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CLINICAL CASE OF A PATIENT WITH EBSTEIN ANOMALY- A RARE NOSOLOGICAL DISCOVERY

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Abstract:

Congenital heart defects (CHD) are among the common diseases in childhood. According to global statistics, the overall incidence of children born with these defects is 6-8 per 1000 live births. There are over 40 nosological forms of CHD, but the most prevalent are 9 forms that constitute approximately 85% of the total. The remaining 15% encompass rare and complex forms.A unique feature, encountered very rarely and constituting a nosological discovery in adult therapeutic practice, is Ebstein anomaly (EA).

Ebstein anomaly (EA) is a complex congenital heart defect (CHD) arising from the displacement of the septal and posterior leaflets of the tricuspid valve (TV). What makes this defect even more unique is the incredible variability in anatomical variants that can be identified.

Ebstein anomaly is found in newborns and can lead to serious health problems, including heart failure, arrhythmia, and the risk of thromboembolism. In some cases, the defect can be so severe that it poses a threat to life and requires immediate medical intervention.

Symptoms of Ebstein anomaly (EA) may include shortness of breath, fatigue, cyanosis of the skin, and swelling of the eyelids, which can arise due to the presence of fluid in the body. The diagnosis of the defect is typically established through echocardiography, enabling physicians to visualize anatomical changes in the heart and determine their severity.

The treatment of EA can vary depending on the severity of the defect and the presence of accompanying complications. For some patients, pharmacological therapy may effectively control symptoms and slow the progression of the defect. In more serious cases, surgical intervention may be necessary, including heart valve reconstruction or the implantation of an artificial valve.

Overall, Ebstein anomaly is a complex and unpredictable heart defect that requires careful observation and management by medical professionals. With ongoing advancements in diagnostics and treatment, it is hoped that the majority of patients with this rare anomaly can achieve an acceptable quality of life and continue to lead an active lifestyle.

According to literature data, Ebstein anomaly is considered a relatively rare pathology, with a frequency among all congenital heart defects not exceeding 1%.

There is a significant likelihood that general practitioners may encounter patients with previously undiagnosed EA. Based on numerous observations, the survival rate of patients who have lived 5-10 years after the onset of the disease is relatively high. Therefore, it is crucial to pay attention to possible symptoms of EA and conduct timely diagnostics to increase the chances of successful treatment and patient survival. Primary care physicians should be particularly attentive and prepared for the possibility of encountering this condition in their patients.

In this article, a clinical case of a patient with a rare congenital anomaly - Ebstein anomaly (EA) - will be discussed. The prevalence of this anomaly among all congenital heart defects is no more than 1%. Ebstein anomaly is a heart defect characterized by the displacement of the tricuspid valve leaflets into the right ventricle, along with tricuspid valve regurgitation and right ventricular myopathy. The diagnosis of this anomaly is challenging, and without timely treatment, it can lead to rapid decompensation and unfavorable outcomes.

Differential Diagnosis of Ebstein Anomaly: The Importance of Early Detection and Timely Treatment

Physicians in the therapeutic field play a crucial role in diagnosing and treating various diseases, including congenital heart defects. However, Ebstein anomaly is a rare condition that can be easily overlooked or masked by other symptoms and syndromes, such as angina or circulatory insufficiency.

The purpose of this article is to draw attention to the differential diagnosis of this anomaly and emphasize the significance of early detection of its symptoms and timely intervention. Considering that Ebstein anomaly is a "silent" disease manifesting symptoms resembling myocardial coronary pathology, physicians must be cautious and attentive to any irregularities or unclear diagnoses.

Upon detecting suspicious symptoms or signs masked by other cardiovascular diseases, primary care physicians should delve deeper into diagnostics, paying particular attention to potential asymptomatic heart defects. Only in this way can accurate and timely diagnosis be ensured, avoiding incorrect treatment or delays.

