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Case Report

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Late Presentation of Ebstein Anomaly: An Unique Case at 67yrs

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Abstract

Ebstein anomaly, a congenital heart condition, can manifest across a broad age spectrum, including adulthood. We present a unique case of a 67-year-old woman with no significant medical history who sought medical attention for severe headaches, vomiting, and respiratory distress, accompanied by elevated blood pressure and rapid pulse. Initial investigations, including troponin T levels, yielded unremarkable results. However, an electrocardiogram unveiled a right bundle branch block (RBBB) pattern, prompting further assessment. A subsequent chest X-ray disclosed an enlarged heart, indicating cardiomegaly, prompting echocardiography. The echocardiogram confirmed the diagnosis of Ebstein anomaly, revealing a 26 mm apical displacement of the tricuspid valve (TV). The patient was managed conservatively with supplemental oxygen and exhibited gradual improvement. This case underscores the infrequent occurrence of Ebstein anomaly in adulthood and underscores the necessity for thorough evaluation in cases of unexplained cardiopulmonary symptoms.

1. Introduction

Ebstein anomaly, a rare congenital heart defect with an incidence of less than 1% among congenital heart conditions, was initially documented by pathologist Wilhelm Ebstein in 1866 [1,2]. He observed this anomaly during an autopsy of a 19-year-old male who experienced exertional dyspnea, palpitations, and a fatal cardiac arrest [2]. Ebstein anomaly is characterized by several key features, including the tricuspid valve being displaced toward the apex of the heart, the attachment of the septal and posterior leaflets to the heart muscle, enlargement and displacement of the tricuspid annulus, dilation of the part of the right ventricle that resembles the atrium, and the anterior leaflet exhibiting redundancy, fenestrations, and tethering [3]. Typically, Ebstein anomaly is often accompanied by additional cardiac issues, such as a secundum atrial septal defect and varying degrees of obstruction in the right ventricular outflow tract. Some newborns may even present with either functional or anatomical pulmonary atresia, both of which rely on ductal circulation [4]. While less common, other associated cardiac defects can include ventricular septal defects, tetralogy of Fallot, transposition of the great arteries, and atrioventricular canal defects. The clinical presentation varies widely, ranging from no apparent symptoms to severe manifestations, depending on factors like the extent of tricuspid valve displacement, the severity of valve regurgitation, the effective volume within the right ventricle, and the presence of associated structural abnormalities [5]. The likelihood of the condition recurring in the children of affected women is higher at 6%, compared to the lower rate of 0.6% observed in the offspring of affected men [6].

2. Case Presentation

A 67-year-old previously healthy woman presented in ER with complaints of headache, vomiting, and breathlessness. Her blood pressure was elevated at 140/90 mmHg, and her pulse rate was 97 beats per minute. Notably, her oxygen saturation (SpO2) measured 87% on room air. The patient had no significant medical or surgical history, and her initial troponin T levels were within normal limits. However, her ECG revealed a right bundle branch block (RBBB) pattern. Further evaluation included a chest X-ray, which demonstrated cardiomegaly. Given these findings, echocardiography was performed, revealing Ebstein anomaly with a notable apical displacement of the tricuspid valve measuring 26 mm. The diagnosis of Ebstein anomaly in this late-adulthood presentation was confirmed.

3. Management and Follow-up:

The patient was initially managed conservatively with supplemental oxygen to improve her oxygen saturation. Gradually, her symptoms improved, and she stabilized clinically. The absence of significant symptoms, such as cyanosis or heart failure, allowed for non-invasive management.

Subsequent follow-up with a cardiologist was scheduled to monitor her cardiac status and provide appropriate guidance. The patient's unique case underscores the importance of considering rare congenital cardiac anomalies in the differential diagnosis of unexplained symptoms in adulthood, even in the absence of a prior medical history.

4. Discussion

Ebstein anomaly, a rare congenital heart defect, typically manifests in childhood or adolescence, with symptoms often appearing during early life. However, in our presented case, the diagnosis occurred at the remarkable age of 67. This late-onset presentation of Ebstein anomaly is an exceedingly unusual occurrence, with very few reported cases in medical literature. The late discovery of this congenital heart defect in an elderly patient raises several important clinical and diagnostic considerations.

The delayed diagnosis of Ebstein anomaly in this case underscores the diagnostic challenges that can arise when congenital heart anomalies present in adulthood. Unlike more common cardiovascular conditions, the presentation of Ebstein anomaly can be highly variable and non-specific, making it easily overlooked or mistaken for other medical conditions. In our case, the patient's initial complaints of headache, vomiting, and difficulty in breathing were non-specific and did not immediately suggest a cardiac etiology.

The initial assessment of the patient included routine investigations, such as troponin T levels, which were within the normal range. However, the key to unraveling the mystery behind her symptoms lay in a more thorough evaluation. An electrocardiogram (ECG) revealed a right bundle branch block (RBBB) pattern, which raised suspicion of an underlying cardiac condition. Subsequent imaging, including chest X-ray and echocardiography, proved invaluable in confirming the diagnosis. Chest X-rays typically reveal an enlarged heart with a notable enlargement of the right side, resulting in a rounded or globular appearance of the cardiac silhouette. Additionally, an elevated cardiothoracic ratio may be observed due to the enlargement of the right atrium [7]. Echocardiography, in particular, played a pivotal role in this case. It provided a detailed visualization of the heart's structures and function, revealing the characteristic features of Ebstein anomaly, including the apical displacement of the tricuspid valve (TV). The degree of displacement, measured at 26 mm in this instance, is a critical parameter for diagnosing and assessing the severity of Ebstein anomaly.

Given the absence of severe symptoms such as cyanosis or heart failure, the patient was managed conservatively with supplemental oxygen to address her initial hypoxemia. This approach allowed her to stabilize gradually, highlighting the importance of tailoring treatment to the individual patient's clinical presentation. Following the stabilization of the patient's condition, a crucial aspect of her management was the initiation of regular cardiological follow-up. Ebstein anomaly is a complex condition that can lead to various cardiac complications over time. As such, close monitoring by a cardiologist is essential to assess the patient's long-term cardiac health and intervene as necessary.

5. Conclusion

We present a unique case of late-onset Ebstein anomaly diagnosed in a 67-year-old woman presenting with cardiopulmonary symptoms. This case underscores the importance of considering rare congenital heart anomalies in the differential diagnosis of adult patients with unexplained symptoms, even in the absence of a prior medical history. Timely and comprehensive evaluation, along with appropriate cardiological follow-up, is crucial for optimal patient management in such rare cases.

Additional Information

1. Consent for Publication: Patient gave consent to participate in the study

2. Availability of Data and Material: Datasets are not publicly available. They can be obtained from the corresponding author upon reasonable request.

3. Competing Interests: Authors declare no conflicts of interest.

4. Funding: The research received no external funding.

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