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CASE REPORT

A CONCEALED MALFORMATION FOR QUINQUAGENARIAN: DEXTROCARDIA

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ABSTRACT

Dextrocardia is a rare congenital malformation in which the heart is located in the right hemithorax, with an incidence of around 1 in 12,000. Dextrocardia usually remains asymptomatic until accidentally diagnosed through echocardiography and chest radiography, which are mostly performed for other reasons. The complex heart anatomy in dextrocardia can pose great challenges, particularly in patients requiring surgical interventions. In this case presentation, we aimed to emphasize that a multidisciplinary approach is recommended in managing patients with dextrocardia. A 54-year-old female patient arrived with nausea, vomiting, fatigue, and tremor complaints. Medical evaluations revealed bradycardia, which developed after the use of propranolol for a panic attack. The patient underwent an angiography due to the existence of non-ST elevation myocardial infarction and Wenckebach's atrioventricular block, which showed the deviation of the left coronary artery to the right and of the right coronary artery to the left, raising suspicion of dextrocardia. Chest radiography confirmed the presence of dextrocardia. The patient received the necessary treatment, since dextrocardia poses minimal risk, the patient was discharged with follow-up recommendations.

Keywords: Arrhythmia, bradycardia, dextrocardia

INTRODUCTION

Dextrocardia is a rare congenital malformation in which the heart is located in the right hemithorax with its apex-base axis shifted towards the right, having an incidence of around 1 in 12,000 pregnancies (1, 2). Not all cases of the heart being located in the right thoracic cavity are referred to as dextrocardia (3). In cardiac dextroposition, the heart is also located in the right thoracic cavity; however, it should be distinguished from dextrocardia. In dextroposition, the heart shifts to the right thoracic cavity due to extracardiac factors, whereas in dextrocardia, the heart develops in the right thoracic cavity due to intrinsic factors (1). A heart in the right thoracic cavity with a leftward axis indicates a mechanically shifted heart rather than dextrocardia (3).

On a simple basis, the primitive cardiac tube, which consists of consecutive sections of sinus venosus, atrium, ventricle, bulbus cordis, and truncus arteriosus, develops into the human heart (1). The primitive cardiac tube later loops to the right (forming D-loop) or to the left (forming L-loop) on approximately the 23rd day of gestation (1, 4). The morphologic right and left ventricles develop from the bulbus cordis and the bulboventricular loop, respectively. Therefore, the direction of the loop determines the position of the ventricles (1). Incomplete rotation of the L-loop to the right hemithorax and failure of the D-loop to migrate to the left hemithorax result in dextrocardia.

Dextrocardia is mostly asymptomatic by its own; therefore, it is mostly detected incidentally through echocardiography (2, 3). Dextrocardia is frequently associated with other congenital visceral anomalies (3). The term situs is used to express both the asymmetric configuration of the viscera and the heart itself and its chambers by their asymmetric configurations. Focusing on the orientation of viscera in the case of dextrocardia, 3 types of situses have been defined: situs solitus, situs inversus totalis, and situs ambiguous (1). *In situs* solitus, all viscera are in their

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normal place, but the heart is located in the right hemithorax. Whereas in the situs inversus totalis, the thoracic and abdominal viscera are the mirror image of normal. On the other hand, *in situs* ambiguous, the viscera are randomly distributed in the abdomen and thoracic cavity (5). Situs solitus with dextrocardia is frequently referred to as isolated dextrocardia (6). The prevalence of congenital heart anomaly is more common in patients with isolated dextrocardia than those with situs inversus totalis (7). Dextrocardia is also a finding in patients with Kartagener syndrome, apart from the anomalies previously mentioned. Kartagener syndrome is a disorder of ciliary motility with an autosomal recessive inheritance. In 50% of cases, it is accompanied by situs inversus totalis, suggesting that dextrocardia may be an outcome of abnormal arrangement of viscera during the embryonic period (2).

The aim of this case report is to represent a patient with isolated dextrocardia.