Furthermore, it is crucial to underscore the management approach for patients post-operative treatment of Ebstein anomaly during the outpatient stage. Adherence to rehabilitation programs and continuous medical monitoring are critical aspects of the postoperative period. This allows for the timely identification and treatment of potential complications, as well as providing patients with necessary support and control.

In conclusion, this article urges physicians to be vigilant in diagnosing and treating asymptomatic heart defects that may be masked by other cardiovascular diseases. Only with a complete understanding of the peculiarities of Ebstein anomaly and the ability to differentiate it from other conditions can physicians provide their patients with the most effective and targeted treatment.

Keywords: Ebstein Anomaly, Congenital Heart Defect, Rhythm Disturbance, WPW Syndrome, Radiofrequency Ablation.

Introduction

Congenital heart defects (CHD) are among the prevalent conditions in childhood. The global incidence of children born with these defects is reported to be 6-8 per 1000 live births, according to world statistics. There are more than 40 nosological forms of CHD, with the most common being 9 forms that constitute 85% of all cases. The remaining 15% includes rare and complex forms [1].

Among the rare forms of congenital heart defects, Ebstein anomaly holds a special place and is considered a rare nosological "discovery" in therapeutic practice.

The anomaly was first described in 1866 by the Prussian pathologist and professor at the University of Göttingen, Wilhelm Ebstein [2]. An interesting fact is that Dr. Ebstein observed the patient in question for only 8 days during his lifetime. However, during the autopsy, Professor Ebstein discovered a previously unknown combination of heart defects, which was later named, in a literal translation, "Ebstein's heart defect."

Professor Ebstein, a disciple of the pathologist R. Virchow, is well-known in cardiological circles primarily for his discovery known as "Virchow's triad." He is also the author of several fundamental scientific works, including manuscripts dedicated to obesity, carbohydrate metabolism disorders, and dehydration. His works also cover the morphology of the kidney and the social hygiene of cities.

However, Professor Ebstein's name became immortalized due to a particular case that has remained in the annals of medical history as "Ebstein anomaly." In this case, the heart of a patient named Joseph Presser, suffering from heart failure, was described, and he succumbed to its decompensation at the age of 19 [2-3].

During the autopsy, it was revealed that the anterior leaflet of the tricuspid valve was enlarged and perforated, while the posterior and septal leaflets were hypoplastic, thickened, and attached to the wall of the right ventricle. The atrialized portion of the right ventricle was dilated, and its walls were thinned. The right atrium was also enlarged, and the oval foramen was open.

The moment of discovery of this "tricuspid valve malformation causing congestive heart failure" occurred when Ebstein was 28 years old.

Currently, two main classifications are widely used in cardiothoracic surgical practice: anatomical and echocardiographic. The anatomical classification is employed for intraoperative assessment, determining the condition of the cardiovascular system during surgery. It allows surgeons to precisely identify anomalies and structural changes in the heart and vessels, as well as assess the complexity of the operation and potential risks.

The other classification, echocardiographic, is used for preoperative assessment and predicting the outcome of the operation. Through this method, detailed information about the anatomy and functional status of the heart can be obtained before surgery. This aids in determining the extent of heart involvement and predicting the patient's prognosis after the operation.

Both classifications are crucial tools in the work of cardiothoracic surgeons. The anatomical classification enables real-time analysis of the heart's condition during surgery, which is a critical factor for decision-making during the operative phase. On the other hand, the echocardiographic classification aids in surgical planning and predicting outcomes, contributing to risk reduction and improved patient prognosis.

Together, these classifications provide a more precise and comprehensive assessment of the cardiovascular system's state before and during surgery. This helps enhance the quality of

surgical interventions and increases the success of the operation, ultimately contributing to the preservation of patients' health and lives.

The complexity of diagnosis, including echocardiography (echoCG), is due to the variety of defect variants distinguished by anatomical types according to A. Carpentier's classification (1988). There are a total of five defect types classified based on the level of fixation and fenestration of the anterior leaflet of the tricuspid valve [4].

