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CASE REPORT

FORENSIC PATHOLOGY - AUTOPSY

Situs inversus totalisli bir olguda spontan koroner arter diseksiyonuna bağlı ani ölüm

Sudden death due to spontaneous coronary artery dissection in a case with situs inversus totalis

Aytül Buğra[1], Ceyhun Küçük[1], Abdul Kerim Buğra[2]

Corresponding author: *Aytül Buğra*
Council of Forensic Medicine, Fevzi Cakmak Mh. Kımız Sk. No:1, 34196, İstanbul, Turkiye
email: aytulsargan@gmail.com

ORCID:
Aytül Buğra: 0000-0001-5640-8329
Ceyhun Küçük: 0000-0001-5372-3753
Abdul Kerim Buğra: 0000-0001-9575-0100

ÖZET

Situs inversus totalis, iç organların karakteristik olarak normal lokalizasyonlarının ayna görüntüsünde olduğu bir konjenital anomalidir. Asemptomatik olabilir ve normal yaşamla uyumlu olabilir. Konjenital kardiyak anomaliler eşlik edebilir. Amacımız aniden ölen 31 yaşındaki kadında tesadüfi bir situs inversus totalis olgusu bildirmektir. Otopside, situs inversus totalis ile spontan koroner arter diseksiyonunun bir arada bulunduğunu görülmüştür. Yapılan literatür taramasında situs inversus totalis'in spontan koroner arter diseksiyonu ile birlikteliği daha önce bildirilmemiştir.

Anahtar Kelimeler: Ani ölüm, koroner arter diseksiyonu, situs inversus totalis, dekstrokardi.

ABSTRACT

Situs inversus totalis is a congenital anomaly in which the visceral organs are characterically in the mirror image of their normal localisations. It can be asymptomatic and compatible with normal life. Congenital cardiac anomalies can be seen in this condition. Our aim is to report an incidental finding of situs inversus totalis in a 31-year-old female with sudden death. Autopsy studies showed that the coexistence of situs inversus totalis and spontaneous coronary artery dissection. As much as we know, the coexistence of situs inversus totalis with spontaneous coronary artery dissection has not been reported before.

Keywords: Sudden death, coronary artery dissection, situs inversus totalis, dextrocardia.

INTRODUCTION

Situs inversus totalis is a congenital anomaly in which the visceral organs are on the other side of the body, instead of their normal localizations. It is firstly described by Marco Severino in 1643.

The incidence of the condition is approximately 1 in 10000 (1). Although most of them are sporadic, it has been shown that 20-25% of the cases have mendelian inheritance and are generally inherited as autosomal recessive (2). Many people with situs inversus are unaware of their anomaly until a radiological exploration or is an incidentally finding in autopsy. Congenital cardiac anomalies occur in 3-5% of individuals with situs inversus, and the most common anomaly is the double outlet right ventricle (3).

Spontaneous coronary artery dissection (SCAD) is spontaneous seperation of coronary artery wall in the absence of iatrogenic injury, trauma, or significant atherosclerosis. SCAD is the cause of sudden cardiac death in 0.5% on autopsy reports (4).

As much as we know, the coexistence of situs inversus totalis with spontaneous coronary artery dissection has not been reported before. In this

1 Morgue Department, Council of Forensic Medicine, Istanbul Turkiye
2 Department of Cardiovascular Surgery, University of Health Sciences, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Hospital, Istanbul, Turkiye

case report, a sudden death due to spontaneous coronary artery dissection in a young woman with situs inversus totalis was presented.

CASE

A 31-year-old woman fell sick at home and died in the hospital where she was taken. The case had no known disease, alcohol use, smoking, or history of surgery before. She also had a 6-year-old healthy child. During the autopsy, no apparent macroscopic pathological feature was observed, and her body was 160 cm in length and 60 kg in weight. The toxicological examination was clear. When the skull was opened, no feature was observed in the cerebrum, cerebellum, and brain stem. When the thorax was opened, the heart appeared in a dextrocardiac position, and the aorta was curved to the right (Figure 1). The right lung had two lobes and

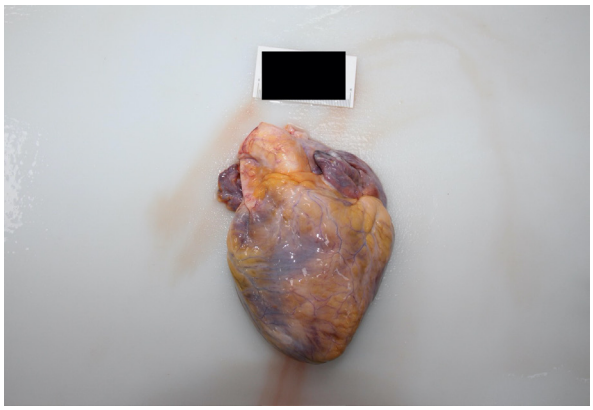


Figure 1. Aortic arch arising to the right

the left lung three lobes. In the abdomen, the liver was left sided, and the spleen was right. The stomach great curvature was right sided. There was no congenital cardiac anomaly. In the transvers sections of the coronary arteries, the circumference of a 15 mm segment of the left main coronary artery from the ostium has hemorrhage (Figure 2).

