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Ventricular tachicardia as a form of presentation of isolated noncompaction of left ventricular myocardium

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ABSTRACT

Isolated ventricular noncompaction left ventricular (LVNC) is a rare genetic disease characterized by left ventricular hypertrabeculation. Clinically manifested as systolic dysfunction, heart failure, ventricular arrhythmias, thromboembolic events and sudden death. It can occur alone or in combination with other congenital heart disease and / or neuromuscular disorders. It is diagnosed by echocardiography and magnetic resonance imaging and has a poor prognosis with a mortality rate of 60-80% at 6 years, half of sudden death (SD). We present a case of LVNC which debuted as ventricular tachycardia.

KEYWORDS

Noncompaction cardiomyopathy;
Ventricular tachycardia.

INTRODUCTION

Isolated noncompaction of left ventricular myocardium (LVNC) is a rare genetic disorder which represents an alteration of the morphogenetic endocardial due to an interruption in the myocardial compaction which occurs in very early stages of embryonic development; it consists of a left ventricular hypertrabeculation (VI) with deep intertrabecular recesses communicating with the ventricular cavity directly but not coronary circulation[1-3].

Clinically manifested as systolic dysfunction, heart failure, ventricular arrhythmias, thromboembolic events and sudden death[4-6]. It can occur alone or in combination with other congenital heart diseases and / or neuromuscular disorders[7,8].

The complementary tests for diagnosis are echocardiography and magnetic resonance imaging (MRI). It has a poor prognosis with a mortality of 60-80% at 6 years[9,10], half of sudden death (SD)[11].

We present a case of LVNC which debuted as ventricular tachycardia (VT).

CASE REPORT

49 year-old male with a history of dyslipidemia and schizophrenia treated with olanzapine 10mg/24 h. Current smoker of 3-4 cigarettes / day with a cumulative

consumption of 46 pack-years.

Finding himself previously well, he presented, in connection with moderate effort, self-limited syncope associated with feelings of instability, thoracic dysesthesias and important vegetative signs.

During the physical examination gravity of the patient was impressive, agitated, dyspneic with undetectable pulse and blood pressure. Electrocardiogram (ECG) is performed (Figure 1) in which VT is appreciated. After sedation with midazolam a 100 J cardioversion is applied which results in the recovery of sinus rhythm (Figure 2) with a general decline in the ST up to 7mm (except in aVR lead) with marked right precordial R waves and frequent ventricular extrasystoles indicating a subcutaneous enoxaparin 80mg, 500mg lysine acetylsalicylate iv and 300 mg of amiodarone in serum glucose to happen in 30 minutes. During transfer to hospital (40 minutes) progressive normalization of the ST segment, persisting T negative waves in the inferior and lateral leads (Figure 3). He was admitted in the ICU with a diagnosis of acute coronary syndrome (ACS).

Investigations. – Chest X-ray: normal. Ultrasensitive troponin T 2515 ng/ml. Coronary angiography: normal. In the cardiology ward, the echocardiogram and cardiac MRI (Figures 4-6) observed slightly dilated left ventricle with lateral hypertrabeculation, severe systolic dysfunction (ejection fraction (EF) 0.27), areas of fibrosis in the inferolateral face, anterior papillary muscle and the inferior interventricular junction, confirming the diagnosis of LVNC with areas of fibrosis.

During admission a cardioverter defibrillator (ICD) was implanted. At discharge treatment with acenocoumarol, carvedilol 6.25mg/12h, enalapril 2.5mg/12h, atorvastatin 40mg/24h, lansoprazole 30mg/24h and torasemide5mg/24h.