Type A: Characterized by a large anterior leaflet of the tricuspid valve (TV) with free movement, unobstructed by interchordal spaces. The displacement of the septal and posterior leaflets of the tricuspid valve is moderate. The atrialized part of the chamber is relatively small, with a thin wall but preserved contractility. The trabecular part of the chamber is normally developed, and the dimensions of the chamber correspond to the norm.

Type B: Characterized by reversed proportions of the volume of the atrialized chamber and the true pumping chamber: the atrialized part of the chamber is larger than the true pumping chamber. The anterior leaflet of the tricuspid valve is large and mobile, with interchordal spaces partially obliterated or obliterated insignificantly. Displaced leaflets of the tricuspid valve are often hypoplastic and attached to the wall of the pumping chamber. The atrialized space of the pumping chamber has large dimensions, and the wall of the atrialized part is thin and non-contractile.

Type C: Characterized by limited mobility of the anterior leaflet of the tricuspid valve due to its fixation to the anterior wall of the pumping chamber by fibrous strips or shortened chords with obliterated interchordal spaces. This limited mobility can create significant obstruction of the right ventricular outflow tract. Displacement of the posterior and septal leaflets of the tricuspid valve is pronounced. The atrialized part of the pumping chamber has large dimensions with a thin, non-contractile wall, while the pumping chamber itself is very small, thin-walled, and functionally inadequate.

Type D is characterized by the entire atrioventricular septum being lined with adherent fibrous tissue of the anterior leaflet and almost completely atrialized, except for a small infundibular portion. The interchordal spaces are obliterated, and the septal and posterior leaflets are absent. The inlet portion of the atrialized chamber represents a non-contractile cavity. The boundaries of the atrialized cavity are challenging to identify, and the infundibular portion of the atrialized chamber is significantly reduced in size with a thin wall.

The echocardiographic classification by Great Ormond Street, also known as the Goza score, was developed by Celermajer and colleagues. It is based on determining the ratio between the atrialized (non-functional) part of the right ventricle and its functional part, as well as measuring the dimensions of the left ventricle in the four-chamber position at end-diastole. This classification has four grades depending on the degree of expression of these parameters.

If the ratio is 0.5, it corresponds to the first degree of classification. The second degree is defined when the ratio is between 0.5 and 0.99. The third degree is characterized by a ratio from 1 to 1.49, and the fourth degree is when the ratio is more than 1.5.

This gradation of the classification allows predicting the natural course of the defect in patients. If the ratio exceeds 1.5, the probability of a fatal outcome is 100%. With a ratio between 1 and 1.4, the likelihood of early fatal outcomes is significantly lower, generally reaching up to 10%, but the mortality rate in early childhood can go up to 45%. In cases where the ratio is less than 1, the probability of a fatal outcome is extremely low, and the survival rate in such patients reaches 92%.

There is a significant likelihood that general practitioners may encounter patients with previously undiagnosed Ebstein's anomaly (EA). Based on numerous observations, the survival rate of patients who have lived 5-10 years after the onset of the disease is quite high. Therefore, it is essential to pay attention to potential EA symptoms and conduct timely diagnostics to increase the chances of successful treatment and patient survival. General practitioners should be particularly attentive and prepared for the possibility of encountering this condition in their patients.

Ebstein's Anomaly (EA) is a complex congenital heart defect (CHD) characterized by the displacement of the septal and posterior leaflets of the tricuspid valve (TV) and remarkable anatomical variability of the defect [5-7].

EA is a serious condition that leads to the formation of a hemodynamic model. This model is based on tricuspid valve insufficiency, resulting in a reduction in extracapillary fluid volume (ECF) and a deficit in pulmonary blood flow. Often, EA is associated with the non-closure of the oval foramen and right-to-left shunting, which has a compensatory nature. Transesophageal echocardiography may be performed to verify the diagnosis of EA and determine the nature of existing hemodynamic changes [6-7].

