Prikaz slučaja / Case report

Apikalna hipertrofijska kardiomiopatija: prikaz slučaja

Apical hypertrophic cardiomyopathy: a case report

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SAŽETAK: Apikalna hipertrofijska kardiomiopatija (AHK) predstavlja rijetku varijantu hipertrofijske kardiomiopatije koju karakterizira zadebljanje najdistalnijeg dijela stijenke lijevog ventrikula. Prvi put ju je opisao Sakamoto 1976. godine u Japanu. U radu *Yamaguchi i sur.* AHK opisana je kao zapanjujući elektrokardiografski zapis u vidu negativnih T-valova u prekordijalnim odvodima te ventrikulografijom prikazana lijeva klijetka koja podsjeća na "as pika". Iako je ovaj entitet češći u Aziji i čini oko 13% do 25% ukupnih hipertrofijskih kardiomiopatija, također je opisan i u zapadnim zemljama, ali sa znatno manjom prevalencijom. Za dijagnozu AHK koristimo EKG, ehokardiografiju, ventrikulografiju, nuklearnu perfuziju srca i magnetsku rezonanciju srca.

Prikazujemo 58-godišnjeg bolesnika kod kojeg je AHK bila neprepoznata deset godina. Zaključno, i u hrvatskoj populaciji možemo naići na ovu rijetku varijantu hipertrofijske kardiomiopatije, a pravilna interpretacija EKG i ehokardiografije ukazuje na ovaj rijedak entitet.

KLJUČNE RIJEČI: apikalna hipertrofijska kardiomiopatija, abnormalan EKG, ehokardiografija

SUMMARY: Apical hypertrophic cardiomyopathy (AHC) is a rare variant of hypertrophic cardiomyopathy characterized by thickening of the most distal part of the left ventricular (LV) wall. It was first described by Sakamoto in 1976 in Japanese patients. AHC was reported as a striking electrocardiographic pattern of giant negative T-waves and angiographic feature of end diastolic LV cavity configuration resembling an ace of spade by *Yamaguchi et al.* Although AHC is more common in Asia and it accounts for about 13% to 25% of all cases of hypertrophic cardiomyopathy, it is much less prevalent in the western population. Diagnostic modalities include ECG, echocardiography, ventriculography, nuclear myocardial perfusion studies and cardiac magnetic resonance imaging.

We present a 58-year-old man with AHC that was unrecognized for the previous 10 years. In conclusion, this rare disease could be found in the Croatian population too, whereas the appropriate interpretation of the ECG and echocardiography is crucial in recognizing this rare, but important form of hypertrophic cardiomyopathy.

KEYWORDS: Apical hypertrophic cardiomyopathy, abnormal ECG, echocardiography.

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Prikaz slučaja

Pedesetosmogodišnji bolesnik s otprije poznatom arterijskom hipertenzijom i tumorom nadbubrežne žlijezde hospitaliziran je u Kliničkoj bolnici Dubrava, Zagreb zbog bolova u prsima koji su imponirali kao stenokardija te osjećaja nepravilnog srčanog rada. Obiteljska anamneza je bila negativna s obzirom na iznenadnu srčanu smrt, kardiomiopatije ili srčano zatajivanje. Višeslojnom kompjuteriziranom tomografijom bubrega i nadbubrežnih žlijezda verificirao se hormonski neaktivan adenom lijeve nadbubrežne žlijezde.

Fizikalni nalaz je bio u granicama normale, kao i srčani biomarkeri te Rtg srca i pluća.

Dominanatan nalaz u EKG-u su bili duboko negativni T-valovi u prekordijskim odvodima (**Slika 1**) koji su se pratili untrag 10 godina zbog čega je bila učinjena i koronarografija koja je bila uredna. U ranijim nalazima transtorakalne ehokardiografije (TTE) opisivana je koncentrična hipertrofijska kar-

Case report

A 58-year-old male with a history of hypertension and tumor of suprarenal gland was referred to Dubrava University Hospital, Zagreb because of the chest pain and palpitation. There was no family history of sudden cardiac death, congestive heart failure or cardiomyopathy. We evaluated the patient with the MSCT of the kidney and suprarenal glands because of the previously suspected tumor of suprarenal gland that was characterized as adenoma.

The results of physical examination was normal, as well as the cardiac biomarkers and chest X-ray.

The dominant finding was an abnormal ECG, which was characterized primarily by deep negative T-waves in precordial leads (**Figure 1**) which were followed-up throughout the last 10 years' period, which is why coronarography was performed showing normal values. Because of that finding, which was also noticed about ten years ago, the left-sided cardiac catheterization was done, with no evidence of coro-

diomiopatija uz urednu morfologiju i funkciju valvularnog aparata.

nary artery disease. Transthoracic echocardiography (TTE) showed concentric hypertrophic cardiomyopathy (HC) of the left ventricle with no valvulopathy.

