Case report: ALCAPA syndrome in a 14 year old Iraqi girl presented with acute myocardial infarction

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ABSTRACT

Anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome is a rare congenital coronary artery anomaly. There are two types of ALCAPA syndrome: the infant and the adult types, each type presents with different clinical findings and outcomes. In infantile type, infants usually suffer myocardial infarction and congestive heart failure, and about 90% of them die within the first year of life if untreated surgically. In rare instances, ALCAPA syndrome presents in adolescents and adults; it is considered as an important cause of sudden cardiac death. The ideal treatment of ALCAPA syndrome is surgical repair performed by restoration of a dual-coronary-artery system.[13] We reported a case of a 14 year old girl presented with an acute myocardial infarction and her coronary angiography revealed ALCAPA syndrome. Four months after ALCAPA diagnosis, she was successfully under went surgical reconstruction without any major complications.

متلازمة الكابا (نشوء شاذ للشريان التاجي من الشريان الرئوي الرئوي) في فتاة ذات اربعة عشر عاما عانت احتشاء العضلة القلبية الحاد

النشوء الشاذ للشريان التاجي الايسر من الشريان الرئوي (الكابا) هس حالة تشوه ولادي نادر للشرايين التاجية. يوجد نوعان من هذا المرض، النوع الاول هو النوع الطفولي الذي يظهر في سن مبكرة والنوع الثاني هو نوع الكبار الذي يظهر في سن المراهقة او البلوغ، وكل منهما له علاماته السريرية ومضاعفاته. في النوع الطفولي نجد بأنه الاطفال الرضع يعانون من احتشاء العضلة القلبية وعجز القلب، وتقريبا ٩٠% منهم يموتون خلال السنة الاولى من الحياة. نادرا" ماتشاهد متلازمة الكابا في سن المراهقة والبلوغ، وقد تكون سببا" مهما للموت الفجائي في هذه الفترة من العمر. تاريخيا"، متلازمة الكابا تم تشخيصها عن طريق القسطرة الاعتيادية، مع ذلك، فأن التطور في مجال التشخيص ادخل الطرق الحديثة كالمفراس الحلزوني والرنين في تشخيص هذه الحالة مما يعطي معلومات دقيقة جدا" عن هذا التشوه الذي يمكن الجراح من اختيار نوع العملية الجراحية المطلوبة لعلاج الحالة. نحن وثقنا حالة لفتاة في الرابعة عشر من عمرها عانت احتشاء حاد للعضلة القلبية وثبتت متلازمة الكابا من خلال اجراء القسطرة القلبية التي اجريت لها في مركزنا والتي بينت نشوء شاذ للشريان التاجي الايسر من الشريان الرئوي وامتلاء هذا الشريان عن طريق شرايين صغيرة رابطة مع الشريان التاجي الايمن، كفاءة البطين الايسر كانت جيدة مع استرجاع حفيف في الصمام الاكليلي، مع عدم وجود سائل غير طبيعي في شغاف القلب. بعد مضي اربعة اشهر من التشخيص اجريت لها عملية جراحية بدون مضاعفات مهمة. بعد مرور سنتين وجدنا بأن كفاءة البطين الايسر طبيعية وبدون اي اعراض قلبية.

INTRODUCTION

The incidence of ALCAPA syndrome is very rare (1 of 300,000 live births)[1,2] and represents about 0.4 percent of congenital cardiac anomalies. It was reported for the first time in 1885 by Brooks[1] and described clinically in conjunction with autopsy findings for the first time by Bland, et al. in 1933[3], therefore, it is also known as Bland-White-Garland-syndrome. Usually, ALCAPA syndrome is an isolated cardiac anomaly which does not manefist prenatally due to favorable fetal physiology that is characterized by equivalent pressures in the aorta and pulmonary artery resulted from a nonrestrictive patent ductus arteriosus (PDA), and the relatively equivalent O_2 saturations resulted from parallel circulations, consequently the perfusion of myocardium will be normal, so that there is no stimulus for the formation of collateral vessels between the right and left coronary artery systems. Shortly after birth, the parallel circulation will be abolished and becomes in one series, the pulmonary artery pressure and pulmonary resistance decrease, and also the O_2 saturation of pulmonary blood flow. Consequently, there will be a decrease in antegrade flow and O_2 saturation of the

