

Anomalous origin of left coronary artery arising from the right coronary cusp presenting with chest discomfort and syncope on physical exercise

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= Abstract =

Anomalous origins of coronary arteries are a rare type of disease among children. These anomalies can be categorized into 3 types according to the anatomical relationship of the aorta and pulmonary trunks. Among these types, the interarterial type, as observed in our case, needs early diagnosis and treatment, because it can increase the risk for the patient, causing sudden cardiac death in young individuals. Although there are controversies concerning the management of anomalous origins of the left coronary artery (LCA) in children, the result can be very beneficial, if treated accurately. Three well-known methods for correction of anomalous origins of LCA are re-implantation, coronary arterial bypass grafting (CABG), and unroofing. We report on the case of a 12-year-old girl who had chest discomfort and syncope with physical exercise and was later diagnosed with an anomalous origin of LCA by transthoracic echocardiography (TTE) and heart computed tomography (CT). She underwent a corrective operation by re-implantation, CABG, and unroofing. (**Korean J Pediatr 2010;53:248-252**)

Key Words : Coronary Vessel Anomalies, Coronary Artery Bypass, Acute Coronary Syndrome, Syncope

Introduction

Anomalies of the coronary arteries among children are very rare. In 2001, according to the research of Bozena et al¹⁾, there were only 18 cases of coronary artery anomalies among echocardiogram tests of 62,320 patients, and there was no single case of left coronary artery (LCA) arising from the right coronary artery (RCA). Although anomalous origin of the coronary artery from the opposite sinus of Valsalva is infrequently encountered, it can cause sudden cardiac death in the young, the risk of which depends on the anatomic course²⁾. The interarterial pathway of anomalous origin coronary artery arising from the contralateral sinus and coursing between the aorta and pulmonary trunk

has been reported to be significantly associated with a high risk of sudden cardiac death³⁻⁵⁾. These anomalies are rarely suspected or diagnosed during life, mostly due to lack of clinical suspicions⁶⁾. Nevertheless, given the ability to perform surgical correction, timely diagnosis and prevention are very important²⁾.

We present a case of LCA, with an anomalous origin of the intramural type, arising from the right coronary cusp (RCC) and coursing between the aorta and the pulmonary trunk.

Case report

A 12-year-old girl presented to the emergency department in another hospital after episodes of chest pain and syncope with physical exercise. At the time of admission, she was hemodynamically stable, and mental status was alert and oriented. Physical examination revealed regular heart beat and clear breath sounds. A resting 12-lead electrocardiogram (ECG) showed ST-segment depression in leads II and III, and aVF initially. Laboratory values in-

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cluded a creatinine kinase of 641 IU/L and a creatinine kinase-MB of 59.3 ng/mL, and elevated ratio of 9.3%. A transthoracic echocardiogram (TTE) indicated an anomalous origin of the LCA from the RCA, which was compressed between the aortic root and the pulmonary trunk. Coronary computed tomography (CT) scan was performed, which showed the same as the TTE (Fig. 1). The patient was transferred to our hospital for further evaluation with precise diagnosis.

Upon her arrival at our hospital, she had no chest discomfort. Her pulse rate was 68 beats/min, respiratory rate was 20 breaths/min, and blood pressure was 113/54 mmHg. Physical examination revealed regular heart beat without murmur and clear breath sounds. A 12-lead ECG showed ST-segment depression in the anterior leads. Laboratory values included a normal leukocyte count, a creatinine kinase-MB of 58 IU/L, and a troponin-T of 0.3 ng/mL, which was elevated.

A TTE indicated a LCA originating from the RCC with interarterial course, and a compressed LCA between the aortic root and the pulmonary trunk (Fig. 2). TTE did not reveal any regional wall motion abnormality or morphologic or functional abnormalities of the left ventricle.

The patient underwent an operation for coronary artery correction and re-perfusion on the sixth day of her hospital stay. It was determined by intraoperative transesophageal echocardiogram that the orifice of the LCA was found to be originating from the right coronary sinus, leaning towards the left commissure rather than the orifice of the RCA. The orifice of the RCA was in its normal location.

The course of the LCA was intramural, which passed between the aorta and the main pulmonary artery. We could not directly dissect the left main coronary artery (LMCA). We performed aortotomy and then incised the compressed portion of the LMCA between the aorta and the main pulmonary artery with a 1.5 mm coronary probe. A new ostium of LCA was created in the correct anatomic location by unroofing.

