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Case Report

Bland White Garland syndrome with aneurysm and collaterals of coronary arteries in an Afghan girl: a case report.

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Abstract

Bland White Garland Syndrome is a very rare congenital heart defect in which the left coronary artery arises abnormally from the pulmonary artery. We present an extremely rare case of Bland White Garland Syndrome with an aneurysm of the left coronary artery in a 14-year-old Afghan girl. The patient was asymptomatic throughout her life except for one attack of exertional chest discomfort. The diagnoses of these anomalies were established by electrocardiography, echocardiography, coronary angiography, and computed tomography of the chest. During her hospital stay and on discharge, she had a stable condition and was referred for surgical management in an advanced setting abroad. Anomalous origin of the left coronary artery from the pulmonary artery may coexist with an aneurysm of the left coronary artery and not exhibit symptoms until adolescence.

INTRODUCTION

Bland White Garland Syndrome (BWGS), also known as anomalous origin of the left coronary artery from the pulmonary artery, is a very rare congenital malformation [1]. The incidence of BWGS is estimated at 1 in 300 000 live births, or 0.25% to 0.5% of all congenital heart disease [1, 2]. The left coronary artery (LCA) in this anomaly originates from the pulmonary artery rather than the aorta. Children typically present with dyspnea, pallor, and a failure to thrive. When collaterals are adequate, symptoms can be absent or relatively minor, allowing growth into adulthood [2, 3]. Adults can sometimes be asymptomatic or, more commonly, have a variety of symptoms such as syncope, chest pain, and sudden death [4, 5]. Echocardiography, coronary angiography, and computed tomography (CT) of the chest are useful tools for the diagnosis of BWGS [3, 5]. When possible, surgery is advised for patients with BWGS. The most popular surgical correction involves direct reimplantation of an anomalous left coronary artery into the aorta by transferring it with a button of pulmonary artery [2, 4]. The patient's survival to their sixth or seventh decade of life without surgery is extremely rare [4]. There is a paucity of data regarding BWGS with aneurysm and collaterals of coronary arteries in asymptomatic adolescents, therefore, such a case is presented.

CASE REPORT

An Afghan 14-year-old girl was referred to the Pediatric Unit of Maiwand Teaching Hospital due to chest discomfort. On arrival, she complained of one attack of localized middle-chest paint that lasted 30 min and was relieved by rest. Her past medical and family history was unremarkable. On general physical examination, a blood pressure of 110/70, a respiratory rate of 20/min, a heart rate of 69/min, and an oxygen saturation of 94% were recorded. By systemic examination of the chest, a soft early systolic murmur (Levine III/VI) was found. No abnormal clinical findings were detected in other systems. Blood investigations were within normal limits. The chest x-ray showed cardiomegaly, with a cardiothoracic ratio of 62.5%, and bilateral costophrenic angles were sharp with no congestion (Fig. 1). The electrocardiography (ECG) showed a heart rate of 69 beats per minute, a normal sinus rhythm, an axis of +50, a PQ of 0.16 s, a QRS of 0.08 s, a QT/QTc of 0.36/0.36 s, and no significant ST segment shift was noted (Fig. 2). On day three of admission, transthoracic echocardiography revealed an ectopic opening of the coronary artery, a blood-stealing phenomenon at the pulmonary artery, and abnormal coronary circulation (Fig. 3), as well as pulsed color doppler confirming abnormal flow within the pulmonary artery. Pulsed-wave doppler showed a left coronary wave pattern (Fig. 4). These echocardiographic findings suggested BWGS. Furthermore, there was all-segment normokinesis, a left ventricular end-diastolic volume (LVEDV) of 72.9 ml, a left ventricular end-systolic volume (LVESV) of 24.9 ml, and a left ventricular ejection fraction (LVEF) of 66.1%. Based on ECG and cardiac echocardiography findings, BWGS was suspected; hence, chest computed tomography (CT) and cardiac catheterization were advised. The chest CT scan of the patient revealed the origin of the left coronary artery from

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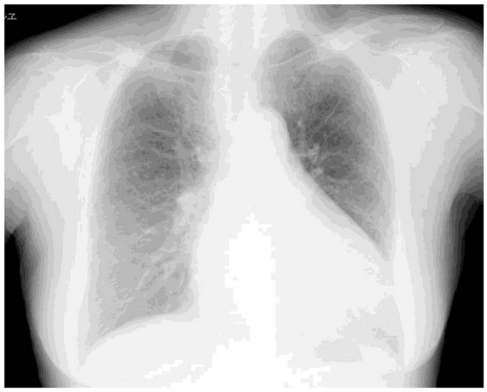

Figure 1. Chest X-ray of the patient shows a cardiomegaly with no lung congestion.

