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MYOCARDIAL INFARCTION IN PATIENT WITH SITUS INVERSUS TOTALIS – PROBLEMS IN DIAGNOSING

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Abstract: *Introduction.* Situs inversus totalisis defined as a total inversion-transposition of all thoracic and abdominal organs, so the position of organs in these cavities is like a mirror image of normally positioned organs.

Case report. The study presents a case of a 49-year-old female with chest pain and hemodynamic instability. She was hospitalized at University Clinical Center of Kragujevac where she passed away two days later. During hospitalization, determined the condition of situs inversus totalis, but she was not diagnosed with myocardial infarction. After autopsy it was concluded that the cause of death was myocardial infarction

Conclusion. In patients with situs inversus totalis special attention should be paid to conducting and interpreting diagnostic procedures and conducting therapeutic procedures as well, since inverse anatomy of some structures demands different approach than usual.

Keywords: situs inversus, myocardial infarction, diagnosis, autopsy.

INTRODUCTION

Situs inversus totalis (SIT) is defined as a complete inversion-transposition of all thoracic and abdominal organs, so the position of organs in these cavities looks like a mirror image of normally positioned organs [1,2]. SIT is a rare, congenital condition with the incidence of 0.02%, more frequent in females than in males with the ratio of 3:2, and it isn't associated with race [3,4]. The structure and functioning of all organs are preserved, so SIT is asymptomatic condition most frequently determined by chance during some diagnostic procedures, such as electrocardiography (ECG), roentgenography (Rdg), ultrasound (US), computerized tomography, and sometimes it's discovered postmortem during autopsy [5]. About 25% of patients with SIT has Kartagener syndrome which belongs to the group of primary ciliary dyskinesia and is characterized, besides SIT, by bronchiectasis and paranasal sinusitis [6,7]. The incidence of some diseases in people with SIT is the same as with other people [4].

CASE REPORT

Clinical summary

49-year-old female was hospitalized at University Clinical Center of Kragujevac because of the sensation of a rapid heartbeat, chest pain, electrocardiographically verified abnormal heart rhythm (supraventricular tachycardia) and hemodynamic instability. ECG report showed sinus rhythm, a frequency of 153/min, reduced amplitude of R in precordial leads and reduced ST depression in II, III, and aVF. Blood pressure was 60/40 mmHg and oxygen saturation 93%. Rdg of lungs indicated pronounced reticulonodular interstition on both sides; costophrenic sinuses were clear, hemi diaphragms were clearly contoured, heart shadow - condition situs inversus (Fig. 1). On the second day of hospitalization, despite cardiopulmonary reanimation, cardiopulmonary arrest ensued.

Autopsy report

Interior report shows inversion of all organs of thoracic cavity as in a mirror. Upon opening thoracic

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cavity, dextraposition of heart, right lung with two lobes and left lung with three lobes are noticed. Right ventricle is with a very hypertrophic muscle layer up to 26mm, with reduced ventricular space. Aorta comes out of the right ventricle, with the auricle 60mm wide, with thin, functional valves. Aortic arch is bended to the right, and aorta's wall is entirely inflexible with signs of advanced atherosclerosis. Right atrial lumen is reduced. There is a bicuspid mitral valve between right atrium and ventricle, which is reduced but functional. Left ventricle is dilated, wall thickness is up to 6 mm, and ventricular cavity of the left heart is twice as big as ventricular cavity of the dominant right heart (Fig. 2). Pulmonary artery emerges from the left ventricle; it's 62 mm wide, with thin, functional valves. Left atrium is dilated. There is a tricuspid valve between left ventricle and left atrium. Coronary arteries have inflexible walls with separate, up to 3 mm thick, atherosclerotic plaques. On the right inverse descending coronary artery, atherosclerotic plaque almost completely closes blood vessel lumen, with fresh bleeding present in the plaque. The inversion of all organs as in a mirror is also noticed in abdominal cavity (Fig. 3). Liver is on the left, below left diaphragmatic cupola, so the large lobe of liver is on the left. Spleen is on the right, below right diaphragmatic cupola. Kidneys are fine-grained surfaces, and there are pus and calculi in renal pelvises. Stomach is also on the right, and appendix is in the left part of abdominal cavity, about 15 cm long.

After autopsy it is concluded that the cause of death is myocardial infarction (Fig. 4), which is the result of difficult and long-lasting atherosclerotic heart and blood vessels disease.

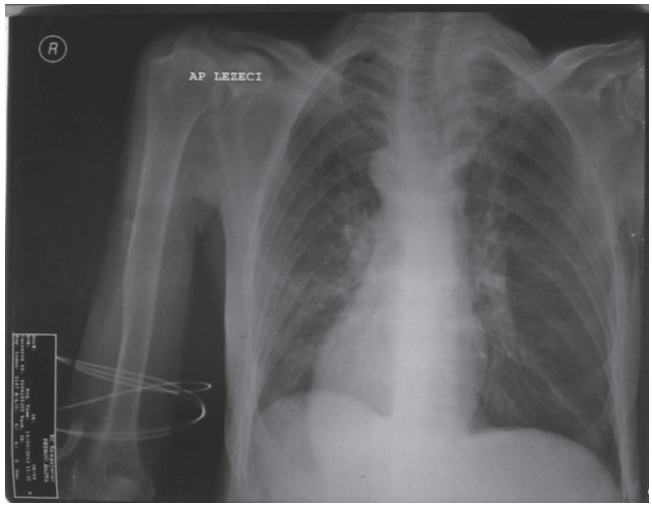

Figure 1. Rdg thorax: heart shadow in the right half of thorax.

