Carcinoid Heart Disease.

INTRODUCTION

Neuroendocrine tumors (NETs) are rare, mostly located in the gastro-entero-pancreatic tract or the broncho-pulmonary system. These tumors originate from neuroendocrine cells. NETs most commonly metastasize to the lymph nodes and liver; rarely in brain, breast, ovaries, adrenal glands, orbita and skin. Cardiac metastases have been sparsely reported in literature and estimated at 1–4 % of cases [1].

In this paper, we report the case of a 67-year-old male who was referred to cardiology department with a history of palpitations and which investigations have led to diagnose myocardial metastasis signing the progression of his NETs.

CASE REPORT

A 67-year-old man, with history of hypertension since 3 years, and family history of a sister who died of colic cancer three years ago.

In 2013, the patient had undergone prostatectomy for a benign prostatic hypertrophy. Two years later, two left inguinal nodes appeared and the pathological analysis of the biopsy revealed metastasis of a low differentiated neuroendocrine tumor with capsulor invasion and vascular emboli. Patient was treated with combined concomitant chemotherapy (cisplatin + etoposide) and radiotherapy with good outcome. The patient was assessed each year by his oncologist.

In 2018, the patient was referred to our service of cardiology to explore palpitations that appeared 6 months ago associated with slight dyspnea. The initial examination has shown no cardiovascular abnormalities, the left inguinal adenopathy has reappeared and its volume has grown compared to first time.

Transthoracic Echocardiography (*figure 1*) has shown two intra myocardial masses, on the apex and in the apical segment of the antero-septal wall measuring respectively 10 / 15mm and 6 / 10mm. No pericardial, endocardial or valvular abnormalities were detected.

We realized a body CT scan that showed multiple tumor locations: right axillary, peritoneal, left adrenal, thoracic anterior wall and perineal tissues.

Given the multiplicity of the affected sites, we decided to proceed with a PET SCAN (*figure 2*) that confirmed the multiple sittings of tumorous masses with hyper metabolism of the metastatic foci on cardiac location, right axillary, right parietal and sternal areas, the thyroid and prostatic gland.

ECG Holter has shown paroxysmal atrial fibrillation without any other rhythm disorder. The biology has shown no abnormalities so far.

10 days after his hospitalization, Mr Ch developed a flaccid paraplegia due to metastatic spinal compression D10-D11 that required a surgical decompression. Then the patient started immediately sessions of adapted palliative concomitant chemo-radiotherapy.

The evolution was marked by the improvement of functional symptomatology of the patient as well as the reduction of all his tumefactions including the cardiac location, until total disappearance after 8 sessions.

Patient has been stabilized under treatment with an uneventful follow-up of 3 years.

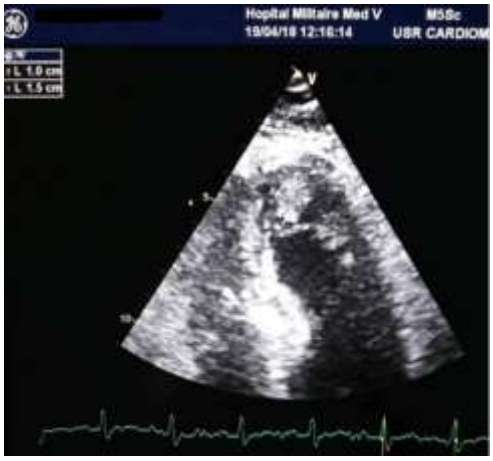

Figure 1: Transthoracic echogardiographic four chambers view showing the presence of the mass in the apex of left ventricule.

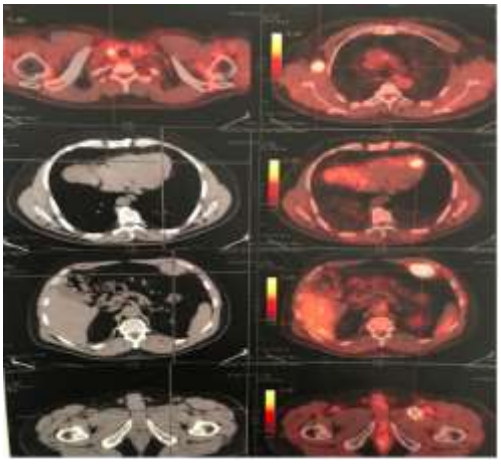

Figure 2: Body PET scan showing multiples metastasis locations with hypermetabolic activity.