DISCUSSION

Cardiomyopathies are heart muscle diseases in which alterations in the structure and function of the myocardium in the absence of coronary artery disease, hypertension, valvular heart disease or congenital heart defects, which can give an explaination[1]. The LVNC or spongiform cardiomyopathy[10] is classified by the American Heart Association[12,13] as a primary genetic cardiomyopathy (TABLE 1), meaning those confined

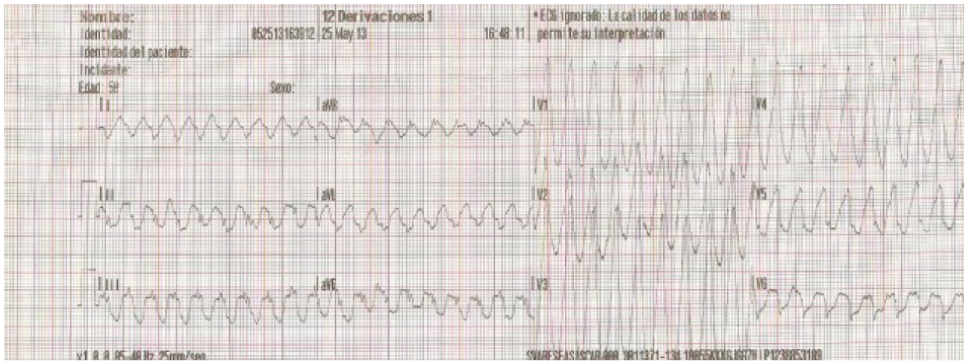

Figure 1 : Initial ECG. Ventricular tachycardia

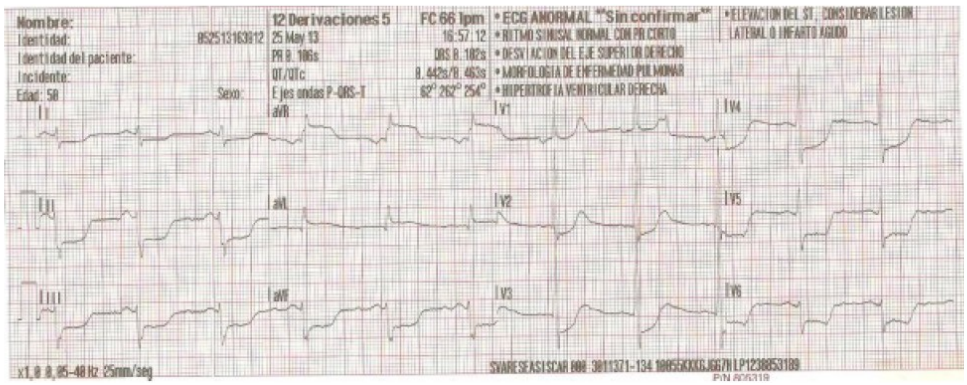

Figure 2 : ECG. Sinus rhythm, ST descent in the lateral and inferior leads, tall R waves in right precordial. Compatible with a subendocardial infarction

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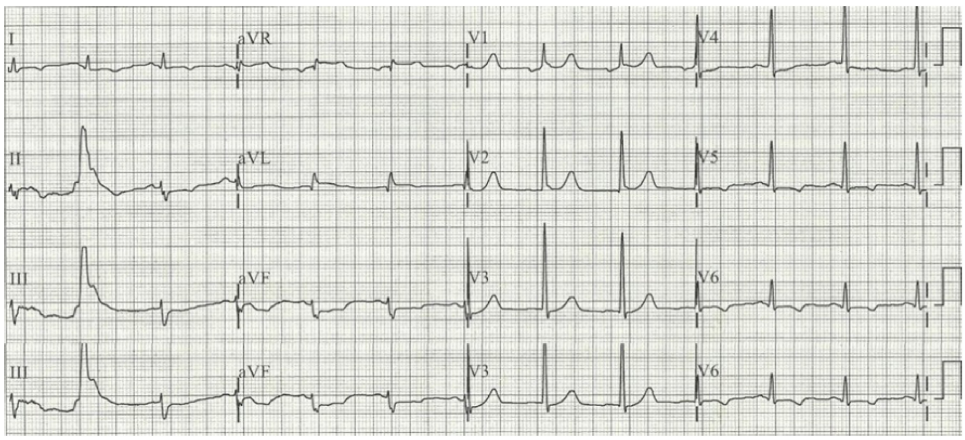

Figure 3 : ECG in hospital. Sinus rhythm, anterior hemiblock, nonspecific repolarization abnormalities in the inferior and lateral face. Negative T-waves in the high lateral face. Tall R-waves in right precordial.

almost exclusively to the heart muscle while the in the secondary´s we see multiple organs effected.

