2010;5(5-6):66. KARDIO LIST

Prikaz slučaja | Case report

Idiopatska dilatacija desnog atrija praćena paralizom atrija i cerebrovaskularnim inzultom

Idiopathic dilatation of right atrium with atrial standstill and stroke

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SAŽETAK: *Prikazujemo slučaj bolesnika u dobi od 26 godina u kojega je dijagnoza idiopatske dilatacije desnog atrija postavljena transtorakalnom ehokardiografijom te nakon što su isključeni svi ostali entiteti koji dovode do sekundarne dilatacije desnog atrija. Nakon asimptomatskog razdoblja godinu dana kasnije bolesnik je doživio ishemijski cerebrovaskularni inzult, vjerojatno uzrokovan embolusom iz lijevog atrija nastalog uslijed paralize atrijske muskulature. Iako se radi o rijetkom poremećaju, idiopatsku dilataciju desnog atrija treba diferencijalno dijagnostički uvijek uzeti u obzir kod obrade uvećanog desnog atrija.*

KLJUČNE RIJEČI: *desni atrij, idiopatska dilatacija desnog atrija, cerebrovaskularni inzult.*

ABSTRACT: *We report a case of a 26 years old man that who was diagnosed with idiopathic right atrial dilatation. The diagnosis was made by transthoracic echocardiography and after all other lesions known to produce the dilatation of right atrium have been excluded. After an asymptomatic period he suffered an ischemic stroke one year later, probably caused by an embolus from the left atrium secondary to the atrial paralysis. Idiopathic dilatation of the right atrium, although a rare disorder, should not be forgotten as differential diagnosis for enlarged right atrium.*

KEYWORDS: *right atrium, idiopathic dilatation of right atrium, stroke.*

CITATION: *Kardio list. 2010;5(5-6):66-69.*

Idiopatska dilatacija desnog atrija rijetka je anomalija, koja može biti otkrivena u bilo kojoj dobi, od fetalne do odrasle dobi. Zbog svoje rijetkosti, vrlo lako biva zamijenjena s drugim, puno češćim, anomalijama koje dovode do sekundarne dilatacije desnog atrija, kao što su Ebsteinova anomalija te kardiomiopatije desne klijetke[1]. Ovdje donosimo prikaz mladog bolesnika s idiopatskom dilatacijom desnog atrija i paralizom atrijske muskulature te preboljenim ishemijskim cerebrovaskularnim inzultom.

Prikaz slučaja

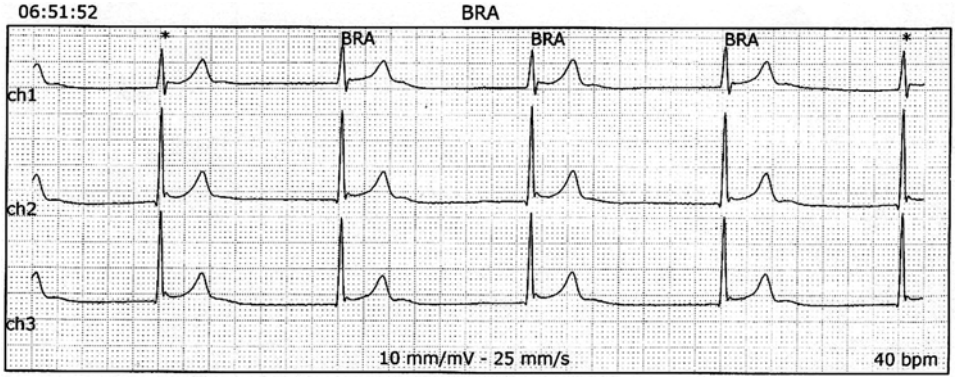
Mladi bolesnik u dobi od 26 godina hospitaliziran je na našem Zavodu radi obrade opetovanih sinkopa u naporu. Obradom je nađen trajni nodalni ritam, bez znakova atrijske električne aktivnosti, s dobrom kronotropnom rezervom (**Slika 1**). Učinjeni su transtorakalni i transezofagusni ultrazvuk srca s kontrastom koji su pokazali jako povećani desni atrij (površine 47 cm^2) s odsutnošću mehaničke ak-

Idiopathic dilatation of right atrium is a very rare anomaly. It can be discovered at any time from fetal to adult age. Since it is rare, it can easily be mistaken for some other, more frequent anomaly, which can produce secondary right atrial dilatation, like Ebstein's anomaly or a right ventricular cardiomyopathy[1]. Here we present a case of a young patient with right atrial dilatation presenting with atrial standstill and ischemic cerebral infarction.