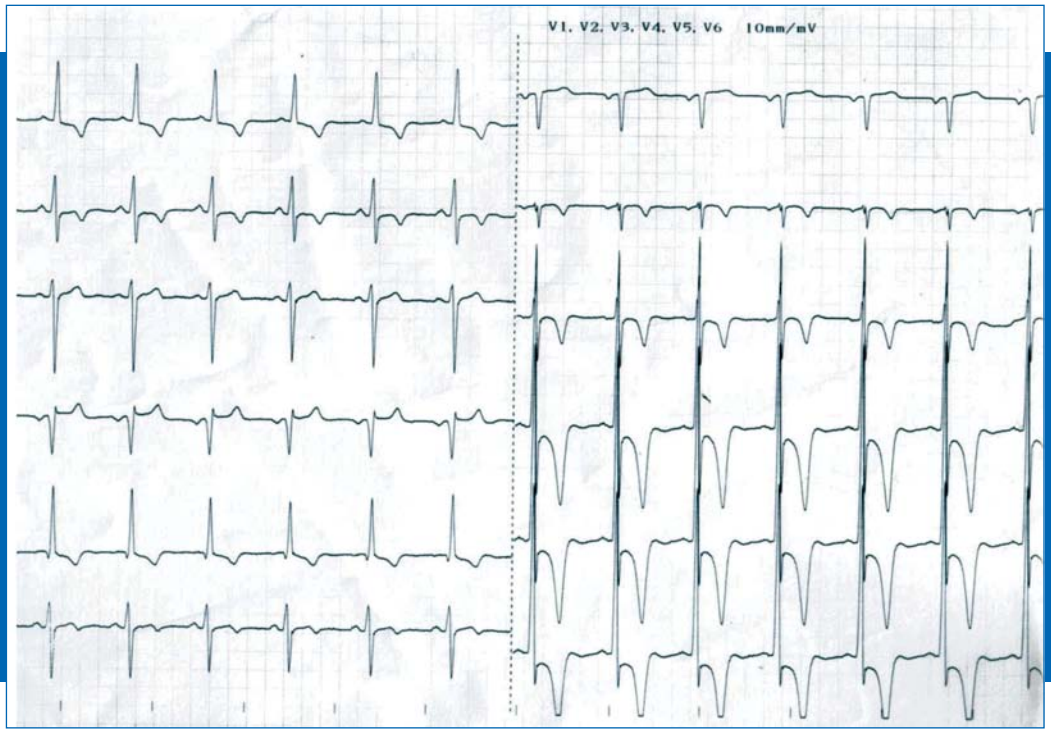

Figure 1.
Abnormal 12-lead ECG.
Deep negative T-waves in precordial leads.

Sada se učinjenim TTE odmah po prijemu utvrdi apikalna hipertrofija lijeve klijetke s debljinom apeksa 43 mm u sistoli i 35 mm u dijastoli te omjerom debljine apeksa i septuma 1,6 (**Slike 2 i 3**). 24-satni Holter EKG nije utvrdio patološke atrijske ili ventrikulske poremećaje ritma.

TTE performed immediately after admission showed apical hypertrophy of the left ventricle with the systolic apex thickness of 43 mm, diastolic apex thickness of 35 mm, and the ratio of maximal apical to posterior wall thickness of 1.6 (**Figure 2, 3**). The follow-up 24-hour Holter ECG monitoring revealed no atrial or ventricular ectopy.