The first anatomical descriptions of LVNC date back to 1932 and they were always associated with complex cyanotic malformations, obstructions in the left ventricular outflow or coronary abnormalities[6,11]. The isolated LVNC was described by Chin in 1990 and is characterized by the absence of other cardiac anomalys because the intertrabecular spaces communicate with the ventricular cavity but not the coronary circulation[6,11,14].

The LVNC is an extremely rare condition, with an estimated adult prevalence in the general population of 0.05% and 0.01%, in the pediatric population[15,16] with an incidence of less than 0.1 per 100,000 in this group[2]. In children it is the third cause of all cardiomyopathies (9.5%), behind the dilated and hypertrophic ones[10,17,18] with an age at diagnosis of 3 months compared to 20-40 years in the adult[11]. In all series more men are affected (60-80% of cases), although the extent of trabeculation is higher in women and patients of African descent[8,11,17,19].

There has been described sporadic and familiar forms of LVNC, the latter representing between 20-50% of all cases[16] are more common in adults[17]. In infants it can be associated (up to a third) with a facial dimorphism[17] (prominent forehead, low-set ears and strabismus). Up to 80 % of adults have neuromuscular disorders (metabolic myopathy, optic neuropathy, muscular dystrophy, muscle enzyme abnormalities and / or abnormal electromyogram[6,8,17,20]), which are uncommon in children[7]. In sporadic forms we find mutations in the mitochondria, and tafazzin Z line. This last mutation, linked to the X chromosome and almost exclusively of childhood, is associated with dilated cardiomyopathy, LVNC, fibroelastosis and Barth syndrome (skeletal myopathy, recurrent neutropenia, growth retardation and aciduria with a low life expectancy)[10,16,17,19]. The LVNC in adults is a genetically distinct disease, transmitted as an autosomal dominant without extracardiac manifestations[4,10,20]. It is possible to identify affected relatives in over 25% of patients[4,6,19], so it is mandatory to perform echocardiography (6) for at least the first-degree relatives[21]. They usually are most often asymptomatic and have better prognosis[4,10,22].

The most likely mechanism responsible for the detention in the fetal myocardial compaction process is mediated by genetic mutations[7]. During early embryonic development, the myocardium is a loose network of interwoven fibers, which are separated by deep recesses which join together the myocardial with the left ventricular cavity[7] to facilitate myocardial oxygenation, because at this stage the coronary circulation has not yet developed[20]. Compaction of this spongy meshwork occurs between the 5th and 8th week of embryonic life, performing from epicardium to endocardium, from base to apex and the septum to the LV lateral wall[7,11,19]. The more committed segments are apical, followed by bottom and side midventricular[11]. The dysfunction because of stoppage in compaction of these cardiac segments explains LVNC clinic.

Although present at birth, symptoms usually appear late, depending mainly on systolic dysfunction[10]. The three major clinical manifestations of LVNC include heart failure, arrhythmias and stroke events[13,16,21,23].

More than two thirds of patients with LVNC present symptomatic heart failure at the time of diagnosis[16,17], the result of subendocardial hypoperfusion, microcirculatory dysfunction and decreased coronary flow reserve in the absence of epicardial coronary lesions[11,16,17,23], which determine reduced EF. Women often exhibit higher levels of dyspnea more with greater involvement of the anterior, posterior and lateral over males with predominance in the apical segment[8].

Arrhythmias are very common. 25% of adults have chronic atrial fibrillation (AF) and 47% ventricular tachyarrhythmias (predictor of SD)[7,17,18]. Arrhythmias in pediatric patients are rare[13,24]. Up to 90% have electrocardiographic abnormalities (ventricular hypertrophy, left bundle branch block (LBBB)[10], ST segment changes and inverted T waves)[17]. The presence of LBBB in children is much lower, which may be due to the endocardial fibrosis progression along the years. Up to 15% of children with LVNC have Wolff-Parkinson-White ECG[7], a finding that is not repeated in adults. In infants we often see high biventricular voltages[17].

Thromboembolic events[25] (stroke, transient ischemic attack, pulmonary embolism and mesenteric stroke) occur in 20-40% of adults[6,7], being anecdotal in children[13]. In etiology mixed thrombus formation in intertrabecular spaces, systolic dysfunction and the presence of AF[17].

