Conclusions

Myocarditis in adolescents almost always mimics bed rest until troponin normalization and sport avoidance. In all patients, the CRP was slightly increased, with values between 7 and 56 mg/dl; hsCRP was also elevated. CPK was increased in all cases, varying between 134–1834 ng/ml. The highest value was in Duchenne patient. CPK-MB was slightly increased in 4 patients and very increased in two. Troponin I was positive in two patients with values 54 and 51 pg/ml and increased values in four, between 669 and 2500 pg/ml. Tropinin I had very high values, from 501 to 30,000pg/ml in one case of multifocal myocarditis.

NTproBNP was altered in all patients, with values between 72 and 1508 pg/ml.

TGO and TGP were elevated, especially TGO. ECG in all cases had the aspect of acute STEMI. Two cases were explored by angioconoronarography with negative result. Ejection fraction was at the lower limit of normal in 3 cases and reduced between 45-49% in the other three. Speckle-tracking was significant in all 6 cases. Holter ECG detected PVC in 2 cases and bradycardia in one.

Cardiac MRI revealed inflamed heart muscle at late gadolinium enhancement.

MRI was repeated at 6 mo and 1 year after, with good results. Viral serology was negative for: parvovirus, coxackie, adenovirus, cytomegalovirus.

Treatment included: AINS, Carvedilol, diuretic, vitamins, bed rest until troponin normalization and sport avoidance 6 months. Treatment options are combination of life-style modification, antiarrhythmic drugs, catheter ablation and ICD implantation.

Here we are illustrating a 15-year old boy who presented with dizziness and syncope. Cardiac diagnostic workup was performed, revealing arrhythmogenic cardiomyopathy. Inverted T waves in V1-V4 leads with epsilon waves and premature ventricular contractions with LBBB were noted in standard ECG.

During the 24-hours holter ECG sustained ventricular tachycardia was registered. Echocardiography and cardiac MRI revealed hypertrabeculated right ventricle with regional dyskinesia and akinesia, asynchronous right ventricle contractions and additionally fibrous replacement of the right myocardial free wall. ICD was implanted after inadequate antiarrhythmic drugs response.

Conclusion cardiac diagnostic workup should be performed as a severe and progressive heart muscle disease can be the underlying cause of dizziness and syncope.

Material and Methods

During a two year interval, a number of 6 adolescent males were admitted in the ED with severe precordial pain. All were between 14-17 yo. They performed lab investigations, ECG, Echocardiography, Speckle-tracking, Holter ECG and cardiac biomarkers. Two were explored by angioconoronarography due to acute STEMI aspect on ECG, to exclude myocardial infarction. Angio MRI of the heart performed in all patients.

Results

Six males were admitted for severe precordial pain, 8 on a scale of 0-10, which appeared within the last 12 hours, with a duration of 1-2 hours, accompanied by palpitations. One boy had Duchenne disease and one thrombophilia. The CRP was slightly elevated in all cases, with values between 7 and 56 mg/dl; hsCRP was also elevated. CPK was increased in all cases, varying between 134–1834 ng/ml. The highest value was in Duchene patient. CPK-MB was slightly increased in 4 patients and very increased in two. Troponin I was positive in two patients with values 54 and 51 pg/ml and increased values in four, between 669 and 2500 pg/ml. Tropinin I had very high values, from 501 to 30,000pg/ml in one case of multifocal myocarditis.

NTproBNP was altered in all patients, with values between 72 and 1508 pg/ml.

TGO and TGP were elevated, especially TGO. ECG in all cases had the aspect of acute STEMI. Two cases were explored by angioconoronarography with negative result. Ejection fraction was at the lower limit of normal in 3 cases and reduced between 45-49% in the other three. Speckle-tracking was significant in all 6 cases. Holter ECG detected PVC in 2 cases and bradycardia in one.

Cardiac MRI revealed inflamed heart muscle at late gadolinium enhancement.

MRI was repeated at 6 mo and 1 year after, with good results. Viral serology was negative for: parvovirus, coxackie, adenovirus, cytomegalovirus.

Treatment included: AINS, Carvedilol, diuretic, vitamins, bed rest until troponin normalization and sport avoidance 6 months. Treatment options are combination of life-style modification, antiarrhythmic drugs, catheter ablation and ICD implantation.

Here we are illustrating a 15-year old boy who presented with dizziness and syncope. Cardiac diagnostic workup was performed, revealing arrhythmogenic cardiomyopathy. Inverted T waves in V1-V4 leads with epsilon waves and premature ventricular contractions with LBBB were noted in standard ECG.

During the 24-hours holter ECG sustained ventricular tachycardia was registered. Echocardiography and cardiac MRI revealed hypertrabeculated right ventricle with regional dyskinesia and akinesia, asynchronous right ventricle contractions and additionally fibrous replacement of the right myocardial free wall. ICD was implanted after inadequate antiarrhythmic drugs response.

Conclusion cardiac diagnostic workup should be performed as a severe and progressive heart muscle disease can be the underlying cause of dizziness and syncope.

Autosomal dominant or autosomal recessive pattern of inheritance are known, latter as part of cardiocutaneous syndromes. The mean age of diagnosis is 30 years, clinical manifestation is rare before puberty. Following hypertrophic cardiomyopathy, it is considered to be the leading cause of sudden cardiac death in adolescence, especially among athletes.