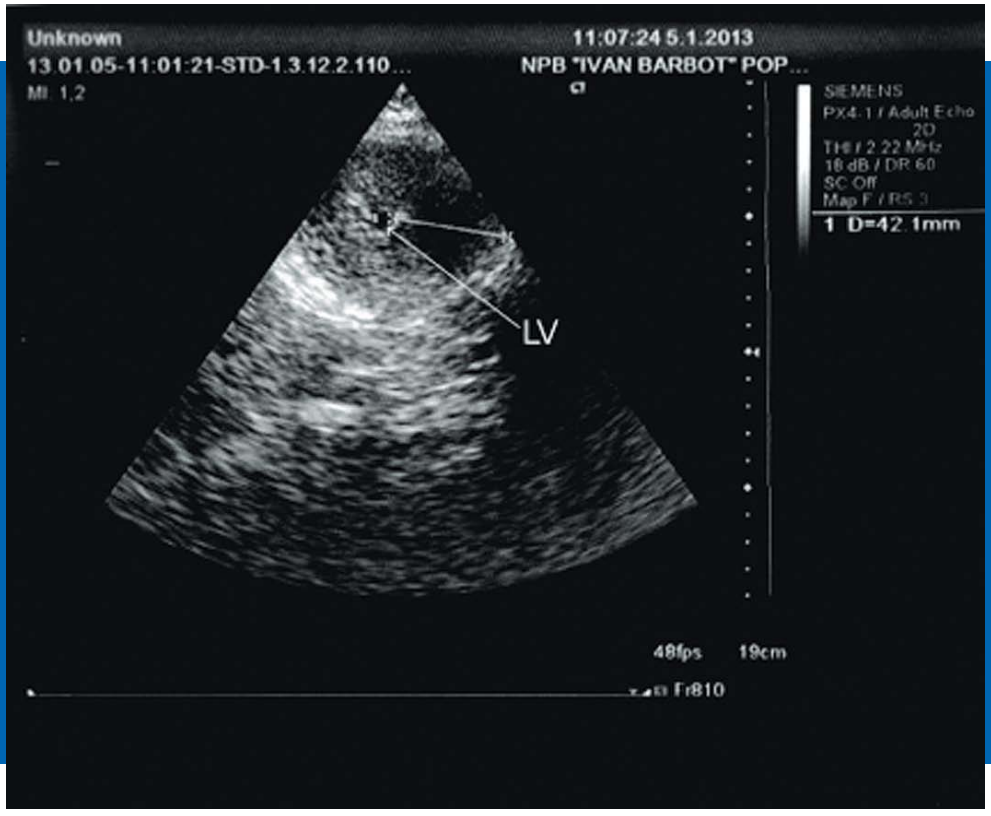

Figure 2.
Transthoracic echocardiography showing left ventricular apical hypertrophy with systolic apex thickness of 42 mm.

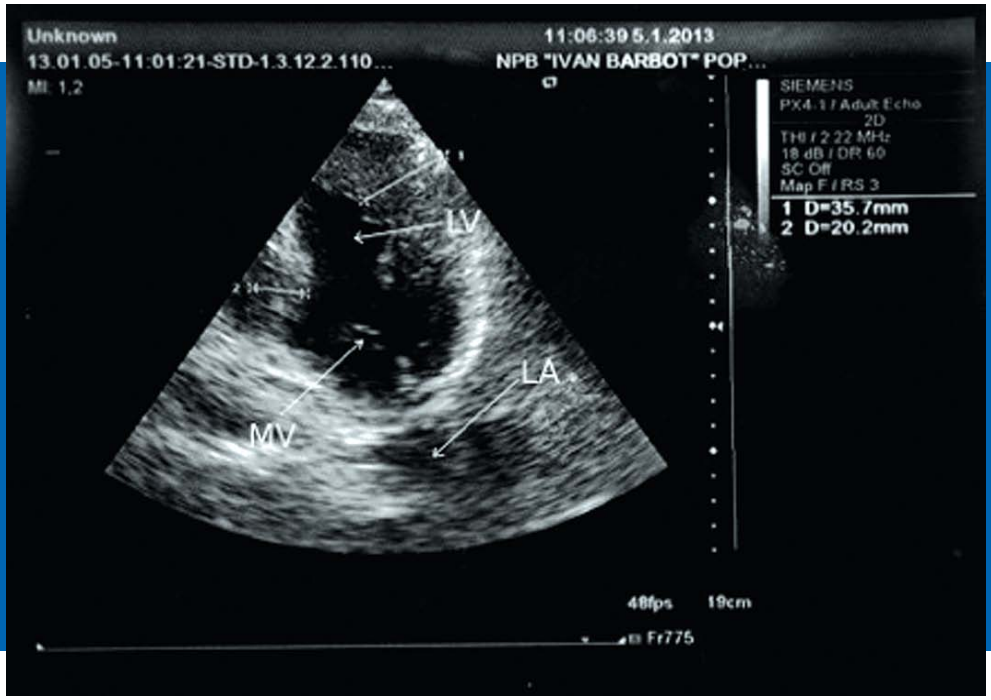

Figure 3.
Transthoracic echocardiography showing left ventricular apical hypertrophy with dyastolic apex thickness of 35 mm.

Bolesniku je dijagnosticirana apikalna hipertrofijska kardiomiopatija (AHK) te je pored terapije ACE inhibitorima uveden beta-blokator.

Diskusija i zaključak

Iako je AHK rijetka bolest, u nekim radovima opisano je autosomno dominantan oblik nasljeđivanja.[1] Obiteljsko nasljeđivanje je češće u asimetričnoj septalnoj hipertrofiji nego u AHK.