The altered geometry of the right atrium and ventricle may serve as the morphofunctional basis for rhythm disturbances in patients with this category. It has been established that in 30-35% of cases in patients with atrial ectopic (AE) rhythm, Wolff-Parkinson-White (WPW) syndrome is diagnosed. A characteristic feature of patients in this category is that accessory atrioventricular connections (AAVC) are mostly localized in the right heart chambers, and half of these patients are diagnosed with multiple AAVCs [8-9].

Today, it is proven that the intake of lithium by the mother can lead to a high likelihood of a congenital heart defect during the child's intrauterine development. According to J. Nora (1976), J. Park et al. (1980), and D. Radford et al. (1985), women who took lithium carbonate-containing medications during the first trimester of pregnancy have a high risk of developing a heart defect, and the birth of children with Ebstein's anomaly (EA) [10].

According to literature data, Ebstein's anomaly (EA) is considered a relatively rare pathology, with a frequency among all congenital heart defects not exceeding 1%. Unfortunately, a significant portion of adult patients remains undiagnosed, while timely diagnosis and surgical correction (valve-sparing operation, prosthesis implantation) could substantially increase their life expectancy. The main reasons for the low detection rate and misdiagnosis include the poor awareness of general practitioners about this defect and the diversity of clinical variants, ranging from asymptomatic courses [11].

Despite improvements in immediate postnatal treatment facilitated by prenatal diagnosis of Ebstein's anomaly, neonatal cardiac surgical correction for such patients, the likelihood of a general practitioner encountering patients born before the development and widespread implementation of prenatal screening is quite high [12-13].

During the medical examination of patients with esophageal atresia, special attention should be given to the prevention of infective endocarditis, regular monitoring, and individual selection of drugs for treating heart failure and arrhythmias.

The clinical manifestations of EA depend on the size and degree of displacement of the tricuspid valve leaflets, the dimensions of the atrialized portion of the right ventricle, and the size of the interatrial communication [14-15].

In childhood, congenital heart defects often go undetected due to mild and inconspicuous symptoms. The most common complaint among other patients is shortness of breath. In the vast

majority of patients, shortness of breath occurs only with physical exertion. 90% of patients experience rapid fatigue. A pathognomonic sign is considered to be episodes of palpitations, observed in 80-90% of patients.

The presence of arrhythmic syndrome in these patients is one of the unfavorable prognostic factors for the course of the disease. Its treatment involves the prescription of continuous antiarrhythmic therapy (AAT), radiofrequency ablation (RFA), or during radical surgical procedures to correct the tricuspid valve defect [16-19].

Clinical Case

On March 21, 2023, a 34-year-old woman, Ms. N, presented with complaints of periodically occurring episodes of rapid heartbeat, occurring independently of external factors, sometimes in the context of psycho-emotional stress, accompanied by bouts of suffocation and sensations of heaviness in the chest.

Over the past two weeks, the patient experienced three such episodes, lasting several hours, which arose and resolved spontaneously. The patient has been troubled by these symptoms since 2001 when the first episode of palpitations occurred, and emergency medical assistance was provided. However, the patient lacks a precise description of the method used to relieve the symptoms.

Following an examination at the regional children's hospital, the patient was diagnosed with Wolff-Parkinson-White syndrome (WPW). Surgical treatment was recommended, but the patient declined.

In August 2005, the patient underwent inpatient treatment at the Tambov Regional Hospital, where a new diagnosis of congenital heart defect - Ebstein's anomaly - was established. In connection with this, a decision was made to refer the patient for further examination and to determine the necessity of surgical treatment for the cardiac arrhythmia disorder at the National Medical Research Center of Cardiac Surgery named after A.N. Bakulev.

Outpatient consultation with a cardiologist, currently undergoing continuous treatment with a dosage of 1.25 mg bisoprolol. However, on March 21, 2023, at 3:00 PM, the patient experienced symptoms such as palpitations and weakness. Due to this condition, emergency medical services were called, and the patient was transported to the Archimedes Luke City Hospital, where further treatment was recommended in the therapeutic department.