Case report

This 26 year old patient was referred to our institution for evaluation of repeated syncopes during effort. Other than this he had no significant previous medical history. Electrocardiogram Holter monitor showed junctional rhythm without any any visible P waves and with good chronotropic reserve (**Figure 1**). Transthoracic and transesophageal echocardiogram with contrast revealed a very large right atrium (area 47 cm^2) and atrial standstill. Tricu-

Figure 1. *ECG of the patient, junctional rhythm without P waves.*



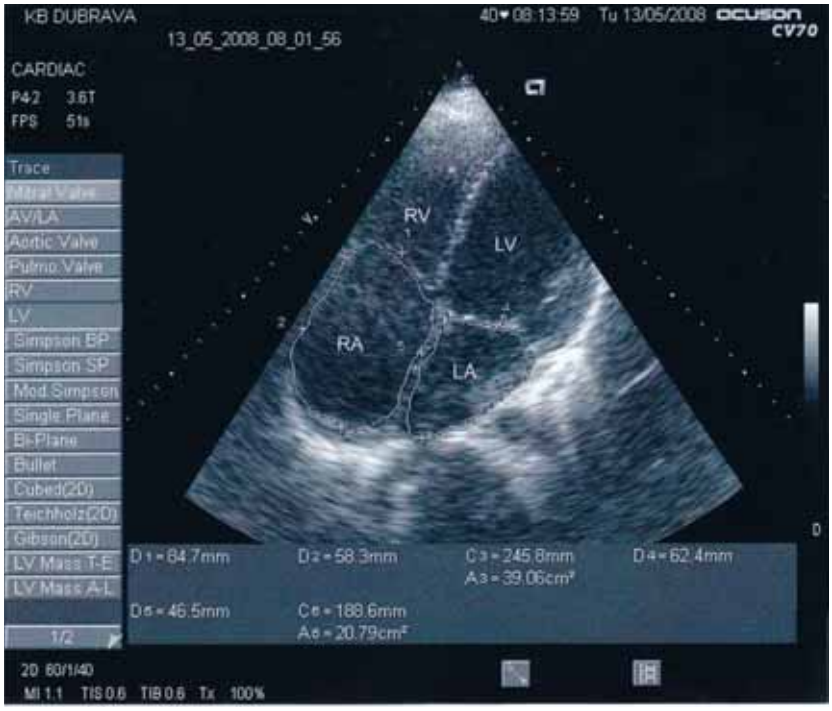


Figure 2. *Transthoracic echocardiogram of the patient: enlarged right atrium area 47 cm².*

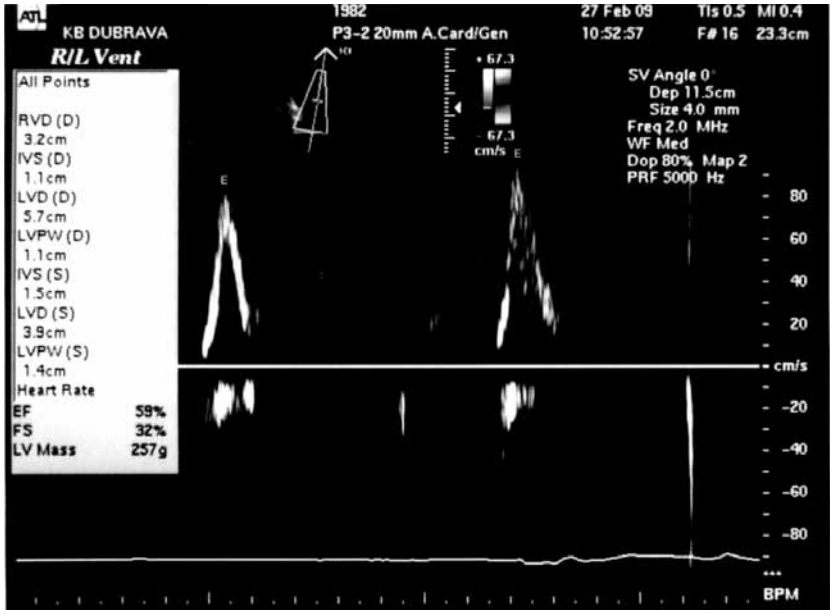


Figure 3. PW-Doppler: flow through mitral aperture is only registered in early diastole.

