Case Report

Situs inversus with congenital absence of left circumflex artery and a superdominant left-sided right coronary artery: A case report

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\textbf{ABSTRACT}

Situs inversus with a coronary artery defect is an extremely rare congenital anomaly, which is often only incidentally detected during cardiac imaging studies, particularly when the patient is complaining of chest pain. We present a case study of a 37-year-old man who presented with a 3-month history of chest pain. A chest X-ray revealed dextrocardia with stomach gas in the lower right diaphragm. A CT examination showed that the right lung had 2 lobes, and the left lung had 3 lobes. This confirmed that the patient had dextrocardia with situs inversus, where the heart, organs and coronary arteries are inverted to the opposite side of the body. It is also known as a mirroring image, as the arrangement of the anatomy is unaffected. The patient's right coronary artery was prominently inverted to the left side, while the left main coronary artery was inverted to the right side and continued toward the anterior interventricular sulcus, eventually becoming the left anterior descending (LAD) artery. The left circumflex artery was absent in the ativoventricular groove. There was also a plaque in the right-sided LAD with no significant stenosis. The patient received conservative medical therapy and had frequent follow-ups to check for potential complications.

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\section*{Introduction}

Situs inversus is a rare congenital condition affecting approximately 1 in 10,000 people, in which the major visceral organs are arranged in a mirror image of their normal positions, which is why it is also known as a mirroring image. People with situs inversus are likely unaware of their condition until they seek medical attention \cite{1}. Nevertheless, the incidence of coronary artery disease (CAD) in situs inversus patients is...
the same as in the normal population [2]. The congenital coronary artery anomaly is rarely observed, diagnosed incidentally, and has only been reported in 0.64–1.3% of patients undergoing angiography [3]. However, the absence of the left circumflex artery (LCx) along with a superdominant right coronary artery (RCA) is an extremely rare anomaly. Yamanaka and Hobb (1990) stated that out of 126,595 patients who underwent coronary angiography between 1960 and 1988, just 4 patients (0.003%) were found to have this condition [4]. Additionally, Srinivasan (2008) found that an absence of the LCx along with a superdominant RCA was found in only 0.067% of 1,495 patients who underwent CT coronary angiography [5].

Case description

A 37-year-old man presented with a 3-month history of chest pain. There was also no significant history of comorbidities found in the patient's medical records. The patient's blood pressure was 110/80 mm Hg, and his heart rate was 84 beats/minute. A chest X-ray and a CT coronary angiogram were performed for initial assessment and to assess the coronary artery, respectively.

The chest X-ray revealed dextrocardia with stomach gas in the lower right diaphragm. A CT examination showed that the right lung had 2 lobes, and the left lung had 3 lobes, confirming that the patient had dextrocardia with situs inversus (Fig. 1).

In this case, because of situs inversus, all heart chambers and coronary arteries are inverted, although the anatomy of the chambers and arteries are unaffected (Fig. 2). The RCA is inverted to the left side and supplies the right atrium and right ventricle. The prominent, left-sided RCA crosses the posterior interventricular groove to the left ventricle and bifurcates into the posterolateral branch (PLB), which supplies the posterior wall, and the posterior descending artery (PDA) branch, which supplies the inferior wall of the left ventricle.

The left main coronary artery (LMCA) is inverted to the right side, continues towards the anterior interventricular sulcus, and becomes the right-sided left anterior descending (LAD) artery. The diagonal (D1) artery becomes the first branch of the right-sided LAD artery and supplies the anterior and lateral part of the left ventricle (Fig. 2). There is no LCx present in the left atrioventricular groove. The small portion of LCx territory in the coronary sulcus is supplied by the D1 artery (Fig. 3).

Our study also revealed a calcified plaque in the proximal right-sided LAD that showed non-significant stenosis determined by a semi-quantitative assessment. Our findings determined that the patient had non-obstructive CAD which did not require revascularization therapy (Fig. 4).