Echocardiography is currently the main diagnostic test. Although there are different criteria the latter are the most referenced in the literature (TABLE 2)[6,14]. LVNC mainly affects the apical and inferior mid-ventricular and to 80 % may have affected lateral sides; in children there maybe septal involvement. Alterations of the segmental motility are frequent. The correlation between ultrasound criteria is low (only 30% of all patients met all the criteria[2]) so that the MRI (Figures 4 to 6), which presents a better spatial resolution of the apex and the left ventricular free wall, has a potential value for the diagnosis, especially in patients where good quality ultrasound images are not achieved[10,18]. With this technique the compacted and non-compacted layers are very well define the with a ratio > 2.3 at the end of diastole with excellent levels of sensitivity, specificity and positive and negative predictive values (86 %, 99 %, 75 % and 99%)[16].

The differential diagnosis must be made with hypertrophic cardiomyopathy, dilated cardiomyopathy, left ventricular hypertrabeculation associated with other cardiac pathologies such as aortic stenosis or poorly controlled longstanding hypertension, thrombi and intramyocardial hematoma, arrhythmogenic right ventricular dysplasia, cardiac metastases or intramyocardial abscesses[6,11,18].

The prognosis of patients with LVNC in symp-

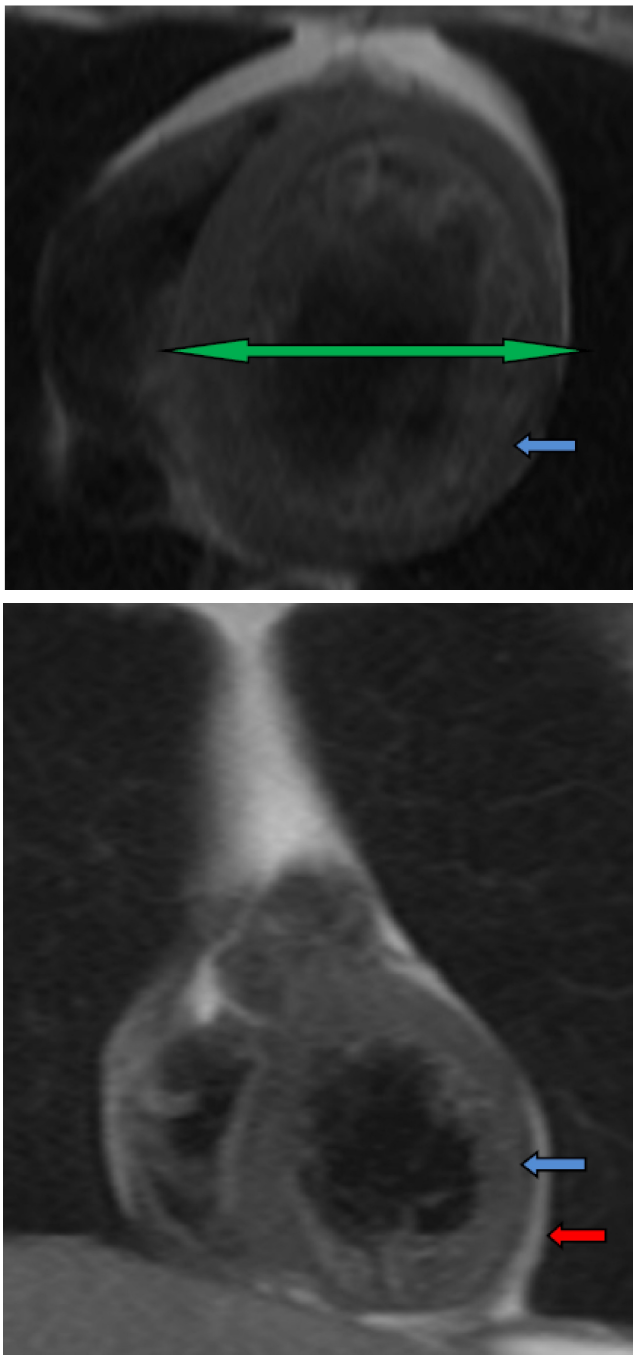

Figure 4 : Cardiac MRI. Myocardial bilaminar in appearance with a compacted layer (red arrow) and non-compacted (blue arrow) with a ratio greater than 2.5 non-compacted/compacted. Ventricular dilatation (green arrow)

