Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

The authors presented a case of a 61 years old male, who was admitted to the Centre for Rare Cardiovascular Diseases with an initial diagnosis of aortic valve stenosis. He complained of mild decrease of exercise capacity. Cardiac echo study revealed LV walls hypertrophy, normal size chambers, dynamic LV outflow tract obstruction (LVOTO) due to mitral anterior leaflet systolic anterior motion (SAM), LVOT gradient of much more than 100mmHg and moderate AV regurgitation. Magnetic resonance confirmed abovementioned findings. His NT-proBNP level was elevated. Holter ECG monitoring recorded paroxysmal AFib, episodes of sinus bradycardia and pauses of max duration of 2,7 seconds. The patient was administered VKA, sotalol 20mg/d, amlodipine 10mg/d, ACEi 10mg/d and statin. The authors are uncertain about the most appropriate further management.

DISCUSSION

Management of HCM patients should include i.a. Assessment of symptoms, laboratory testing (NT-proBNP and high sensitivity cardiac troponin T), ECG, echo, CMR, identification of LVOTO, cardiopulmonary exercise testing, family history and assessment of risk of sudden cardiac death are required in management of HCM patients. In symptomatic patients with LVOTO, the aim of treatment is to improve symptoms by using drugs, surgery, alcohol ablation or pacing.

EXPERT'S OPINION

This is a mildly symptomatic, elderly patient with HOCM. Maximal LVOT gradient is of >100mmHg. High resting LVOT gradient implicates the need for deciding whether he requires septal reduction therapy (SRT). According to the ESC Guidelines (2014) the SRT is not recommended in this patient at this point. Pharmacological treatment should include non-vasodilating betablocker (eg. sotalol) to improve symptoms and to control arrhythmia. ICD implantation is not recommended in this patient based on ESC HCM SCD risk formula – calculated risk of SCD at 5 years is 3,2.

Coronary angiography should be considered in this patient to assess the coronary arteries anatomy together with septal branches. The patient should remain in follow-up with assessment of symptoms, ecg, echo, exercise test and laboratory testing every 6-12 months.

REFERENCES

Elliott PM, Anastasakis A, Borger MA et al. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). Eur Heart J. 2014;35:2733-79.

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