CASE REPORT

A 54-year-old female patient was admitted to the emergency room after complaining of nausea, vomiting, fatigue, and tremors. However, she didn't complain from chest pain. Medical evaluations revealed that bradycardia developed after using propranolol for panic attacks. Physical examination revealed rhythmic first heart sound (s1) and second heart sound (2) without murmurs and symmetrical chest expansion at respiration. The patient was observed to be well-oriented and fully cooperative. An electrocardiogram (ECG) was performed in the emergency department, revealing intermittent Wenckebach's atrioventricular (AV) block and sinus bradycardia with normal sinus rhythm (Figure 1). Therefore atropine 1 mg was administered. The patient was then transferred to the coronary care unit (CCU) due to suspicion of non-ST elevation myocardial infarction (NSTEMI). Sequential troponin tests revealed values of 25 pg/mL, 257 pg/mL, and 637 pg/mL, respectively. While the initial value was within the normal range (<40 pg/mL), the elevated subsequent values and upward trend indicate NSTEMI (8).

Wenckebach's AV block was observed during monitoring in the CCU. The 24-hour Holter monitoring recorded minimum, mean, and maximum heart rates of 55, 78, and 92 beats per minute, respectively. The transthoracic echocardiogram revealed moderate tricuspid regurgitation, and the estimated pulmonary artery pressure was measured at 55 mmHg, indicating elevated pressure. Due to the presence of Wenckebach's AV block and NSTEMI, the patient underwent angiography. Spontaneous coronary artery dissection (SCAD) was seen on the distal part of the left anterior descending artery (LAD). Angiography revealed no stenosis in the coronary arteries that could lead to myocardial infarction (MI). Nevertheless, it was observed that the left coronary artery deviated to the right and vice versa, raising suspicion of dextrocardia (Figure 2). The chest radiography confirmed the presence of dextrocardia (Figure 3). Due to the remission of life-threatening conditions and dextrocardia posing minimal risk, the patient was discharged (2). However, due to the onset of epigastric pain and dyspnea, another ECG was performed, revealing a negative T wave in leads V1 and V3. Newly developed symptoms were remitted, and the patient was discharged with a prescription for perindopril-hydrochlorothiazide, benidipine, and atorvastatin, along with a follow-up recommendation.

The right anterior oblique view shows the occluded LAD and a non-critical stenosis of the diagonal artery.

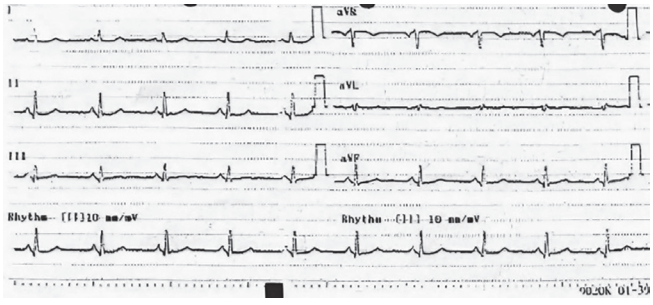

Figure 1: ECG performed in the emergency room showing that ST-T wave changes in leads V1-V3. ECG taken at 25 mm/s, 10 mm/mV.

ECG: Electrocardiogram

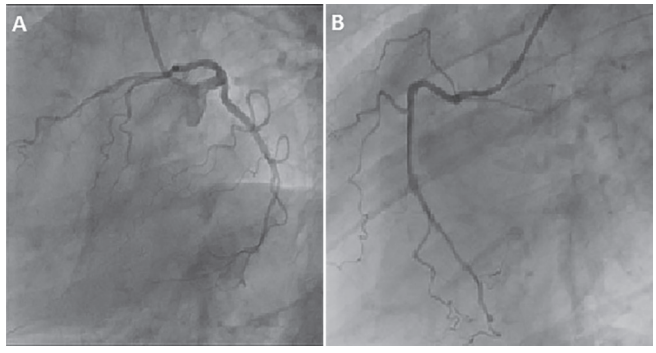

Figure 2: Left coronary angiogram (A) and right coronary angiogram (B) showing deviated coronary arteries.

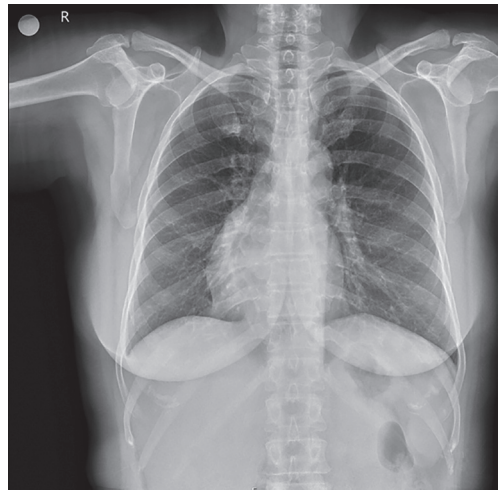

Figure 3: Radiography of the patient showing the heart located in the right thoracic cavity.