Dijagnostički kriteriji za AHK uključuju: asimetričnu hipertrofiju lijeve klijetke koja predominantno zahvaća apikalnu i stražnju stijenku lijevog ventrikula s debljinom apeksa ≥15 mm i omjerom debljine apikalne i stražnje stijenke ≥1,5, s obzirom na nalaz TTE ili MRI. Bolest je češća u muškaraca, a srednja životna dob u kojima se najčešće pojavljuje je 41,1±14,5 godina. Oko 54% bolesnika s AHK imaju simptome, a najčešće su to bol u prsima, palpitacije, dispneja i sinkopa.[2]

Najčešći nalaz u 12-kanalnom EKG predstavljaju negativni T-valovi u prekordijalnim odvodima koji se mogu registrirati u 93% bolesnika te hipertrofija lijeve klijetke koju nalazimo u 65% bolesnika. TTE pokazuje hipertrofiju apeksa lijeve klijetke i predstavlja prvu stepenicu u dijagnozi apikalne hipertrofije. Međutim, u slučaju da ovom metodom ne dobijemo konačnu dijagnozu, MRI postaje zlatni standard za dijagnozu ovog entiteta.[3] Apikalni tumori, trombi u lijevoj klijetki kao i endomiokardijalna fibroza te koronarna bolest srca mogu imitirati ovu bolest.[4-7]

Bolesnici s apikalnom hipertrofijom mogu biti simptomatski i asimptomatski. Verapamil, beta-blokatori i antiaritmici mogu se koristiti u liječenju simptomatskih bolesnika.[2] Verapamil i beta-blokatori djeluju na simptome bolesti dok se amiodaron i prokainamid upotrebljavaju za liječenje atrijske fibrilacije i ventrikulskih aritmija.[8,9] Ugradbeni kardioverter defibrilator se preporuča u visoko rizičnih bolesnika s anamnezom prethodnog kardijalnog aresta ili postojanih epizoda ventrikularne tahikardije, sinkope ili obiteljske anamneze iznenadne srčane smrti.[10]

The patient was diagnosed with benign form of apical HC and received treatment with ACE inhibitors and beta-blockers.

Discussion and Conclusion

Apical HC is frequently sporadic; however, a few families have been reported with autosomal dominant inheritance.[1] Positive family history is more common in patients with asymetric septal hypertrophy than with apical HC.

The diagnostic criteria for apical HC include demonstration of asymmetric LV hypertrophy, confined predominantly to the LV apex, with apical thickness ≥15 mm and ratio of maximal apical to posterior wall thickness ≥1.5, based on an TTE or magnetic resonance imaging (MRI). The mean age of presentation of apical HC is 41.1±14.5 years and it is most commonly seen in males. About 54% of patients with apical HC are symptomatic and the most common presenting symptom is the chest pain, followed by palpitations, dyspnea and syncope.[2]

The most frequent ECG findings are negative T-waves in precordial leads which are found in 93% of patients, followed by LV hypertrophy in 65% of patients. TTE shows hypertrophy of LV apex and is the initial diagnostic tool for apical HC. Although the TTE is most commonly the initial diagnostic tool, if inconclusive, cardiac MRI is now emerging as "the gold standard" for diagnosis of apical HC.[3] Apical HC can mimic other conditions, including apical cardiac tumors, LV apical thrombus, endomyocardial fibrosis and coronary artery disease.[4-7]

Patients with apical HC can be symptomatic and asymptomatic. Verapamil, beta-blockers and anti-arrhythmic agents are used in symptomatic patients.[2] Verapamil and beta-blockers are found to be beneficial in improving symptoms, while amiodaron and procainamid are used in the treatment of atrial fibrillation and ventricular arrhythmias.[8,9] Implantable cardioverter defibrilator is recommended for high risk HC patients with previous cardiac arrest or sustained episodes

Za razliku od drugih varijanti hipertrofijskih kardiomiopatija, prognoza AHK je relativno benignija. Ukupna smrtnost iznosi 10,5%, a kardiovaskularni mortalitet je 1,9% nakon praćenja od 13.6±8,3 godina.[2] Iako je rizik od iznenadne srčane smrti i kardiovaskularnih događaja češći u asimetričnoj septalnoj hipertrofiji nego u apikalnoj, bolesnici s apikalnom hipertrofijom također mogu razviti iznenadne, po život opasne komplikacije.

U zaključku naglašavamo da je bitno ne previdjeti dijagnozu ove forme hipertrofijske kardiomiopatije te iako je prognoza kod ovog oblika hipertrofijske kardiomiopatije znatno povoljnija nego kod drugih, potrebno je redovito pratiti i adekvatno liječiti ove bolesnike.

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of ventricular tachycardia, syncope or a family history of sudden cardiac death.[10]

Unlike some other variants of hypertrophic cardiomyopathy, the prognosis of apical HC is relatively benign. The overall mortality rate is 10.5% and cardiovascular mortality is 1.9% after follow-up of 13.6±8.3 years.[2] Although sudden cardiac death and cardiovascular events occur more frequently in asymmetric septal hypertrophy than in the apical one, those patients may also develop sudden life-threatening complications.

In conclusion, it is important not to overlook the diagnosis of this form of HC; and although the prognosis in this form of HC is much better these patients are to be regularly followed-up and properly treated.

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