The postoperative condition was stable. An ECG was performed before discharge, which showed normalization of the ST-segments. TTE was performed on the fourth postoperative day. We could see the remains of a proximal

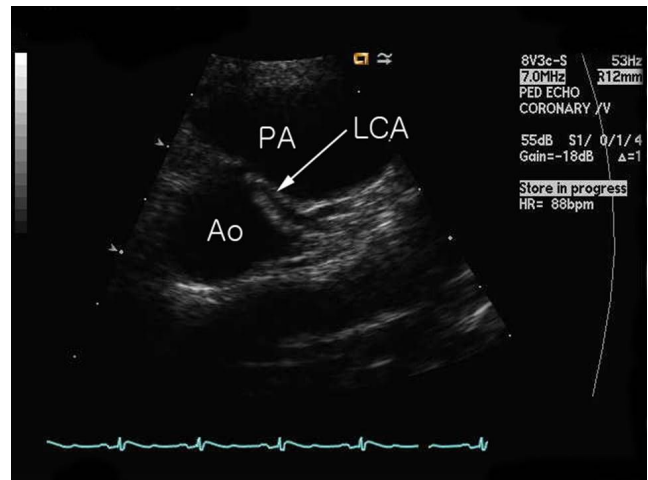


Fig. 2. Preoperative transthoracic echocardiography reveals that the left coronary artery arises from the right coronary cusp with an interarterial course and a compressed LCA between the aortic root and the pulmonary artery. Abbreviations : LCA, left coronary artery; Ao, aorta; PA, pulmonary artery.



Fig. 1. Preoperative coronary computed tomography reveals an anomalous origin of the left coronary artery from the right coronary artery, which is compressed between the aortic root and pulmonary trunk. Abbreviations : LCA, left coronary artery; Ao, aorta; PA, pulmonary artery.

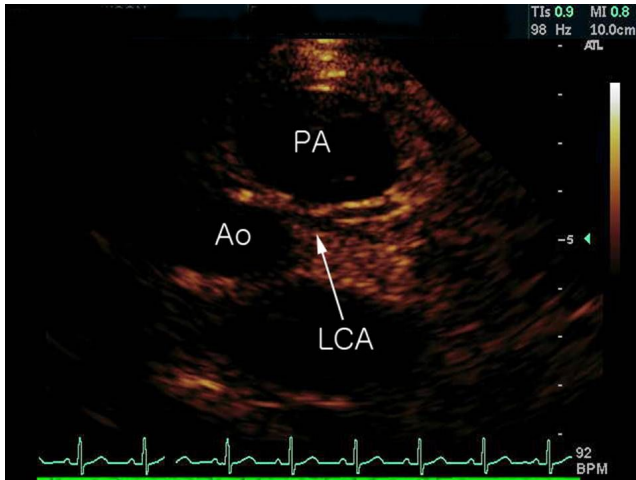


Fig. 3. Postoperative transthoracic echocardiography reveals a proximal narrowing of the left main coronary artery after the operation. Abbreviations: LCA, left coronary artery; Ao, aorta; PA, pulmonary artery.

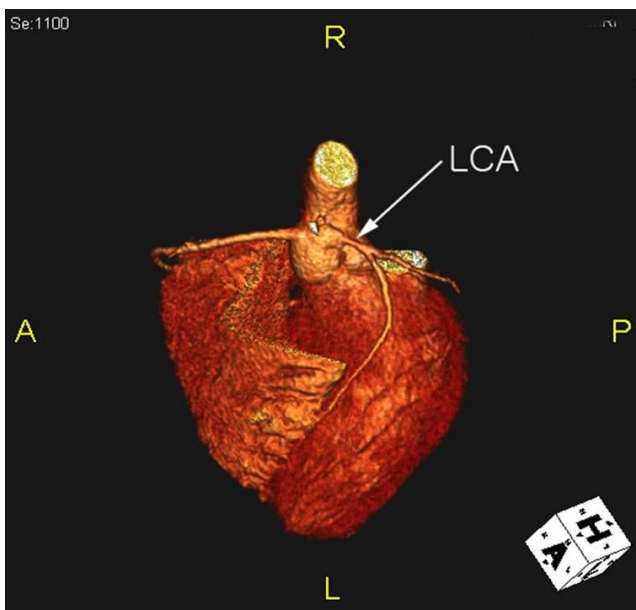


Fig. 4. Postoperative heart computed tomography reveals a proximal stenosis of the left main coronary artery at about 20–30% of the diameter of the LCA after the correction of an anomalous origin of the LCA from the RCC. Abbreviations: LCA, left coronary artery; RCC, right coronary cusp.

LMCA narrowing (proximal: $d=1.9$ mm, distal: $d=3.2$ mm, left anterior descending (LAD): $d=2.7$ – 2.9 mm, left circumflex artery: $d=3.1$ mm) and supra-
valvular narrowing of the ascending aorta ($d=16.5$ mm, Fig. 3). We performed a postoperative coronary CT scan to check for stenosis of the LMCA on the eighth postoperative day. CT scan revealed a mild stenosis of the LMCA along about 20–30% of the

diameter of the LMCA (Fig. 4). The patient was discharged home on the fourteenth day after operation on aspirin and clopidogrel bisulfate.

Although the patient was recovering without any significant post symptoms, the immediate post operative TTE and heart CT revealed stenosis of the LMCA (20–30%). We followed up with a heart CT eight months after the operation, and it revealed progression of stenosis of the LMCA (50–60%).

Ten months after the operation, a treadmill test was performed, the result of which was negative. It was decided to maintain current medications (aspirin and clopidogrel bisulfate) and perform routine check up by TTE, heart CT, and treadmill test.