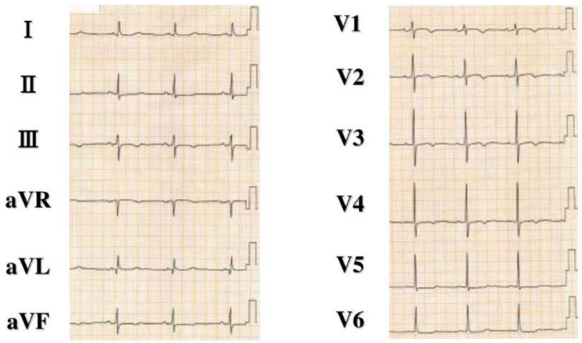

Figure 2. ECG of the patient demonstrating a heart rate of 69 bpm, a normal sinus rhythm, with no significant ST shift.

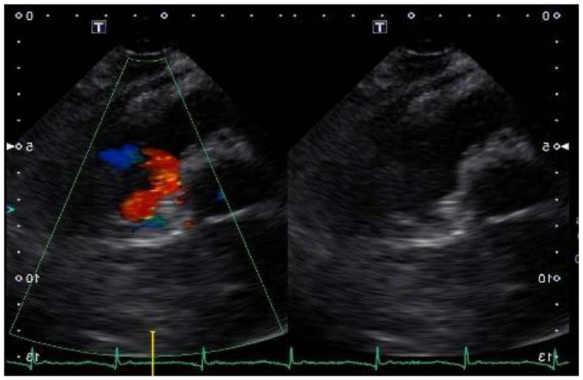

Figure 3. Transthoracic echocardiography with doppler study demonstrating, ectopic opening of the coronary artery and blood-stealing phenomenon at the pulmonary artery.

the pulmonary artery, along with an aneurysm of the LCA (Fig. 5). Coronary angiography showed an aneurysm or dilatation of the left coronary artery (Fig. 6). She had a stable condition during her three-day hospital stay and on discharge and was referred to an advanced setting abroad for the surgical management of BWGS. It was impossible to follow up once she left the country.

DISCUSSION

Bland-White-Garland syndrome, or anomalous origin of the left coronary artery from the pulmonary artery, was first reported in 1933 by Bland, White, and Garland [6]. BWGS is a highly uncommon congenital defect that affects 1 in 300 000 newborns. It typically manifests in infancy, 10% of children with this congenital

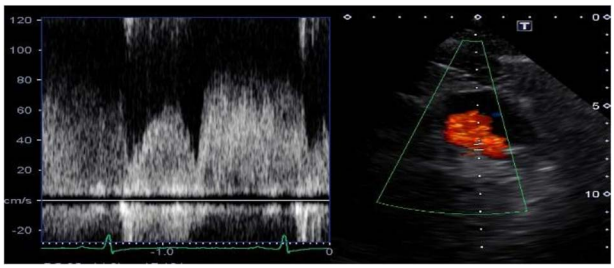

Figure 4. Pulsed-wave doppler echocardiography confirmed the left coronary wave pattern which peaked during diastole. These echocardiographic findings suggested BWGS.

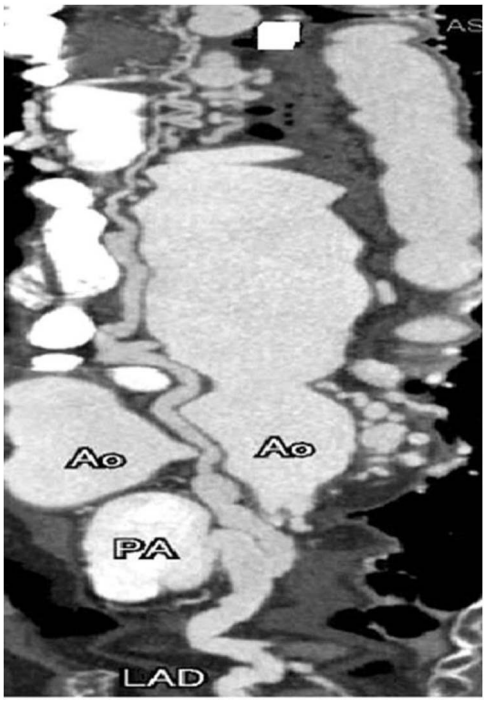

Figure 5. Chest CT scan of the patient showing anomalous origin of the left coronary artery from the pulmonary artery along with an aneurysm of the LCA.