Palpitations, dizziness and syncope can be symptoms of the disease, though sudden cardiac arrest may be the first clinical presentation. Diagnosis is made by combining medical history, ECG, morphological abnormalities (echocardiography and/or cardiac MRI), endomyocardial biopsy and genetic tests. Standardized diagnostic criteria (major and minor), revised in 2010 were made to increase sensitivity and specificity. Treatment options are combination of life-style modification, antiarrhythmic drugs, catheter ablation and ICD implantation.

Here we are illustrating a 15-year old boy who presented with dizziness and syncope. Cardiac diagnostic workup was performed, revealing arrhythmogenic cardiomyopathy. Inverted T waves in V1-V4 leads with epsilon waves and premature ventricular contractions with LBBB were noted in standard ECG.

During the 24-hours holter ECG sustained ventricular tachycardia was registered. Echocardiography and cardiac MRI revealed hypertrabeculated right ventricle with regional dyskinesia and akinesia, asynchronous right ventricle contractions and additionally fibrous replacement of the right myocardial free wall. ICD was implanted after inadequate antiarrhythmic drugs response.

Conclusion cardiac diagnostic workup should be performed as a severe and progressive heart muscle disease can be the underlying cause of dizziness and syncope.

Arrhythmogenic cardiomyopathy (arrhythmogenic right ventricular dysplasia) is a hereditary heart muscle disease characterized pathologically by dystrophy and fibrofatty replacement of the right ventricular myocardium, consequently causing ventricular dysfunction and life-threatening cardiac arrhythmias. The right ventricle is predominately affected, but approximately 70% of hearts studied during autopsy reveal left ventricle involvement. Positive family history is seen in 30% cases.

Autosomal dominant or autosomal recessive pattern of inheritance are known, latter as part of cardiocutaneous syndromes. The mean age of diagnosis is 30 years, clinical manifestation is rare before puberty. Following hypertrophic cardiomyopathy, it is considered to be the leading cause of sudden cardiac death in adolescence, especially among athletes.

Palpitations, dizziness and syncope can be symptoms of the disease, though sudden cardiac arrest may be the first clinical presentation. Diagnosis is made by combining medical history, ECG, morphological abnormalities (echocardiography and/or cardiac MRI), endomyocardial biopsy and genetic tests. Standardized diagnostic criteria (major and minor), revised in 2010 were made to increase sensitivity and specificity. Treatment options are combination of life-style modification, antiarrhythmic drugs, catheter ablation and ICD implantation.

Here we are illustrating a 15-year old boy who presented with dizziness and syncope. Cardiac diagnostic workup was performed, revealing arrhythmogenic cardiomyopathy. Inverted T waves in V1-V4 leads with epsilon waves and premature ventricular contractions with LBBB were noted in standard ECG.

During the 24-hours holter ECG sustained ventricular tachycardia was registered. Echocardiography and cardiac MRI revealed hypertrabeculated right ventricle with regional dyskinesia and akinesia, asynchronous right ventricle contractions and additionally fibrous replacement of the right myocardial free wall. ICD was implanted after inadequate antiarrhythmic drugs response.

Conclusion cardiac diagnostic workup should be performed as a severe and progressive heart muscle disease can be the underlying cause of dizziness and syncope.

Very rare case of Noonan Syndrome, Type 2

LA Gandara*, NV Zhurkova, EN Basargina, AA Pushkov, TD Degtayreva, VP Miroshnichenko, OB Kondakova, KV Savostyanov. National Medical Research Center of Children’s Health of the Ministry of Health of the Russian Federation, Moscow, Russian Federation

Background

Noonan syndrome, type 2 (NS2) is rare autosomal recessive disorder of RASopathies group, caused by mutations in the LZTR1 gene. NS2 characterized by a typical face, short stature, broad, short neck, congenital heart disease, developmental delay. The most common heart disease in children with NS2 is hypertrophic cardiomyopathy.

Patients and Methods

Patient is a boy, 15 years old with short stature, developmental delay at 1 first year of life and heart disease. He had distinctive facial features of NS: downslanting palpebral fissures, epicantalic folds, hypertelorism, low-set ears, short neck, wing-like folds on the neck, pectus deformity. Hypertrophic cardiomyopathy was identified at 1 month old. Now patient have obstructive, hyperthrophic cardiomyopathy, cardiac arrhythmia: ventricular extrasystoles, 4A Lown, intraventricular block combined with bundle branch block, transient WPW. Syncopal episodes.

Surgical correction of obstructive hypertrophic cardiomyopathy: septal myomectomy was performed on the child due to the high risk of developing sudden death syndrome. Target areas of the exome were investigated by next generation sequencing (NGS). Bioinformatic analysis was carried out using ACMG recommendation. Validation of the identified variants was carried out by the Sanger method.

Abstracts

Arrhythmogenic cardiomyopathy (arrhythmogenic right ventricular dysplasia) is a hereditary heart muscle disease characterized pathologically by dystrophy and fibrofatty replacement of the right ventricular myocardium, consequently causing ventricular dysfunction and life-threatening cardiac arrhythmias. The right ventricle is predominately affected, but approximately 70% of hearts studied during autopsy reveal left ventricle involvement. Positive family history is seen in 30% cases.