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tomatic forms is not good[7]. In one of the first series of patients mortality during the following 6 years was 80%[9], in another 60% of patients suffered a SD or underwent transplantation in the first 6 years after diagnosis[6]. The prognosis is determined by the degree and progression of heart failure, embolic events and arrhythmias. The SD, arrhythmias and emboli are less common in pediatric patients. Among the features that have been associated with poor outcomes are the presence of a higher diastolic diameter at diagnosis, functional class III-IV heart failure patients, permanent or persistent atrial fibrillation and bundle branch block[16].

In the absence of randomized studies, the 2008 AHA guidelines recommend implantation of a cardioverter defibrillator (ICD) in all patients with LVNC to reduce the risk of sudden death (class IIb, level of evidence C)[5]. With the advancement of knowledge of the LVNC it is observed that the disease includes a wide clinical spectrum, from no symptoms to advanced ventricular dysfunction. For mild and asymptomatic cases, close monitoring seems appropriate. Patients who develop mild to moderate heart failure should be treated according to routine clinical practice guidelines (beta blockers, ACE inhibitors, antiplatelet agents, diuretics, etc.). The β-blockers are the mainstay of pharmacological treatment[11]. When associated with AF and increased ventricular dysfunction, anticoagulation must be considered[5]. Some authors recommend anticoagulation in all adults with LVNC[17] while other groups reserved for patients with EF < 40 %, history of embolism or AF[7]. If there is advanced heart failure, cardiac resynchronization therapy or transplantation seems the only alternatives. Finally, for patients who have recovered from a cardiac arrest, recurrent (or poorly tolerated) VT, ICD implantation is recommended[5].

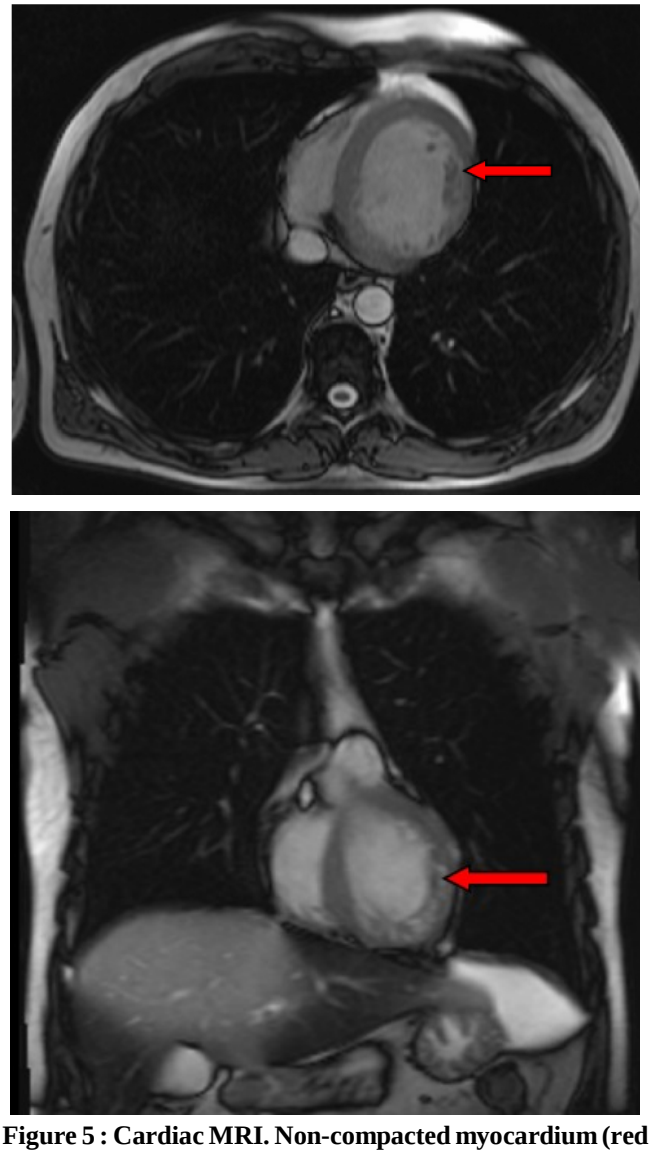

Figure 5 : Cardiac MRI. Non-compacted myocardium (red arrow)

Ethical responsibilities

Protection of people and animals: The authors declare that for this investigation, no experiments have been completed on humans or animals.