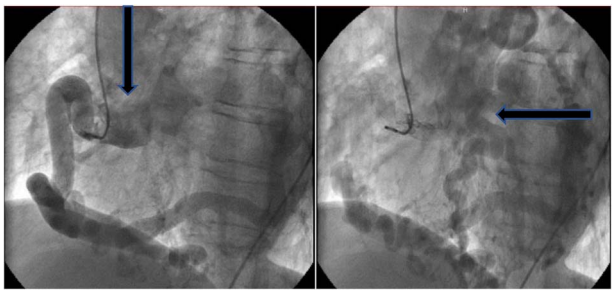

Figure 6. Coronary angiography demonstrating an aneurysm of the left coronary artery with huge collaterals from RCA to LCA.

defect survive to adulthood [5]. The anomaly is usually isolated but has occasionally been associated with other congenital heart defects such as patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot, or coarctation of the aorta [7]. During fetal life, antegrade flow from the pulmonary artery to the coronary artery occurs because of high pulmonary artery pressure, allowing for satisfactory myocardial perfusion from the pulmonary artery through the anomalous coronary artery [7–10]. However, after birth, the pulmonary artery contains desaturated blood at

a pressure that rapidly falls below systemic pressure [4, 7, 9]. Therefore, the left ventricle, with its huge demand for oxygen, is perfused with desaturated blood at low pressure [11]. This predisposes to myocardial ischemia, especially during exertion such as feeding or crying [4, 7, 11]. Collateral vessels develop between the right and left coronary arteries and may provide adequate perfusion to the left myocardium [2, 4, 11]. At this point, blood flow is also reverted in the LCA because the PA has a lower pressure than the arterial collaterals. Coronary steal syndrome is a term used to describe this phenomenon [11]. If collaterals are sufficient, symptoms may be absent or only mild. Adults might have no symptoms, but more frequently, they can have a variety of symptoms, including syncope, chest pain, and sudden death [2, 4, 5]. The final stage in late adulthood, involves an overloaded collateral artery that repeatedly experiences ischemia due to the steal phenomenon from the RCA to the LCA and pulmonary artery [9]. BWGS can be diagnosed through the use of computed tomography (CT) of the chest, coronary angiography, and echocardiography [3, 5]. Surgery is suggested for BWGS as soon as possible [2, 4]. A dilatation of a blood vessel with a luminal diameter greater than 1.5 times that of the adjacent normal segments is known as a coronary artery aneurysm (CAA). Children rarely experience this abnormality, and the main cause of CAA is Kawasaki disease [12].

In the mentioned case of an asymptomatic adolescent girl, the BWGS and an aneurysm of the LCA were diagnosed by echocardiography, CT scan, and angiography. Transthoracic echocardiography with doppler study demonstrated, ectopic opening of the coronary artery and blood-stealing phenomenon at the pulmonary artery (Fig. 3). The pulsed-wave doppler echocardiography confirmed the left coronary wave pattern which peaked during diastole (Fig. 4). These echocardiographic findings suggested BWGS. Chest CT scan of the patient showed anomalous origin of the left coronary artery from the pulmonary artery along with an aneurysm of the LCA (Fig. 5). Coronary angiography demonstrated an aneurysm of the left coronary artery with huge collaterals from RCA to LCA (Fig. 6). Polak et al. reported a case of anomalous origin of the left coronary artery from the pulmonary artery in a 16-year-old girl with a history of heart failure in infancy [13]. An anomalous origin of the left coronary artery from the pulmonary artery was evaluated by Yazdi et al. The patient was a 25-year-old who had experienced cardiac arrest; a chest CT scan revealed a right coronary artery aneurysm [1]. Sadoma et al. reported an infant with BWGS and heart failure [11]. Our case differs from the reports of Polak et al., Yazdi et al., and Sadoma et al. by the presence of a left coronary artery aneurysm, an asymptomatic course, and normal heart function. Kawasaki disease is the most important cause for coronary artery aneurysms in children and adolescents, and should be considered in the differential diagnosis of CAA [12]. However, our case lacks all criteria for this differential diagnosis. Therefore, BWG is the most appropriate diagnosis, and CAA may be associated with high blood flow between the RCA and LCA, as well as steal phenomenon. The existence of a left coronary artery aneurysm, an asymptomatic course, and normal heart function in an adolescent distinguish our case of BWGS from the previous published reports [1, 4, 6, 8, 9, 11, 13]. In this case, the patient was advised to undergo surgical management of BWGS in an advanced setting abroad. The surgical management is crucial [2, 4], but it can be difficult in settings with limited resources.

CONCLUSION

BWGS is an extremely rare anomaly in adolescents. This anomaly may lead to the establishment of collaterals between the RCA and LCA and aneurysm of the coronary artery due to high blood flow and the steal phenomenon. The BWGS may be asymptomatic until adolescence as a result of numerous collaterals between the RCA and LCA.

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CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

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ETHICAL APPROVAL

This study was approved by the Department of Neonatology, Kabul University of Medical Sciences.

CONSENT

Written informed consent was obtained from the patient's next of kin for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

GUARANTOR

Mansoor Aslamzai, MD, Professor of Neonatology.

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