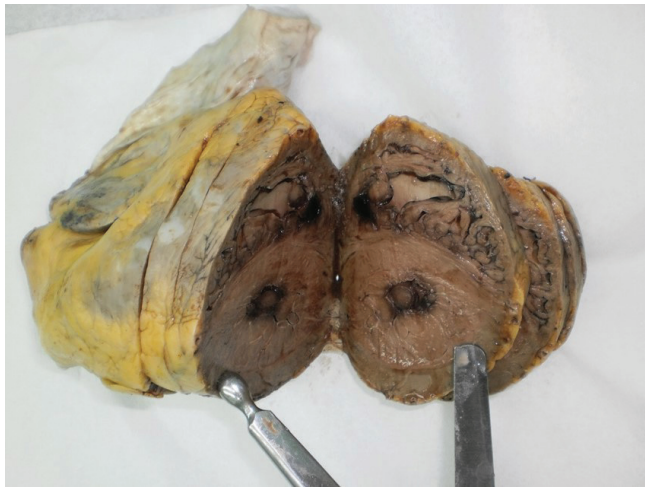

Figure 2. Cross section of the myocardium, heart hypertrophy.

Figure 3. The inversion of all organs as in a mirror in abdominal cavity.

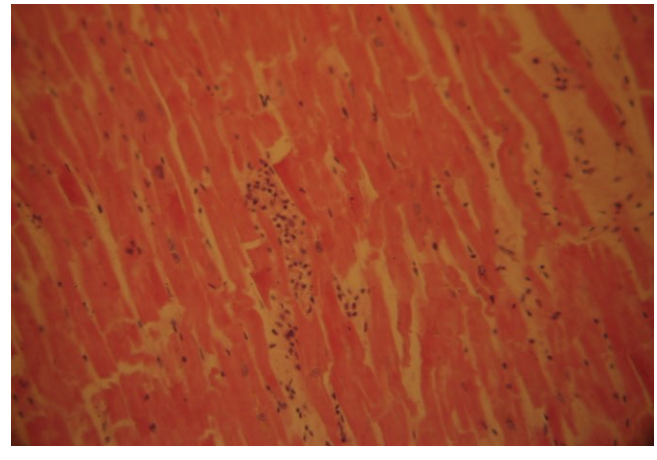

Figure 4. Myocardial infarction.

DISCUSSION

Etiopathogenesis of SIT is even today insufficiently interpreted. It has autosomal recessive inheritance, whereby several genetic mutations have the role in etiopathogenesis. During the embryological development, a 270-degree clockwise rotation instead of normal 270 degree anti-clockwise of the developing thoracic-abdominal organs results in mirror image positioning of the abdominal and thoracic viscera [8].

Situs inversus can be classified as situs inversus with levocardia and situs inversus with dextrocardia. Situs inversus with levocardia is a condition where heart axis passing through base and apex is oriented to the left, vena cava inferior and right atrium are on the opposite side [9]. Dextrocardia is a condition where heart axis is oriented to the right and it is present in SIT in about 39% of patients, it also appears in situs solitus in about 34%, and in about 26% in situs ambiguous [2]. Our patient had dextrocardia, extreme hypertrophic right ventricle with aorta and dilated left ventricle with pulmonary artery coming out of it. Coronary arteries showed signs of advanced atherosclerosis, with bleeding in atherosclerotic plaque. Hypertrophy of the right ventricle, left heart dilatation, and atherosclerotic alterations are explained by the long-term untreated hypertension. Lung's anatomy in patients with SIT is also altered and in accordance with the inversion of all thoracic organs. Our patient had a left lung with three lobes, the right one with two lobes.

Myocardial infarction of our patient wasn't diagnosed on time due, and SIT shouldn't be influential in determining diagnosis. Literature describes problems associated with diagnostic and therapeutic procedures in patients with organs' inversion. Correct interpretation of ECG can help in diagnosing dextrocardia or SIT. Our patient's ECG didn't completely show signs of dextrocardia, but only reduced R wave in precordial leads, which wasn't enough for determining real diagnosis. Only after Rdg of thorax, inversion of thoracic organs with heart's dextraposition could be confirmed. Thorax US, abdomen US or computerized tomography can confirm SIT, which reduces the chance of wrong diagnoses and eases some therapeutic procedures. Alzand *et al.* describes patients with dextrocardia and situs inversus totalis presenting with acute chest pain irradiating to the right arm [10]. He stresses the importance of performing a reversed electrocardiogram in patients with dextrocardia, for correct diagnosis of an acute anteroseptal myocardial infarction.

Symptomatology of certain organs' systems can be manifested on unexpected body parts, so one should be cautious when interpreting results. In SIT patients, inverse anatomy of some structures demands different diagnostic and therapeutic approach than usual, which is described in several case reports of laparoscopic cholecystectomy [11,12] and laparoscopic appendectomy [13]. Since organs inversion doesn't lead to the inversion of all structures, but certain nerves stay in normal anatomic position, the pain will be projected on not only to the side with organs inversion, but to the opposite side as well [13]. Our patient had abdomen and urinary tract US reports, and transposition of all abdominal organs was established with renal parenchyma damage and calculus.

With SIT patients it's a great challenge to perform angiography, since the largest problem is to differentiate between coronary arteries auricles due to inverse positions [14]. It is also challenging to reconstruct blood vessels of inverse organs in order to adjust them to normal anatomic structures [15].

In conclusion, generally, the problems of unplaced diagnoses, wrong diagnoses, or complications during diagnostic and therapeutic procedures can be associated with doctors' mistakes. Therefore, it is important for doctors to be aware of SIT existence and to confirm it on time. If there are limitations in using diagnostic procedures to confirm the existence of the condition, it should be taken into account that SIT can be confirmed by thorough physical examination including auscultation and percussion of thorax and abdomen with adequate ECG.

Conflict of interest

The authors declare that they have no conflict of interest.

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