DISCUSSION

Dextrocardia can remain asymptomatic throughout life and generally has a good prognosis (2, 9). Notably, there is a case in the literature of a patient being diagnosed after the age of 100 (9). In this case, our patient remained unaware of her condition until the age of 54 years. Moreover, in cases of dextrocardia accompanied by other congenital anomalies, symptoms may vary depending on the associated anomalies (2). Since most cases are asymptomatic, patients are typically diagnosed incidentally through echocardiography and chest radiography performed for other reasons (9). The altered anatomy of the heart in dextrocardia can pose great challenges, particularly in patients requiring surgical interventions such as cardiac catheterization, transcatheter aortic valve replacement, ablation for arrhythmias, pacemaker insertion, or placement of defibrillators. This situation necessitates that physicians consider the abnormal positioning of the heart and other organs during invasive and surgical procedures. Therefore, a multidisciplinary approach is recommended in managing patients with dextrocardia, emphasizing the importance of extra attention in planning the treatment (10).

The malpositioning of the heart can increase susceptibility to specific arrhythmias, such as atrial fibrillation and atrial flutter, which may lead to later-onset complications (11). Naaraayan et al. (10) reported that according to the National Inpatient Sample database, the prevalence of arrhythmia in patients presented to the hospital with dextrocardia is 44.62%, whereas in patients without dextrocardia, this prevalence is 26.69% with a p-value of <0.001. Therefore, it is important that physicians further examine patients with arrhythmias to determine if dextrocardia is present.

Beta-blockers are the most common cause of bradycardia, causing the production of slow action potentials and conduction of atrioventricular impulses (12). A retrospective cohort study (13) revealed that the most common reason for unplanned hospitalizations among elderly patients due to adverse drug reactions was bradycardia, with beta blockers and digoxin being the most common culprits. In another study (14), it was reported that cardiac iatrogenic complications are a significant factor in admissions to intensive cardiac care units, as was the case with our patient. They reported that 91% of these cases were associated with bradyarrhythmia caused by anti-arrhythmic agents such as beta-blockers. Whereas beta-blockers are known to be responsible for arrhythmias in 9.5% of cases and ischemic heart diseases in 38.8% of cases, our patient suffered from both arrhythmia and NSTEMI (15).

The development of MI does not necessarily require an obstruction or stenosis in coronary arteries (16). MI occurring with less than 50% stenosis in the coronary arteries is referred to as myocardial infarction with non-obstructive coronary arteries (MINOCA), responsible for 10% of acute coronary syndrome cases. Conditions such as coronary spasm, SCAD, and coronary microvascular dysfunction may lead to MINOCA (17). SCAD was observed in this patient, leading to MI. SCAD cases are predominantly observed in females, with a rate of 90% (18). MI was responsible for elevated troponin levels in our patient. However, elevated troponin levels do not specifically indicate MI. Several conditions, such as sepsis, hypovolemia, atrial fibrillation, and renal failure, can also cause an elevation in troponin levels (19). This was not the case with our patient. However, other causes of elevated troponin levels should be considered during diagnosis.

In patients with isolated dextrocardia, no treatment or lifestyle changes are required, and they can lead a normal life. However, in cases of dextrocardia accompanied by other congenital anomalies such as tetralogy of Fallot, severe valvular abnormalities, and structural heart defects, surgical intervention should be considered (2). In this case report, we presented a patient with undiagnosed dextrocardia until the age of 54 years. Since dextrocardia is often asymptomatic, it may be overlooked in radiography if not specifically assessed. Such cases are relatively common in the literature. Consequently, physicians should consider dextrocardia in their patients and be aware of its important role in the diagnostic and treatment process.

Footnote

Ethics Committee Approval: *N/A*

Informed Consent: *Informed consent was obtained from the patient.*

Conflict of Interest: *One author of this article, Dengiz Koray Şahintürk, is a member of the editorial board of the Turkish Medical Student Journal. However, he did not take part in any stage of the editorial decision of the manuscript. The editors who evaluated this manuscript are from different institutions. The other authors declared no conflict of interest.*

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