tivnosti oba atrija i granično široke lijevu i desnu klijetku jako dobre kontraktilnosti (**Slike 2** i **3**). Trikuspidni zalistak je bio uredne morfologije uz trivijalnu regurgitaciju. Nije nađeno elemenata Ebsteinove anomalije: nije bilo pomaka trikuspidalnih zalistaka prema apeksu kao niti odstupanja od njihove normalne morfologije, dok je desna klijetka bila uredne veličine. Uredna debljina i morfologija stijenke desne klijetke, kao i odsutnost njezine akinezije i segmentalne hipokinezije nije dalo uporišta za sumnju na Uhlovu anomaliju ili aritmogenu displaziju desne klijetke. Također treba napomenuti da tijekom telemetrijskog monitoriranja nije zabilježena nikakva ventrikularna ektopijska aktivnost podrijetlom iz desne klijetke kao niti ventrikularna tahikardije podrijetlom iz desnog izgonskog trakta, inače tipične za ove dvije anomalije. Kontrastnom ehokardiografijom isto tako nije nađeno znakova za patološki spoj na razini ve-

spid valve was having normal morphology with only trivial regurgitation. There were no features suggesting Ebstein's anomaly (**Figures 2** and **3**): there was no downward displacement of the tricuspid valve into the right ventricle, no morphological changes of tricuspid valves while the size of the right ventricle was normal. Normal thickness and morphology of the right ventricular wall with absence of its akinesia or segmental hypokinesia provided no evidence for Uhl's anomaly or arrhythmogenic right ventricular dysplasia. It should also be mentioned that during telemetric monitoring no ventricular ectopy originating from right ventricle, and in particular no right ventricular outflow tract tachycardia typical for these two anomalies, were recorded. Contrast echocardiography could not demonstrate any evidence for shunting at the level of large vessels, cardiac cavities or lungs. Systolic pressure in the right ventri-

likih žila, srčanih šupljina ili pluća. Sistolički tlak u desnoj klijetki je iznosio 25 mmHg. Isključivši time sve entitete koji bi mogli imati za posljedicu sekundarnu dilataciju desnog atrija, zaključeno je da se radi o idiopatskoj dilataciji desnog atrija. Nalaz je potvrđen i višeslojnom kompjutoriziramom tomografijom s kontrastom (**Slika 4**). Bolesnik je otpušten uz preporuku redovitih kardioloških kontrola, izbjegavanja napora i uzimanja antikoagulantne terapije, čega se nije pridržavao. Osam mjeseci kasnije pacijent biva hospitaliziran na neurološkom odjelu druge ustanove zbog ishemijskog cerebrovaskularnog inzulta u području lijeve arterije cerebri medie uz desnostranu hemiparezu i motoričku disfaziju. Tada je učinjena kardiološka obrada kojom nije verificiran trombotički proces u lijevom atriju te je ponovno isključen atrijski septalni defekt i otvoreni foramen ovale. Dva tjedna po otpustu bolesnik je ponovno hospitaliziran u našoj ustanovi zbog opetovanih sinkopa. U 24-satnom holteru EKG tada je ponovno verificirana odsutnost atrijske aktivnosti uz osnovni nodalni ritam, no zabilježena je i asistolna pauza od 3,8 sekundi radi čega je indicirana ugradnja trajnog elektrostimulatora srca, što je bolesnik odbio te je otpušten uz preporuku antikoagulacijske terapije.

cle measured 25 mmHg. Having excluded all other entities known to produce secondary dilatation of the right atrium, it was concluded that patient has idiopathic dilatation of the right atrium. Echocardiographic findings were also confirmed by multislice computed tomography with contrast (**Figure 4**). The patient was discharged with recommendations to come for regular follow-ups, and take varfarine in therapy and avoid any efforts. Regretably, he failed to comply with this medical advice. Eight months later he was referred to a stroke unit of another institution for left middle cerebral artery territory ischemic stroke accompanied by right sided hemiparesis and motor dysphasia. Cardiological diagnostic work up failed to detect any thrombotic mass in the left atrium, and no patent foramen ovale or atrial septal defect could be demonstrated. Two weeks following the discharge, he was again referred to our institution due to repeated syncopes. The Holter monitor revealed again junctional bradycardia with atrial standstill, only this time also 3.8 seconds long asystolic pauses were recorded. Therefore he was advised to undergo permanent cardiac electrostimulator implantatnion which he declined, and was discharged with anticoagulation therapy.