From the medical history: born full-term, normal growth and development. No lag in physical or mental development compared to peers. Living conditions are satisfactory. Adequate and regular nutrition. Material and household conditions are satisfactory. Allergic history includes urticaria in response to antibiotics of the penicillin group.

Past illnesses, injuries, surgeries: Appendectomy in 2001. Denies HIV, syphilis, hepatitis, tuberculosis.

During the objective examination, the patient's general condition is satisfactory. Body temperature at the time of examination is 36.6°C. The patient's physical activity is active, and the level of consciousness is clear. The patient has a height of 170 cm and a weight of 75 kg, corresponding to a normal body build (normosthenic). The patient's skin is pale-pink, clean, and moderately moist. Visible mucous membranes have a pink color with no signs of contamination. Tissue turgor is maintained. Subcutaneous adipose tissue in the patient is excessively developed. The thyroid gland is not enlarged. There is no enlargement of regional lymph nodes; they exhibit a soft-elastic consistency, are mobile, non-tender upon palpation, and are not adherent to the surrounding tissues. The patient's muscles are developed satisfactorily, causing no tenderness

upon palpation and lacking asymmetric signs. The skeletal system is also satisfactorily developed, showing no deformities, and no tenderness is noted upon palpation. Joint configuration remains unchanged, with no tenderness upon palpation, normal skin covering the joints, and fully preserved joint functions.

Nasal breathing is unobstructed. The chest has a cylindrical shape without deformities, uniformly participating in the act of breathing. The predominant type of breathing is thoracic. The respiratory rate is 17 breaths per minute. Accessory musculature does not participate in the act of breathing. No tenderness is detected upon palpation of the chest. Vocal fremitus is symmetrical on both sides. Percussion sound is clear, pulmonary, and the lung borders are within the normal range. Auscultation reveals vesicular breath sounds throughout all lung regions. No crackles are present. Bronchophony is unchanged.

During the objective examination, the chest above the heart area is not deformed. The apical impulse is limited, palpable in the fifth intercostal space 1 cm lateral to the midsternal line. The borders of relative cardiac dullness are not expanded. Heart sounds are clear and rhythmic, with a moderate systolic murmur at the base of the xiphoid process. Vascular murmurs are not audible during auscultation.

The pulse rate is 140 beats per minute, rhythmic. The blood pressure on the left is 130/90. The heart sounds are muffled and rhythmic.

The appetite is good. Oral cavity sanitation has been performed. The tongue is moist and not coated. The abdomen is soft on palpation, painless, with no signs of peritoneal symptoms. Peristalsis is preserved. The liver does not protrude beyond the costal margin, painless on palpation, with a rounded edge and a soft-elastic consistency. The spleen is not palpable. The renal area is unchanged and painless on palpation. The percussion tenderness sign along the lumbar region is negative on both sides. Physiological reflexes are normal.

Nervous system: No signs of meningeal symptoms or focal neurological symptoms were detected.

The diagnosis was confirmed by transthoracic echocardiography, from the apical four-chamber position, visualizing atrialization of the right ventricle, atrial dilation, tricuspid and pulmonary regurgitation of 2-3 degrees, and an open oval window. Systolic pressure in the pulmonary artery was within the normal range, indicating good heart function. A slight mitral regurgitation of the first degree was detected, suggesting some deviation in the functioning of the heart valves. The inferior vena cava valve was elongated, which may be related to some disturbances in the circulatory system. The ejection fraction is 75%, indicating good contractile heart function. The end-diastolic dimension of the left ventricle is 3.3 cm, and the end-systolic dimension remains unspecified.

On the ECG, the patient exhibits supraventricular tachycardia at a rate of 166 beats per minute, rightward deviation of the QRS axis, and complete right bundle branch block (CRBBB). Myocardial changes typical of anterior-septal wall ischemia of the left ventricle are also observed, along with ST segment depression in leads II, III, aVF, V4-V6 up to 1 mm.