Confidentiality of data: The authors declare that in this article the patients personal information does not

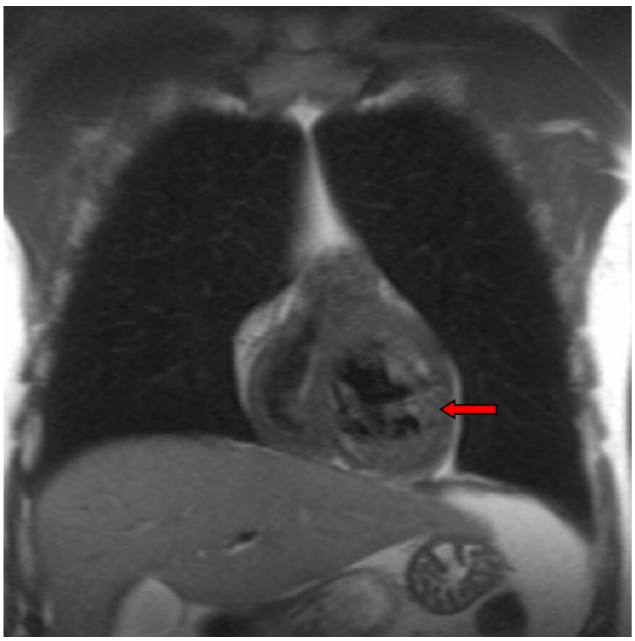

Figure 6 : MRI. Left ventricular dilation LVNC criteria (red arrow)

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TABLE 1 : Classification of cardiomyopathy[12]

Primary Cardiomyopathies		
Genetic	Hypertrophic Cardiomyopathy	
	Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia	
	Noncompaction of ventricular myocardium	
	Glycogen storage diseases	
	Conduction System Disease	
	Mitochondrial, Cardiomyopathy	
	Ion Channelopathies	Brugada Syndrome
		Long-QT Syndrome
		Short-QT Syndrome
		Catecholaminergic Polymorphic Ventricular Tachycardia
		Asian sudden unexplained nocturnal death syndrome,
		Idiopathic Ventricular Fibrillation
Mixed (genetic and acquired)	Dilated Cardiomyopathy	
	Primary Restrictive Nonhypertrophied Cardiomyopathy	
Acquired	Myocarditis (Inflammatory Cardiomyopathy)	Toxins and drugs
		Infectious
		Hipersensibilidad
		De células gigantes
	Stress-provoked	(“Tako-Tsubo”) Cardiomyopathy
	Peripartum (postpartum) cardiomyopathy	
	Tachymiocardiopathy	
	Cardiomyopathy of infants of insulindependent diabetic mothers.	
Secondary Cardiomyopathies		
Infiltrative	Amyloidosis, Gaucher disease, Hurler’s disease, Hunter’s disease	
Storage	Hemochromatosis, Fabry’s disease, Glycogen storage disease (type II, Pompe), Niemann-Pick disease	
Toxicity	Drugs, heavy metals, chemical agents	
Inflammatory (granulomatous)	Sarcoidosis, Endomyocardial fibrosis, Hypereosinophilic syndrome (Löeffler’s endocarditis)	
Endocrine	Diabetes mellitus, Hyperthyroidism, Hypothyroidism, Hyperparathyroidism, Pheochromocytoma, Acromegaly	
Cardiofaciales	Noonan síndrome, Lentiginosis	
Neuromuscular/neurological	Friedreich’s ataxia, Duchenne-Becker muscular dystrophy, Emery-Dreifuss muscular dystrophy. Myotonic dystrophy, Neurofibromatosis.	
Nutritional deficiencies	Beriberi (thiamine), pallagra, scurvy, selenium, carnitine, kwashiorkor	
Autoimmune/collagen	Systemic lupus erythematosis, Dermatomyositis, Rheumatoid arthritis. Scleroderma, Polyarteritis nodosa.	
Electrolyte imbalance		
Consequence of cancer therapy	Anthracyclines: doxorubicin (adriamycin), daunorubicin, Cyclophosphamide	
	Radiation	

appear.

