Right atrial myxoma in a patient with Carney syndrome: a case report

Biljana Hržić*,
Nikolina Slamek,
Katarina Grandavec,
Martina Vidak,
Petra Kušenić,
Magdalena Kunić,
Ivica Benko

Dubrava University Hospital, Zagreb, Croatia

KEYWORDS: Carney complex, cardiac myxomas.

CITATION: Cardiol Croat. 2024;19(11-12):572. | https://doi.org/10.15836/ccar2024.572

***ADDRESS FOR CORRESPONDENCE**: Biljana Hržić, Klinička bolnica Dubrava, Avenija Gojka Šuška 6, HR-10000 Zagreb, Croatia. / Phone: +385-1-5540-677 / E-mail: bhrzic@kbd.hr

ORCID: Biljana Hržić, https://orcid.org/0000-0001-5441-0900 • Nikolina Slamek, https://orcid.org/0000-0002-2975-8793
Katarina Grandavec, https://orcid.org/0000-0002-6421-6471 • Martina Vidak, https://orcid.org/0009-0006-1973-8509
Petra Kušenić, https://orcid.org/0009-0008-8617-9932 • Magdalena Kunić, https://orcid.org/0000-0001-7402-8135
Ivica Benko, https://orcid.org/0000-0002-1878-0880

Introduction: Carney Complex (CNC) is a rare, autosomal dominant genetic syndrome characterized by the development of benign connective tissue tumors (myxomas), skin pigmentation changes, endocrine abnormalities, and tumors in other tissues. This syndrome is associated with mutations in the PRKAR1A gene, which plays a crucial role in the regulation of cellular growth. Cardiac myxomas are rare in the general population, with an incidence of 0.5–1 per million people per year, but they are frequently observed in patients with Carney Complex, appearing in 20–40% of cases. These myxomas are a leading cause of death among CNC patients, often being multiple and sometimes present at birth. Recurrence of myxomas in these patients is linked to the excessive secretion of growth hormone. Surgical treatment is the primary therapeutic approach for atrial myxomas, and regular follow-up is essential for preventing complications and managing the risk of recurrence.[1,2]

Case report: This case involves a 46-year-old female patient who was hospitalized due to a tumor in the left atrium. She had a history of Hashimoto's thyroiditis, iron deficiency anemia, and mild allergic asthma. The tumor, measuring 37x47 mm, was attached by a stalk to the interatrial septum, and was diagnosed at another healthcare facility. Clinical signs and symptoms raised suspicion of Carney Complex, leading to a thorough endocrinological evaluation. After a cardiology and cardiothoracic surgery consultation, surgical intervention was deemed the optimal treatment approach. The surgery was successfully performed, and the patient was discharged in good condition with recommendations for ongoing follow-up.

Conclusion: Nursing care for patients with atrial myxomas and Carney Complex requires a specialized, individualized approach due to the complexity of the condition. It is crucial to recognize symptoms early, provide necessary support, and ensure a safe recovery. The focus is on continuous assessment, multidisciplinary collaboration, and patient support to minimize the risk of complications and achieve optimal treatment outcomes.

RECEIVED:
September 20, 2024

ACCEPTED:
October 31, 2024

LITERATURE

1. Pitsava G, Zhu C, Sundaram R, Mills JL, Stratakis CA. Predicting the risk of cardiac myxoma in Carney complex. Genet Med. 2021 Jan;23(1):80-85. https://doi.org/10.1038/s41436-020-00956-3
2. Bouys L, Bertherat J. MANAGEMENT OF ENDOCRINE DISEASE: Carney complex: clinical and genetic update 20 years after the identification of the CNC1 (PRKAR1A) gene. Eur J Endocrinol. 2021 Mar;184(3):R99-R109. https://doi.org/10.1530/EJE-20-1120