Discussion

A person with a normal anatomical configuration has 3 main coronary arteries: the RCA, the LAD, and the LCx. The RCA originates from the right coronary sinus of Valsalva, which passes anteriorly to the right. It occurs between the right auricle and the pulmonary artery and descends vertically into the right atrioventricular sulcus from which it supplies the right atrium and right ventricle. It also supplies the posteroinferior third of the interventricular septum and some posterior parts of the left ventricle [6]. The LMCA arises from the left coronary sinus of Valsalva and separates into the LAD and LCx. The LAD continues toward the apex of the heart in the anterior interven-
The congenital coronary artery anomaly is rarely observed, diagnosed incidentally, and has only been reported in 0.64–1.3% of patients undergoing angiography [3]. However, the absence of the LCx along with a superdominant RCA is an extremely rare anomaly [8]. Moreover, in the absence of the LCx, the RCA grows into a prominent (or superdominant) vessel, which crosses the posterior interventricular groove and bifurcates into the PLB and PDA branches. From there, it supplies the posterior and inferior walls of the left ventricle that are normally supplied by the LCx [7]. It is imperative that this anomaly is identified, as it may mimic atherosclerotic CAD [9].

The accepted hypothesis for this anomaly is that it is a steal phenomenon [6,9,10]. This phenomenon, which can be triggered by increased physical activity, causes an increased arterial demand from the area which is normally supplied by the interventricular sulcus and gives diagonal (D1) branches. D1 descends diagonally across the anterior surface of the left ventricle and supplies the anterior wall of the left ventricle as well as the interventricular septum. Lastly, the LCx courses towards the left and supplies the posterior and inferior parts of the left ventricle [6,7].

Fig. 2 – CT coronary angiography volume-rendering technique images showing the inverted chambers and arteries. (A) The anterior view shows the left ventricle on the right side and the right ventricle on the left side. (B) The top view shows the RCA on the left side and the LMCA on the right side. (C) The prominent, left-sided RCA supplies the interventricular sulcus and the posterior and inferior walls of the left ventricle, while (D) the right-sided LAD supplies the anterior and lateral walls of the left ventricle.
LCx territory and results in transient ischemia to other coronary arterial territories [9,10,11]. In this study, the patient presented with chest pain, which was assumed to be transient ischemia caused by increased oxygen demand.

Over the past years, invasive coronary angiography has been considered as a standard for the evaluation of coronary artery anomalies. However, CT coronary angiography is recently emerging as a reasonable first-line tool for the assessment of coronary anatomy [11,12]. CT coronary angiography is suitable to detect the anatomical course of coronary artery anomalies and define their relationship to other cardiac and non-cardiac structures. Furthermore, the 3-dimensional post-processing rendering systems and multiplanar reconstruction (MPR) techniques provide better result in coronary artery evaluation [13].

This patient did not require any specific treatment. The plaque in the right-sided LAD was also judged to not require interventional therapy as the stenosis was non-significant. The patient received conservative medical therapy and had frequent follow-ups to check for potential complications.

Fig. 3 – CT coronary angiography volume-rendering technique images showing the right-sided LMCA continuing as the right-sided LAD in the anterior interventricular groove, with the D1 and D2 branches supplying the anterior and lateral walls of the left ventricle.

Fig. 4 – The multiplanar reconstruction (MPR) technique showing calcified plaque in the proximal LAD with nonsignificant stenosis.
Conclusion

In conclusion, situs inversus with a congenital absence of the LCx is an extremely rare and distinctive anomaly. It is often only detected incidentally during cardiac imaging studies, particularly if the patient is suffering from chest pain. Even though the absence of the LCx is considered a minor problem and the plaque in the proximal LAD causes non-significant stenosis, clinicians should nevertheless be aware of and be able to identify anomalies such as these in order to ensure the best course of treatment.

Consent and ethic committee approval

Written consent has been obtained from the patient as there is no patient identifiable data included in this case report. This study has met the ethical principle and already got approval from Research Ethics Committee from Dr. Soetomo General Hospital, Surabaya.

Patient consent

Informed consent obtained for publication of a case report: Written informed consent was obtained from the patient for the publication of this case report.

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