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Fourth Recurrence of Cardiac Myxoma in a Patient with Carney Complex

A 54-year-old male with previously known diagnosis of Carney complex and acromegaly was admitted with exertional dyspnea, chest pain, and palpitation. He had a prominent jaw and painless swelling of temporal part of the right upper eyelid (Figure 1A). Further physical examination revealed a solitary pedunculated cystic mass at the right flank area (Figure 1B and C). Electrocardiography showed sinus rhythm with pathological Q waves in the inferior leads. Transthoracic echocardiography exposed a multilobulated large mass in the left atrium with irregular borders, filling almost the whole chamber and protruding into the mitral valve during diastole (Figure 2A, B and Supplementary Video 1). There was global left ventricular hypokinesia with reduced ejection fraction. We learned that he had

E-PAGE ORIGINAL IMAGE



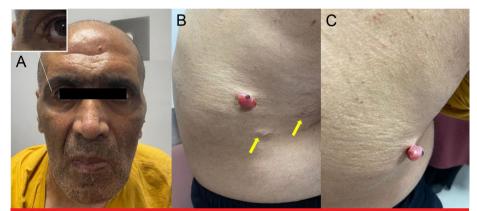


Figure 1. (A): Patient's face with a prominent jaw and a focused image of eyelid swelling. (B): Solitary pedunculated cystic mass compatible with cutaneous manifestation of Carney complex at the right flank area. Arrows show previous excision scars due to recurrent lesions. (C): Posterolateral sight of the mass and its pedicle.



Figure 2. (A): Parasternal long axis view on transthoracic echocardiography, demonstrating a large mass in the left atrium. (B): Apical 4 chambers view on transthoracic echocardiography, demonstrating a large mass in the left atrium, filling almost the whole chamber.



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Damla Yalçınkaya Öner¹ Uğur Nadir Karakulak²

¹Department of Cardiology, Ankara Training and Research Hospital, Ankara,

²Department of Cardiology, Faculty of Medicine, Hacettepe University, Ankara, Turkev

Corresponding author: Damla Yalcınkaya Öner

⊠ damlaykaya@gmail.com

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3 previous heart surgeries, the last of which was 8 years ago, owing to recurrent atrial myxoma. He also had history of right inguinal orchiectomy due to previous Sertoli cell tumor, bilateral adrenalectomy due to primary pigmented nodular adrenocortical disease, transsphenoidal hypophysectomy due to pituitary macroadenoma, and thyroidectomy due to benign thyroid nodule and recurrent excisions for eyelid myxoma. Since he had low attendance to regular clinical follow-ups and low adherence to medication, he had multiple admissions because of cerebrovascular accident and adrenal insufficiency. Emergent reoperation was planned for the patient and successful myxoma resection was performed.

Carney complex is a rare syndrome characterized by cutaneous manifestations, cardiac and extra-cardiac myxomas, and multiple endocrine neoplasms. Here, we report a patient with multiple recurrence of cardiac and cutaneous myxomas who meets almost all of the major criteria.

Informed Consent: Informed consent was obtained from patient's quardian.

Supplementary Video 1: Parasternal long axis view on transthoracic echocardiography, showing a mobile, multilobulated large mass in the left atrium with irregular borders, which is protruding into the mitral valve during diastole.