No macroscopic pathological features were observed in the heart valves, cavities, and myocardial sections. Histopathological examination of the sample taken from the coronary artery revealed separation in the artery wall and hemorrhage in this area (Figure 3). There was no inflammatory cell infiltration around the vessel and in the adventitia, and no evidence to support fibromuscular dysplasia in the coronary artery wall with Masson trichrome stain. There were no specific findings in the histopathological examination of the other organs. The cause of sudden death was hearth failure due to coronary artery dissection.



Figure 2. Transverse section of the hemorrhagic-segment of the coronary artery (arrow)

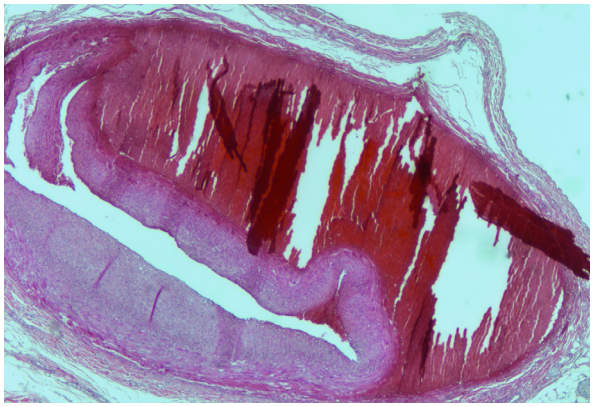


Figure 3. (H&E, x40): Intimal thickening with dissection and bleeding in the coronary artery wall between media and intima

DISCUSSION

Situs inversus totalis is more common with dextrocardia and congenital heart disease is seen in 3-5% in situs inversus with dextrocardia. Right aortic arch is present in 80% of these patients. Cases are usually asymptomatic and have a normal life (5). Our case was also in her 30s and died suddenly with sickness while continuing her life normally. In our case, the aortic arch was on the right side, and there was no congenital heart disease. The rate of coronary anomalies among all cardiac anomalies in situs inversus totalis cases is uncertain as very few cases have been reported. In addition, because of the rarity of this situation, the prevalence and types of late complications cannot be predicted. In the study by Kim et al., the left and right coronary ostium originate from the left coronary cusp (3). In our case, spontaneous dissection was observed in the histopathological examination of the left main coronary. In our literature review, we did not find any case in which spontaneous coronary artery dissection and situs inversus totalis were seen together.

Spontaneous coronary artery dissection is, by definition, a non-traumatic and non-iatrogenic separation of the coronary artery wall. This separation can be between intima and media, or between media and adventitia. Dissociation within the arterial wall can result in the formation of an intramural hematoma that compresses the arterial lumen, decreased antegrade blood flow, and subsequent myocardial ischemia or infarction (6). The true incidence and prevalence of SCAD in the general population are uncertain. According to recent studies, SCAD was detected in 1-4% of all acute coronary syndromes by coronary angiography and was found to be the cause of sudden cardiac death at a rate of 0.5% in autopsy reports. In our case, it was detected during the autopsy as a result of sudden death. More than 90% of SCAD cases are women with little or no cardiovascular risk factors (4). The cause of SCAD is usually multifactorial with underlying predisposing arteriopathies, environmental triggers, and stress factors. Fibromuscular dysplasia is the most common condition associated with SCAD. Pregnancy, infertility treatments, postmenopausal hormone treatments, oral contraceptives may cause weakening of the vessel wall. Excessive physical activity, high dose hormone therapy, intense emotional stress, and sympathomimetic medications can accelerate the dissection process (4,8-15). Our case died in the hospital where she was hospitalized due to her sickness at home. According to the information received from her relatives, she does not have any chronic disease and hasn't known whether she received hormonal therapy. No abnormalities were detected in the toxicological examination. SCAD commonly affects normal or weakened artery walls and causes anterograde and retrograde separation. Average length is reported to be 45 mm (7). In our case, the dissected arterial segment length was 15 mm. In spontaneous coronary artery dissection, a mixed type of inflammatory cell infiltration rich in eosinophils can be seen around the vessel (16). In our case, there was no inflammatory cell infiltration around the vessel wall. The case was admitted to the hospital immediately after the onset of symptoms and died within hours. Since death occurs immediately after dissection, we may not have seen inflammation.

In conclusion, our case is a case not previously described in the literature, in which spontaneous coronary artery dissection which causes sudden death coexistence with situs inversus totalis. In sudden death cases developing at a young age, coronary artery dissection should always be kept in mind, especially in women. In the light of autopsy studies and postmortem histopathological examinations, a definite diagnosis can be made in sudden death cases. Thus, the true incidence of the cases can be revealed more clearly by contributing to the literature.

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