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Case Report

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Carney Complex: "The Complex of Cardiac Myxoma"

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Introduction

The Carney complex (CNC) is a rare dominantly inherited syndrome characterized by spotty skin pigmentation, endocrine overactivity and myxomas. About 500 patients have been registered by the NIH-Mayo Clinic (USA) and the Cochin center (France) [1]. Cumulative reports from these centers, plus information from the Cornell center in New York, indicate that there are about 160 index cases of CNC presently known since the disorder was first described in the medical literature in 1985 [2].

It affects males and females in equal numbers and can present at any age with the mean age at diagnosis being 20. Cardiac myxomas can develop in any cardiac chamber and may be multiple. They comprise main feature of Carney Complex in 30-60% of cases and may be responsible for the high rate (16%) of sudden death historically reported in CNC families [2].

Carney Complex is characterized by spotty skin pigmentation, cardiac (heart) myxomas (tumors composed of mucous connective tissue), skin myxomas, endocrine tumors or over-activity, and schwannomas.

Case Report

A case of 56-year-old female patient with past medical history of two ischemic strokes, myocardial infarction, GERD, atrial myxoma resection, Addison disease, and hypothyroidism, who presents to the ER due to intermittent suprapubic pain that started a month ago. The patient described it as localized, stabbing in nature, 10/10 in intensity, 20 minutes duration, without alleviating or aggravating factors. She had associated nausea, non-bloody emesis, diarrhea that improved with loperamide, hematuria, decreased urine output, and dyspnea on exertion. The patient was admitted with the diagnosis of gastroenteritis, but had an echocardiogram performed due to dyspnea on exertion expressed during review of system.

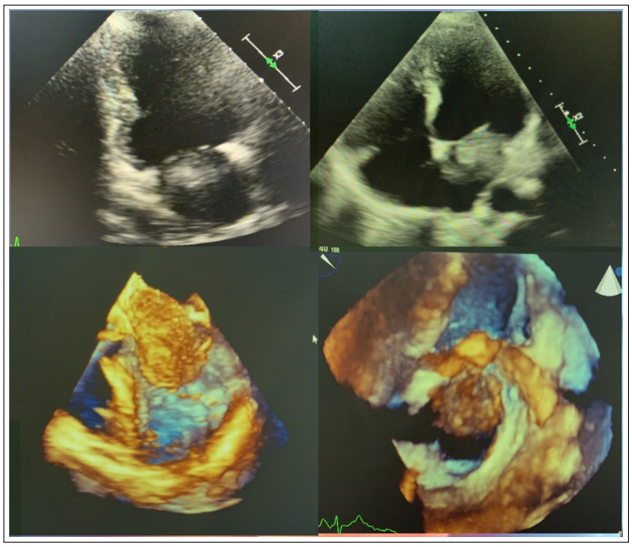
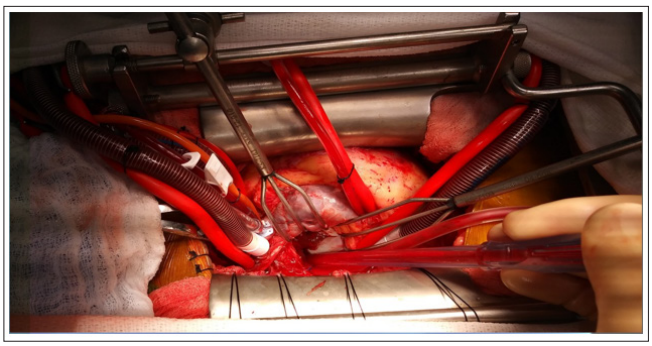


Figure 1: An echocardiogram revealed a 4cm x 4cm mass at the posterior mitral valve annulus around the posterior commissure, causing an obstruction. Due to her history and the unusual position of the mass, it was deemed a recurrent left atrial myxoma.



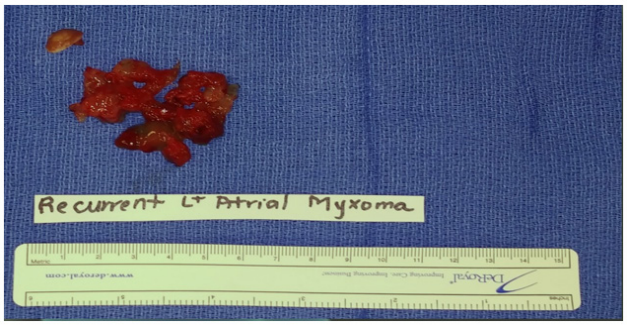


Figure 2, 3: Resection of the myxoma was performed and the stalk was excised from the mitral valve annulus, including the endocardium.

The post- operative course was complicated by post-cardiotomy heart failure requiring hemodynamic support. Finally, the patient improved and eventually was discharged to a skilled nursing facility for rehabilitation.

Discussion

Approximately 500 affected individuals have been reported since the disorder was first described in the medical literature in 1985. This is thus a rare syndrome. Carney complex differs from Carney triad. Carney triad encompasses three types of tumors: a gastric stromal sarcoma, functioning extra-adrenal paragangliomas, and pulmonary chondromas [1]. Cardiac myxomas may cause strokes due to embolism in addition to heart failure [2]. It is therefore important to screen regularly at least once a year by echocardiogram. 1Patients with a history of cardiac myxoma, should be screened annually.

Conflicts of Interest

None of the authors have any relevant financial disclosures

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