Discussion

The incidence of an anomalous origin of the coronary artery has been reported by 0.2–0.3% of angiography and 0.3–0.5% of autopsy series^{7, 8}. These anomalies are clinically very important because they can lead to sudden cardiac death in the young³. Anomalous origins of the LCA can be divided into three different types, according to the anatomical relationship of the aorta and pulmonary trunks^{9–11}. Type 1 anomalous origin of LCA is defined as one crossing the anterior free wall of the right ventricle. Type 2A interarterial, courses between the aorta and the pulmonary trunk, type 2B, septal, adopts an intramuscular pathway through the septum. Type 3 (retroaortic), passes to the posterior of the aorta. Among these, type 2A is remarkable, as it is related to exertional angina, syncope and sudden cardiac death¹¹. In 1974, Cheitlin *et al*¹² found many cases of patients with sudden death during physical exercise with anomalies of LCA, and reported that sudden death is more likely when the coronary artery passes between the aorta and the pulmonary artery.

Even in Korea, there have been multiple cases of myocardial infarction while exercising, and some were pediatric cases^{2, 9, 13}. Although a mechanism for sudden cardiac death has not been established in these anomalies, there is clear evidence that coronary arterial flow is compromised by compression of the coronary artery, so-called interarterial entrapment, restriction at the site of its orifice from the sinus of Valsalva, or kinking of the coronary artery itself^{14, 15}. Other causes of sudden cardiac death include the travel length of the intramural course and the angle of the orifice¹⁵.

In diagnosing these coronary artery anomalies, the most important thing is not to “rule out” the anomaly, but to “rule in” the exact location of the origins of the coronary arteries¹⁵⁾. As previously described, diagnostic tools can be used to identify anomalous vessels. However, other three tools have their own challenges; angiography being invasive, CT employing ionizing radiation, and magnetic resonance imaging (MRI), while convenient and precise, costing substantially more. Therefore TTE remains the most commonly available and practical tool to examine these anomalies^{14, 15)}. After ruling in these coronary anomalies, we need an understanding of the factors that determine and regulate myocardial blood flow by using ECG, myocardial perfusion imaging, stress echocardiogram and positron emission tomography.

Early diagnosis and surgical treatment are important in these anomalous origins of coronary arteries, because these coronary artery anomalies can be corrected for a complete recovery^{2, 15)}. Surgical correction of significant coronary arterial abnormalities can be undertaken with excellent results. Re-implantation of the anomalous left coronary artery arising from the pulmonary trunk, and surgical ablation of coronary arterial fistula are part of the routine care for children treated in pediatric cardiac centers. However, there are controversies concerning the management of anomalous origin coronary arteries from the opposite sinus of Valsalva with a course between the arterial trunks. When the coronary artery takes an interarterial course, we can surgically treat these patients by re-implantation, coronary arterial bypass grafting (CABG), and coronary arterial unroofing¹⁶⁻¹⁸⁾. CABG is unlikely to be successful, as the coronary vascular bed at risk is well supplied most of the time, it can lead to injury itself and will not provide adequate perfusion during episodes of ischemia. In re-implantation, it is difficult to mobilize the proximal part of the coronary artery. The unroofing procedure is straightforward, and eliminates ischemic potential by removing the intramural course, at the same time eliminating any contribution from an abnormal coronary arterial orifice. The procedure involves unroofing the anomalous coronary artery into the aortic lumen^{18, 19)}. However, in this patient the orifice of the LCA was found to be originating from the right coronary cusp, leaning towards the left commissure rather than the orifice of the RCA. This was the first case where CABG and re-implantation were undertaken for correction of the anomalous origin of the LCA and unroofing for anatomical

correction of the anomalous location of the orifice of the LCA.

The patient was doing well without complications after the operation, and on medications.

한글 요약

12세 여아에서 운동 중 발생한 흉통 및 실신 - 왼쪽 주 관상동맥의 이상 기시의 진단 및 수술적 치료 1례

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관상동맥의 이상 기시는 소아에서 드문 질환이다. 이러한 이상 기시들은 대동맥과 폐동맥간과의 해부학적인 관계에 의해 3가지 형태로 분류할 수 있다. 우리 환자의 경우와 같이 기형 동맥이 대동맥과 폐동맥사이로 주행하는 경우 젊은 연령에서 급사의 위험도가 증가하기 때문에 빠른 진단과 치료가 필요하다. 관상동맥의 이상 기시를 교정하는 방법으로는 잘 알려진 re-implantation, 관상동맥 우회술과 unroofing의 세가지 방식이 있고, 아직까지 좌 관상동맥의 이상 기시의 치료에 대해서는 많은 견해들이 있으나 일단 수술적 교정이 되면 좋은 결과를 보인다.

우리는 운동 중 발생한 흉통과 실신을 주소로 내원한 12세 여아에서 심초음파와 관상동맥 전상화 단층촬영을 통하여 좌 관상동맥의 우 관상동맥동으로부터의 이상 기시를 진단받고 re-implantation, 관상동맥 우회술, 그리고 unroofing 방법을 통해 치료 받은 1례를 경험하여 이를 보고하고자 한다.

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