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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 prevalence around 1:500 [1, 2]. Around 70% of HCM-patients have either resting or provocable left ventricle outflow tract (LVOT) obstruction, thus presenting as hypertrophic obstructive cardiomyopathy (HOCM). Atrial myxomas are much rarer finding, although most frequent benign cardiac tumors. The surgical incidence of atrial myxomas is 0.5/million population/year [3] and the approximate incidence 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) are only casuistically reported. Most probably

the first HCM-atrial myxoma case was presented by Hiasa et al. [7]. Recently Gi et al. [8] reported sixth such case, as well as presented 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 tertiary high-volume heart center. In the studied period (January 2008–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 atrial myxoma prevalence among all hospitalized HCM-patients.

RESULTS AND DISCUSSION

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

Patient 1

55-year-old HOCM-patient with paroxysmal atrial fibrillation (AF), non-sustainable ventricular tachycardia and after implantation of implantable 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 dynamic LVOT-gradient up to 60 mm Hg. Systolic function of both ventricles was preserved. The patient was scheduled to percutaneous treatment of AF by radiofrequency ablation. Pre-interventional work-up by both transesophageal echocardiography (TEE) and cardiac computed tomography (cCT) revealed inconclusive appearance of tumor (17 × 11 × 15 mm) attached to the inter-atrial septum (IAS), near foramen ovale (which was patent) and protruding into left atrium (Figure 1). The patient underwent surgical treatment. The tumor was removed with adjacent part of the interatrial septum via the right atrial approach. Next, pulmonary vein ostia were isolated with monopolar electrode. Finally myectomy of the hypertrophic IVS was done via trans-aortic approach. Histopathological diagnosis of the removed tumor was myxoma (Supplementary material, *Figure S1*). Post-operative course was

uneventful. Pressure gradient across LVOT was 20mmHg one year after surgery. cCT done 9 years after resection of atrial myxoma did not reveal any recurrence of intra-cardiac tumor (Supplementary material, *Figure S2*).

Patient 2

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. LVOT-gradient was 80mmHg Systolic function of both ventricles was preserved. Both TTE and TEE revealed oval tumor (13 × 9 mm) loosely attached to IAS in the right atrium (Supplementary material, *Figure S3A*)[*]. Patient underwent surgical IVS-myectomy, mitral valve replacement and tumor removal. Again, histopathological examination revealed myxoma[*]. Six years later 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 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 LVOT relief. 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 complimentary 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), 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 and 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.

CONCLUSIONS

These two patients add to the very limited literature of HCM coexisting with atrial myxoma. Previous six reports were focused on the casuistic coexistence of these two anomalies. This

paper presents the first systematic study of atrial myxomas among large HCM cohort, giving additional findings, not presented before, namely visualization of atrial myxoma in HOCM patient as well as the prevalence of atrial myxoma among HCM cohort (0.14%). Finally, myxoma may be attached on both sides of IAS in HCM-patients.

*TTE and TEE as well as histopathology done in 2002 — available only echocardiographic report along with with printout of TEE image and histopathological finding.

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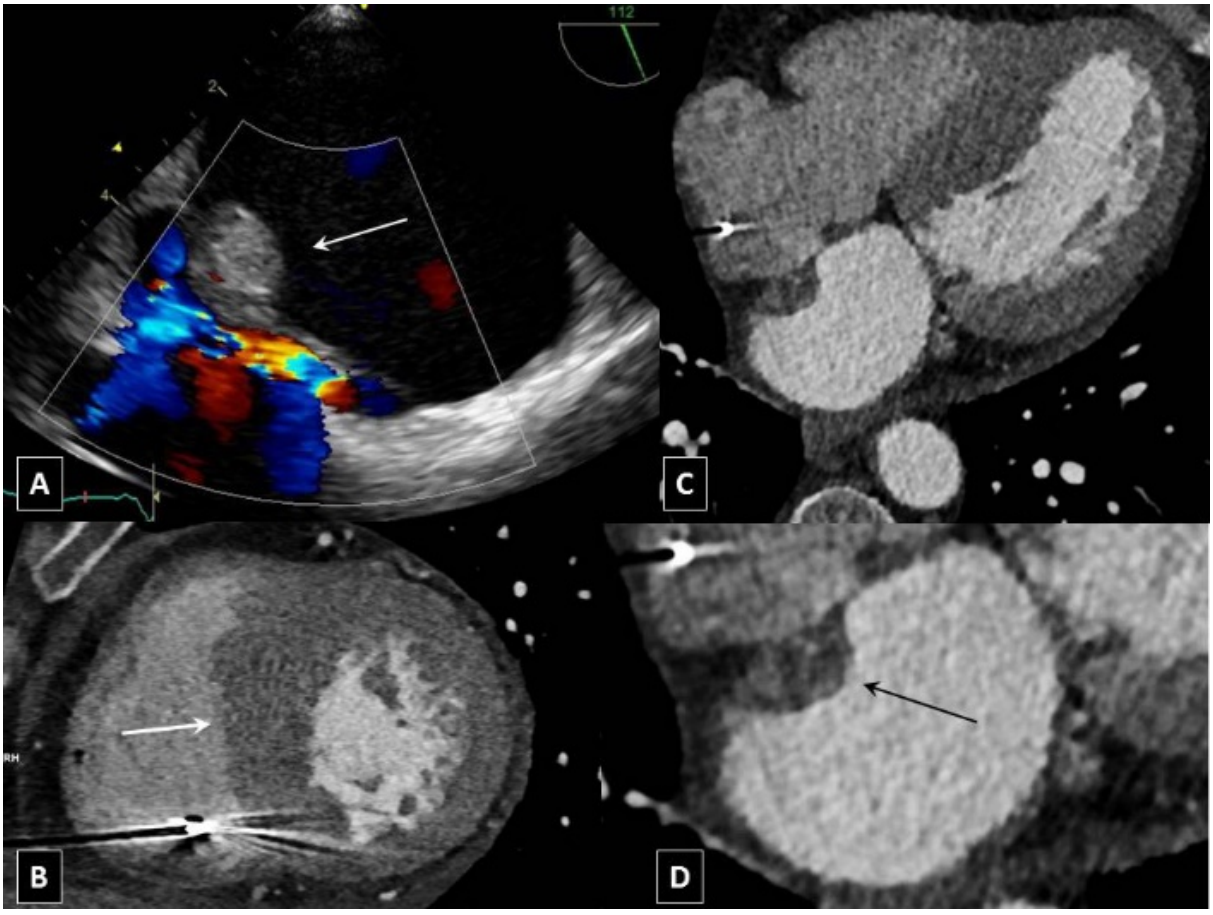

Figure 1. Patient 1. **A.** Transesophageal echocardiography. White arrow shows a tumor attached to the inter-atrial septum in the left atrium, near the patent foramen ovale. **B.–D.** Computed tomography. **B.** Short axis view. White arrow indicates hypertrophic interventricular

septum. **C.** Four-chamber view. **D.** Magnification of the panel “C” with focus on the left atrium. White arrow indicates tumor in the left atrium attached to the interatrial septum