The presence of tachyarrhythmic syndrome in patients with Ebstein's anomaly is considered an adverse risk factor determining the severity of the disease course.

In light of this, specialists have decided to refer this patient to the National Medical Research Center for Cardiovascular Surgery named after A.N. Bakulev for additional examination and to determine the need for surgical intervention. The medical center bears significant responsibility for making the correct decision, considering all medical data and the patient's condition. It is crucial to conduct all necessary investigations and consultations before

making a final decision on further treatment. The hope for a positive outcome and improvement in the patient's condition is based on the professionalism and experience of the physicians at the National Medical Research Center for Cardiovascular Surgery named after A.N. Bakulev. Each case requires an individual approach and finding an optimal solution, which is the primary goal of the medical center's activities.

Discussion

The malignant nature of the arrhythmic syndrome plays a pivotal role in the occurrence of fatal outcomes in patients with arrhythmic syndrome. According to current care recommendations for patients with congenital heart defects from the American Heart Association and the American Association for Cardiovascular and Thoracic Surgery, absolute indications for surgical intervention in patients with arrhythmic syndrome include heart failure of functional class III-IV according to the NYHA classification, the presence of atrial fibrillation, and capillary blood oxygen saturation below 80%.

In the presented case of a young patient with Ebstein's anomaly in combination with manifesting Wolff-Parkinson-White syndrome (WPW), there is a high risk of developing secondary atrial fibrillation. Clinical data obtained during the examination allowed us to classify her as a high-risk group for sudden cardiac death and implied the progression of heart failure symptoms. In choosing the treatment strategy for this patient, we anticipated that performing radiofrequency catheter ablation, including the elimination of the WPW syndrome, would reduce the likelihood of developing atrial fibrillation and symptoms of heart failure.

Radiofrequency catheter ablation of Wolff-Parkinson-White (WPW) syndrome in patients with Ebstein's anomaly is a highly effective and safe method for treating arrhythmic syndrome in this patient population. WPW syndrome is characterized by the presence of an accessory pathway in the atrioventricular (AV) bundle, which can lead to the generation of reentrant arrhythmias, including atrial fibrillation. In patients with Ebstein's anomaly, where there is anomalous attachment of the tricuspid valve and a defect in the interatrial septum, the presence of WPW syndrome can significantly increase the risk of developing cardiac arrhythmia.

Conclusion

Arrhythmic syndrome in patients with Ebstein's anomaly is one of the adverse prognostic factors for the course of the disease. Its treatment is carried out by prescribing continuous antiarrhythmic therapy (AAT), performing radiofrequency catheter ablation (RFCA), or during radical surgical intervention to correct the tricuspid valve defect.

The low detectability and misdiagnosis of the defect are associated with several main reasons. Firstly, many general practitioners are not sufficiently informed about this defect, which complicates its timely detection. Additionally, the defect presents with various clinical variants, including asymptomatic courses, further complicating its diagnosis.

The absence of both effective pharmacological treatment and surgical medical intervention for treating the pathology rapidly leads to decompensation and increases the likelihood of an unfavorable prognosis and sudden death. Therefore, it is crucial to diagnose Ebstein's anomaly (AE) at the earliest stages, document the patient's symptoms, objective examination data, and instrumental study findings, accurately assess anatomical and hemodynamic abnormalities, and subsequently refer the patient to a cardiothoracic surgeon for operative treatment. The healthcare provider must be attentive to characteristic signs and symptoms indicating arrhythmia and be prepared to offer the patient the most effective and timely treatment. Only in this way can the best outcome be ensured and possible complications in the future be prevented.

Despite the improvement in immediate postnatal treatment through prenatal diagnosis of Ebstein's anomaly, and the neonatal cardiothoracic correction performed on such patients, the probability of encountering a general practitioner with patients born before the development and widespread implementation of prenatal screening is quite high.

During the medical examination of patients with Ebstein's anomaly, special attention should be paid to the prevention of infectious endocarditis, regular monitoring, and the individual selection of drugs for treating heart failure and arrhythmias.

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