Right to privacy and informed consent: The authors declare that in this article the patients personal information does not appear.

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To Jeremy Brenan who helped us in this English

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TABLE 2 : Jenni LVNC diagnostic ultrasound criteria[14]

Coexisting cardiac abnormalities were absent
A two layer structure was seen, with a compacted thin epicardial band and a much thicker non-compacted endocardial layer of trabecular meshwork with deep endomyocardial spaces. A maximal end systolic ratio of non-compacted to compacted layers of > 2 is diagnostic.
The predominant localisation of the pathology was to mid-lateral, apical, and mid-inferior areas. The pathological preparations confirmed the echocardiographic findings. Concomitant regional hypokinesia was not confined to the non-compacted segments.
There is colour Doppler evidence of deep perfused intertrabecular recesses.

version.

REFERENCES

[1] L.Monserrat, M.Hermida Prieto, A.Castro Beiras; Avances en Miocardiopatía dilatada: Del genotipo al fenotipo clínico. Rev.Esp.Cardiol.Supl., **7**, F-13F **(2007)**.

[2] S.K.Kohli, A.A.Pantazis, Y.S.Shan, B.Adeyemi, G.Jackson, W.J.McKenna, S.Sharma, P.M.Elliott; Diagnosis of left-ventricular systolic dysfunction: time for a reappraisal of diagnostic criteria?. European Heart Journal, **29**, 89-95 **(2008)**.

[3] O.Sousa, G.Silva, F.Sampaio, M.Olivira, H.Gonçalves, J.Priomo, J.Ribeiro, C.Fonseca, A.Alburuerque, L.Simões, V.Gama; Isolated left ventricular non-compaction: a single-center experience. Rev.Port.Cardiol, Accesible en http://dx.doi.org/10.1016/j.repc.2012.06.010, **32(3)**, 229-38 **(2013)**.

[4] A.Lorsheyd, M.J.MCramer, B.K.Velthuis, E.J.P.Vonken, J.Van der Smagt, P.Van tintelen, R.N.W.Hauer; Familial occurrence of isolated non-compaction cardiomyopath. European Journal of Heat Failure, **8**, 826-31 **(2006)**.

[5] N.D.Galizio, J.L.González, L.E.Favaloro, M.Díez, A.Fernández, E.Guevara, A.A.Palazzo, F.Roblés, J.H.Casabé; Miocardiopatía no compactada. Estratificación de riesgo de muerte súbita para indicación de cardiodesfibrilador automático implantable. Rev.Argent.Cardiol., **79**, 14-20 **(2011)**.

[6] L.Veira da Rosa, V.Cury Salemi, L.Machado Alexandre, D.Mady; Miocardiopatía no compactada: una visión actual. Arq.Bras.Cardiol., **97(1)**, e13-e19 **(2011)**.

[7] C.Correa Zalazar, L.M.Lescano, P.Gallo, R.Barreto, F.Núñez Burgos; Ventrículo izquierdo no compactado. Rev.Fed.Arg.Cardio., **38**, 86-91 **(2009)**.

[8] C.Stöllberger, G.Blazek, M.Winker-Dworak, J.Finsterer; Diferencias de sexo en la ausencia de compactación ventricular izquierda con y sin trastornos neuromusculares. Rev.Esp.Cardiol., **61(2)**, 130-6 **(2008)**.

[9] L.S.Briongos Figuero, P.Bachiller Luque, T.M.Pérez Sanz, A.Vara Castrodeza; Miocardiopatía no compactada en paciente con infección VIH. Semergen, **64**, 233-5 **(2010)**.

[10] T.García-Paredes, M.D.Fernández-Zamora, J.Mora-Ordóñez, J.A.Ferriz Martín, J.Muñoz-Bono, A.Vera-Almazán et al.; Miocardiopatía no compactada: una causa poco frecuente de taquicardia ventricular. Med. Intensiva [revista en la Internet]. http://scielo.isciii.es/scielo.php?script=sci_arttext&pid=S0210-56912007000500008&lng=es, **31(5)**, 261-264 **(2007)**.

