Myxoma in patients with hypertrophic obstructive cardiomyopathy. Retrospective single-center analysis

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INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a relatively frequent cardiac disease, with a prevalence around 1:500 [1, 2]. Around 70% of HCM patients have either resting or provocable left ventricular outflow tract (LVOT) obstruction, thus presenting as hypertrophic obstructive cardiomyopathy (HOCM). Atrial myxomas are much rarer although the most frequent ones are benign cardiac tumors. The surgical incidence of atrial myxomas is 0.5/million population/year [3], and the approximate prevalence of cardiac tumors in adults is up to 0.2% in some autopsy series [4–6]. Most atrial myxomas are located in the left atrium. Next, patients with coexisting HCM and atrial myxoma (HCM-atrial myxoma) have been described casuistically. Most probably the first HCM-atrial myxoma case was presented by Hiasa et al. [7]. Recently Gi et al. [8] reported a sixth such case and presented a systematic review of this topic. We present two patients with these entities from a retrospective single-center registry.

METHODS

We retrospectively screened the discharge summaries filled in the electronic database from a tertiary high-volume heart center. In the studied period (January 2008 to November 2020), 103 330 patients were hospitalized; among them — 1437 HCM patients. The following keywords (with their grammatical variations and abbreviations) were used to identify HCM and atrial myxoma: "hypertrophic cardiomyopathy", "hypertrophic obstructive cardiomyopathy", and "myxoma".

Statistical analysis

Statistical analysis was limited to the simple calculation of the prevalence of atrial myxoma among all hospitalized HCM patients.

RESULTS AND DISCUSSION

Two female patients with a history of HCM-atrial myxoma were identified.

Patient 1

A 55-year-old HOCM patient with paroxysmal atrial fibrillation (AF), non-sustainable ventricular tachycardia, and after implantation of a cardioverter-defibrillator for primary prevention of sudden cardiac arrest, was admitted due to recurrent AF episodes. Transthoracic echocardiography (TTE) showed asymmetric hypertrophy of the interventricular septum (IVS) up to 26 mm, systolic anterior motion (SAM) of the mitral leaflet, mild mitral regurgitation (MR), and a dynamic LVOT gradient up to 60 mm Hg. The systolic function of both ventricles was preserved. The patient was scheduled for percutaneous treatment of AF by radiofrequency ablation. Pre-interventional work-up by both transesophageal echocardiography (TEE) and cardiac computed tomography (cCT) revealed the inconclusive appearance of a tumor (17 × 11 × 15 mm) attached to the inter-atrial septum (IAS), near the foramen ovale (which was patent), and protruding into the left atrium (Figure 1). The