DISCUSSION

NETs are commonly asymptomatic, but they can be symptomatic due to the excretion of hormones. Most frequent sites of metastases are lymph nodes and liver in more than 80 % of the patients

When metastases are present, small intestinal serotonin-producing tumors may lead to the carcinoid syndrome, a clinical syndrome characterized by flush, diarrhea, and bronchoconstriction. Long-standing serotonin secretion may lead to endocardial fibrosis especially of the right heart; it may concern the valves and their proximity and is responsible of a valvular retraction leading to regurgitation rather valvular stenosis. In fact, Serotonin stimulates in vitro the proliferation of fibroblasts via its 1B-receptor, and thus carcinoid heart disease at later stages of the disease [2].

Significantly higher urinary levels of 5-HIAA (a serotonin metabolite) have been reported by several authors in patients with carcinoid heart disease; [3]

Cardiac metastasis are exceptional [4-6] (4% of metastatic locations), however, they must be searched for systematically because they represent ultrasound features that allow a reliable diagnosis when their diameter reaches 1cm. They are tumors located in the right or left myocardium, homogeneous, well defined and non-infiltrating. They may be present without involving the valves. [7]

According to one study [8], 38.8% of 18.751 autopsies (patients who died in hospital between 1994 and 2003) demonstrated a neoplastic process of which 9.1% (662 autopsies) also included cardiac metastasis. The analysis of these cardiac lesions showed a pericardial invasion in 2/3 of cases, myocardial in 25% of cases and endocardial in 5% of cases. This important series did not show any predominance of man-woman. The primary tumors most commonly found were mediastinal tumors and melanomas with some cases of pulmonary adenocarcinoma and mesothelioma.

The clinical presentation of a cardiac metastasis is most often silent. The symptomatology depends on its extent and location. In they are pericardial, serous or hematological effusion may be observed. When it concerns the myocardium, one will rather observe disturbances of the rhythm or disorders of conduction like our patient whose ECG Holter has shown paroxysmal atrial fibrillation. In cases of intracavitary implantation, neoplastic embolism or cardiac decompensation may be observed. Right intracavitary localizations are frequent and favored by low pressure, slowed blood flow and less contractile force, which explains the rarity of implantations in the left heart and valvular system. [9]

The clinical evolution is usually disappointing, and patients with cardiac metastasis die within a year of the diagnosis, on average. [8,9]

The poor prognosis related to cardiac involvement is well established. In Pellikka's study [3], the 3-year survival of patients with cardiac carcinoid syndrome was of 30% compared to 60% for those without heart damages. Pellikka et al [3] in a series of 74 patients with evidence of carcinoid

heart disease reported that overall survival was significantly worse for patients with cardiac involvement; median survival was only 1.6 years. Himelman and Schiller [10] also noted similar shortened survival. However, Pandya et al [7] reported a higher rate of survival of 9.5±4 years from time of diagnosis of the carcinoid syndrome and of 6.3±5 years from time of diagnosis of metastatic carcinoid heart tumor.

Our patient has a follow-up for 3.5 years. He was stabilized under palliative radio-chemotherapy, his myocardial carcinoid metastasis as the other locations have responded favorably without the need for biopsy or surgery; echocardiographic controls have shown regression until total disappearance of cardiac damages as well as on CT scan. Unlike some authors [3] who reported that despite treatment that resulted in significant reductions of urinary levels of 5 - HIAA, there was no regression of the carcinoid heart lesions in any of their series worsening thus the prognosis of carcinoid heart disease.

CONCLUSION

Cardiac involvement in patients with carcinoid disease carries important prognostic implications. Though under diagnosed, the treating clinicians should assess regularly these patients by serial examinations, as several develop typical features of carcinoid heart disease during follow-up.

Echocardiography and MRI are methods of choice for diagnosis and follow-up, while PET-CT may contribute to earlier detection. Echocardiographic assessment should include a search for carcinoid cardiac metastases, even in the absence of carcinoid valvular disease. Their management is challenging and requires multidisciplinary team to optimize the outcomes. The prognosis is often reserved with higher mortality.

CONSENT

The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest to report.

ETHICAL APPROVAL

We confirm that the manuscript has been read and approved by all named authors. The protection of intellectual property associated with this manuscript had been in our consideration.

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