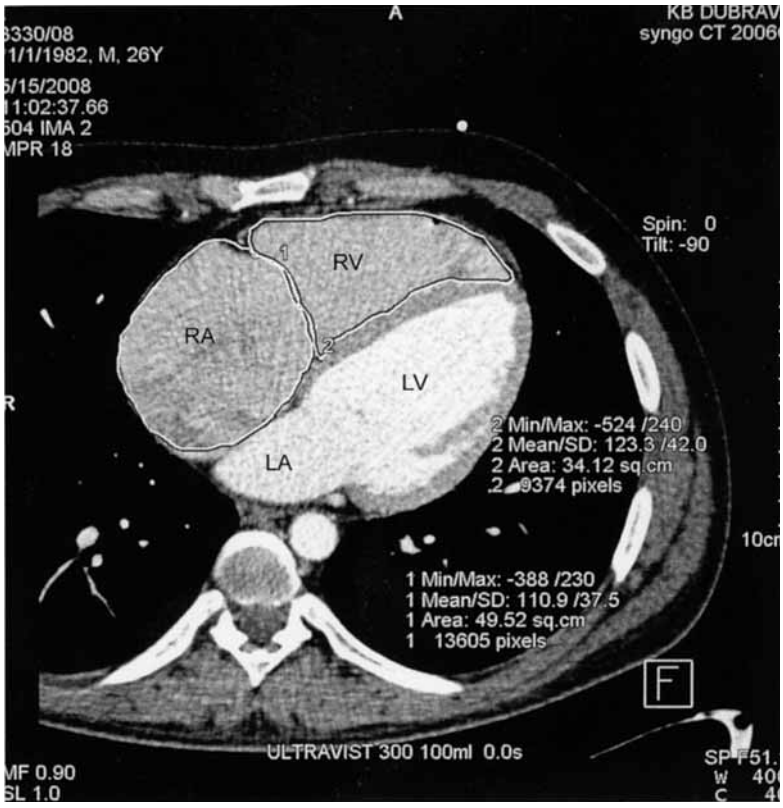


Figure 4. *MSCT of the thorax, also confirming right atrial dilatation.*

Rasprava

Idiopatska dilatacija desnog atrija vrlo je rijetka anomalija, opisana je i obiteljska sklonost[2]. U pravilu je bolesnici dobro podnose, no mogu nastati i komplikacije[3]. Paraliza atrijske muskulature je jedna od njih, a karakterizirana je odsutnošću atrijske atkivnosti na standardnom i intrakardijalnom elektrokardiogramu te gubitkom mehaničke atrijske aktivnosti. Paraliza atrija može uzrokovati sinkope, cerebrovaskularne incidente i srčano zatajivanje. Aktivacija srca u odsutnosti atrijske aktivnosti ovisi o spojnom ritmu.

Discussion

Idiopathic dilatation of the right atrium is a very rare anomaly, familial occurrence has also been reported[2]. Usually it is very well tolerated, although complications may occur[3]. Atrial standstill is one of them, and it is characterized by lack of atrial activity on electrocardiogram with absence of atrial mechanical activity. Atrial standstill may cause syncope, cerebrovascual accidents and heart failure. Cardiac activation depends then on a junctional escape rhythm.

Liječenje asimptomatske idiopatske dilatacije desnog atrija je medikamentozno, dok je kirurški pristup rezerviran za bolesnike sa simptomima. U bolesnika s paralizom atrija i nestabilnim nodalnim ritmom, preporuča se implantacija VVI ili VVIR elektrostimulatora uz antikoagulacijsku terapiju.

U ovom slučaju posebnost je idiopatska dilatacija desnog atrija praćena paralizom atrija te cerebrovaskularnim inzultom, do sada je u literaturi opisan samo jedan ovakav slučaj[1]. Uzrok moždanog udara vjerojatno je bila embolizacija tromba podrijetlom iz lijevog atrija uslijed njegove paralize. Za adekvatnu terapiju neophodna je točna dijagnoza. Okosnicu dijagnostike predstavljaju slikovne metodu, u prvom redu transtorakalna i transezofagusna ehokardiografija.

Zaključno, idiopatska dilatacija atrija, iako rijedak poremećaj, ne smije se diferencijalno dijagnostički zanemariti kod obrade proširenog desnog atrija. Dijagnoza se postavlja metodom isključivanja češćih anomalija srca, poput Ebsteinove anomalije, koje mogu uzrokovati sekundarnu dilataciju desnog atrija, a obzirom na opisanu obiteljsku učestalost preporuča se i probir članova obitelji.

Received: 8th Mar 2010

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Treatment of asymptomatic idiopathic dilatation of right atrium is medical, while surgical procedures are reserved for symptomatic patients. In those patients who have atrial standstill and unstable junctional rhythm, implantation of VVI or VVIR mode cardiac pacemaker with anticogulation therapy is recommended.

In our case the peculiarity is the association of atrial standstill with idiopathic dilatation of right atrium along with a stroke, to our knowledge only one such case has been reported so far[1]. In this case the cause of the stroke is most probably an embolus from the left atrium secondary to the atrial paralysis.

To conclude, accurate diagnosis is essential for adequate treatment. The diagnostic mainstays are imaging methods, most importantly the transthoracic and transesophageal echocardiography. Idiopathic dilatation of the right atrium, although a rare disorder, should not be forgotten as differential diagnosis for enlarged right atrium. Diagnosis is made by systematic exclusion of other, more frequent cardiac anomalies, known to produce secondary right atrial enlargement, most importantly, the Ebstein anomaly, and due to reported familial occurence the screening of relatives is recommended.

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