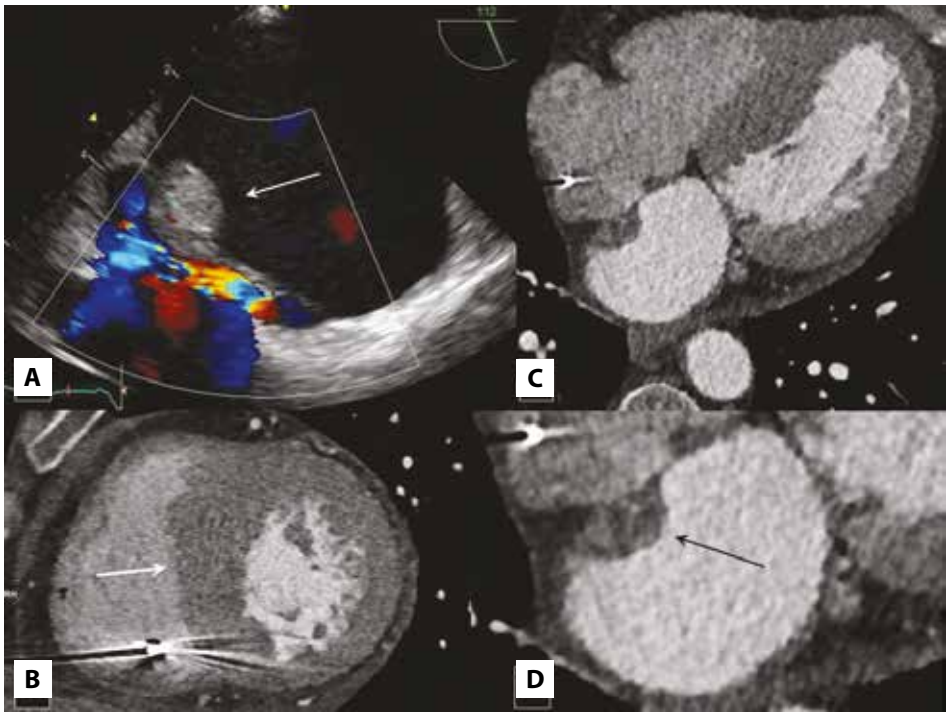

Figure 1. Patient 1. **A.** Transesophageal echocardiography. The white arrow shows a tumor attached to the interatrial septum in the left atrium, near the patent foramen ovale. **B–D.** Computed tomography. **B.** Short axis view. White arrow indicates hypertrophic interventricular septum. **C.** Four-chamber view. **D.** Magnification of panel **C** with focus on the left atrium. White arrow indicates tumor in the left atrium attached to the interatrial septum

patient underwent surgical treatment. The tumor was removed with an adjacent part of the interatrial septum via the right atrial approach. Next, pulmonary vein ostia were isolated with a monopolar electrode. Finally, myectomy of the hypertrophic IVS was done via a trans-aortic approach. Histopathological diagnosis of the removed tumor was myxoma (Supplementary material, *Figure S1*). The postoperative course was uneventful. The pressure gradient across LVOT was 20 mm Hg one year after surgery. cCT done 9 years after resection of atrial myxoma did not reveal any recurrence of the intra-cardiac tumor (Supplementary material, *Figure S2*).

Patient 2

A 50-year-old HOCM patient was admitted for alcohol septal ablation. Pre-interventional TTE showed asymmetric IVS-hypertrophy (20 mm), SAM, and mild-to-moderate MR. The LVOT gradient was 80 mm Hg. The systolic function of both ventricles was preserved. Both TTE and TEE revealed an oval tumor (13 × 9 mm) loosely attached to the IAS in the right atrium (Supplementary material, *Figure S3A*)*. The patient underwent surgical IVS-myectomy, mitral valve replacement, and tumor removal. Again, histopathological examination revealed myxoma*. Six years later, an implantable cardioverter-defibrillator was implanted for primary prevention of sudden cardiac arrest. Repeated TTE nor cCT done during 19 years of clinical follow-up did not reveal recurrence of the intra-cardiac tumor (Supplementary material, *Figure S3B*).

Incidental findings of IAS-attached tumors in these patients changed previously scheduled therapeutic strategy from percutaneous RF-ablation (first patient) and alcohol septal ablation (second patient) to open-heart surgery for both tumor removal and relief of the LVOT obstruction. In the first case, TEE appearance was more suggestive of angioma than myxoma, and cCT appearance was thrombus-like. Both TEE and cCT have good resolution. Nevertheless, both these modalities have their limitations in differentiating intra-cardiac tumors. The role of complementary imaging modalities of cardiac masses is discussed in detail elsewhere [9].

Next, familial myxomas (Carney complex and its subsets LAMB syndrome and NAME syndrome) are inherited via autosomal dominant transmission. In many cases (but not all), the Carney complex is due to mutations of the PRKAR1A gene [10]. HCM, as a genetically heterogeneous disease and familial trait, is seen in approximately 60% of HCM patients. Among the known causal genes, MYH7 and MYBPC3 are the two most common ones, and they are responsible for approximately half of familial HCM — see detailed review elsewhere [11]. Other ultra-rare alterations in the MYBPC3 gene could represent a novel founder pathogenic variant in the Polish HCM cohort [12]. So far, no potential genetic links have been found between familial myxomas and HCM.

* TTE and TEE, as well as histopathology, done in 2002 — only an echocardiographic report is available along with a printout of the TEE image and histopathological findings.

CONCLUSIONS

These two patients add to the very limited literature of HCM coexisting with atrial myxoma. The previous six reports were focused on the casuistic coexistence of these two anomalies. This paper presents the first systematic study of atrial myxomas among a large HCM cohort, providing additional new findings, namely visualization of atrial myxoma in HOCM patients as well as the information on the prevalence of atrial myxoma among the HCM cohort (0.14%). Finally, myxoma may be attached on both sides of the IAS in HCM patients.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

Conflict of interests: None declared.

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