[11] S.Lissasoain; Ventrículo izquierdo no compactado. Artículo de revisión. http://www.cardiomil.com.uy/archivos/vizqnocompactado.pdf

[12] B.J.Maron, J.A.Towbin, G.Thiene, C.Antzelevich, D.Corrado, D.Amett, E.Seidman, J.Young; Contemporary Definitions and Classification of the Cardiomyopathies. Circulation, **113**, 1807-1816 **(2006)**.

[13] A.Sabaté Rotés, V.M.Huertas Quiñones, P.Betrián, J.Carretero, L.Jiménez, J.Girona, S.Uriel, R.Perich, R.Collell, F.Prada, D.C.Albert; Miocardiopatía no compactada: características clínicas, evolutivas, pronósticas en edad pediátrica. Resultado de un estudio multicéntrico. An Pediatr (Barc), **(2001)**.

[14] R.Jenni, R.Schneider, C.Attenhofer Jost, P.A.Kaufmann; Echocardiographic and pathoanatomical characteristisc of isolated left vntricular non-compaction: A step towards clasification as a distinct cardiomypathy. Heart, **86**, 666-671 **(2001)**.

[15] N.Espinola Zavaleta, M.Elena Soto, L.Muñoz Castellanos, S.Játiva Chávez, C.Keirns; Non-compacted cardiomyopathy: clinical-echocardiographic stude. Cardiovascular Ultrasound, Accesible en www.cardiovascularultrasound.com/content/4/I/35, **4**, 35 **(2006)**.

[16] J.Ganame, A.Ayres, R.H.Pignatelli; Ventrículo izquierdo no compactado: Una forma de

miocardiopatía recientemente reconocida (Left Ventricular Noncompactation). Insuf Cardiaca, **3(1)**, 125-30 **(2006)**.

[17] C.A.Dumont; No compactación del miocardio ventricular. 5º Congreso Virtual de Cardiología. http://www.fac.org.ar/qcvc/llave/c085e/dumontc.php

[18] T.García Paredes, M.D.Fernández Zamora, J.Mora Ordóñez, J.A.Ferrriz Martín, J.Muñoz Bono, A.Vera Almazán, G.Quesada García; Miocardiopatía no compactada: Una causa oco frecuente de taquicardia ventricular. Med Intensiva, **31(5)**, 261-4 **(2007)**.

[19] L.Monserrat Iglesias; Miocardiopatía no compactada: una enfermedad en busca de criterios. Rev.Esp.Cardiol., **61(2)**, 112-5 **(2008)**.

[20] S.E.Hughes, W.J.McKenna; New insights into the pathology of inherited cardiomyopathy. Heart, **91**, 257-64 **(2005)**.

[21] M.Martín, E.Santamarta, C.Corros, E.M.Benito, D.León, E.Velasco, A.García-Campos, M.L.Rodríguez, J.M.De la Hera, V.Barriales, A.Saiz, J.L.Lambert; Left ventricle non-compactation cardiomyopathy; different clinical scenarios and magnetic resonance imaging findings. Arch.Cardiol.Mex., **83(1)**, 3539 **(2013)**.

[22] R.T.Murphy, R.Thaman, J.Gimeno Blanes, D.Ward, E.Sevdalis, E.Papra, A.Kiotsekolglou, M.T.Tome, D.Pellerin, W.J.McKenna, P.M.Elliot; Natural history anf familial characteristics of isolated left ventricular non-compactation. European Heart Journal, **26**, 185-92 **(2005)**.

[23] E.Vizzardi, S.Nodari, M.Metra, L.Dei Cas; Non-compactation of the ventricular myocardium. Heart International, **3-4**, 178-83 **(2006)**.

[24] A.Enríquez, R.Baeza, L.Gabrielli, S.Córdova, P.Castro; Miocardiopatía no compactada: una serie de 15 casos. Rev.Med.Chile., **139**, 864-71 **(2011)**.

[25] Trejo Gabriel, J.M.Galán, N.Sánchez Mata, Y.Bravo Anguiano, J.M.Téllez García; Miocardiopatía no compactada presentándose como ictus de repetición. Rev.Clin.Esp., **210(7)**, 365-70 **(2010)**.