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anomalous left coronary artery, causing ischemia in myocardium. This results in initiation of collateral circulation between the right and left coronary systems, and reversion of flow in the left coronary artery flow and enters the pulmonary trunk because of the low pulmonary arterial pressure, resulting in what is called coronary steal phenomena that causes a fixed ischemia or myocardial infarction. 85% of all ALCAPA cases present within the first 2 months of life. The clinical presentation and course of ALCAPA include severe left-sided heart failure with significant mitral valve regurgitation due to ischemia of papillary muscle, permanent fibrosis, or left ventricular dilatation. The presentation at late stage is very rare and usually with significant cardiac dysfunction.[14-16] A case report of a 14 year old girl with ALCAPA syndrome will be discussed.

CASE PRESENTATION

A 14-year-old girl presented with dyspnea and chest pain for two days that was misdiagnosed and treated as a case of musculoskeletal chest pain. She experienced worsening left-sided chest pain and subsequently referred to our cardiac center. She had no coronary risk factors and no family history of premature coronary artery disease or congenital heart disease. Physical examination was unremarkable apart from sinus tachycardia. ECG showed sinus tachycardia with ST segment depression at anterolateral and inferior leads-Non ST elevation myocardial infarction (Figure-1). Chest X-ray showed no cardiomegaly. Cardiac troponin was elevated Echocardiography showed preserved left ventricular function (LV ejection fraction=65%) with anterior hypokinesia and mild mitral regurgitation, no pericardial effusion. A subsequent cardiac catheterization in Basrah Cardiac Center confirmed the diagnosis of ALCAPA with retrograde filling in left coronary artery through collaterals arising from an enlarged right coronary artery, significant left-to-right shunting left main coronary artery into main pulmonary artery trunk (Figure-2). The patient underwent surgical treatment in India by ligation of left coronary artery ostium with left internal mammary artery graft to obtuse marginal artery and right internal mammary artery to left anterior descending artery. She had no major complications or ischemic symptoms two years after surgical operation. A follow up echocardiography showed normal size cardiac chambers and normal left ventricular systolic function and absence of valvular abnormalities.

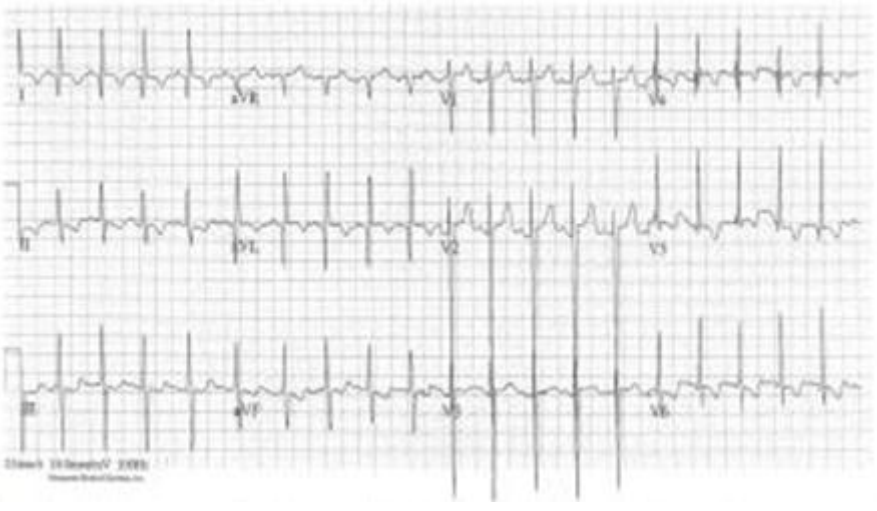

Fig 1. ***ECG at time of presentation: Sinus Tachycardia with ST depression at anterolateral and inferior leads.***

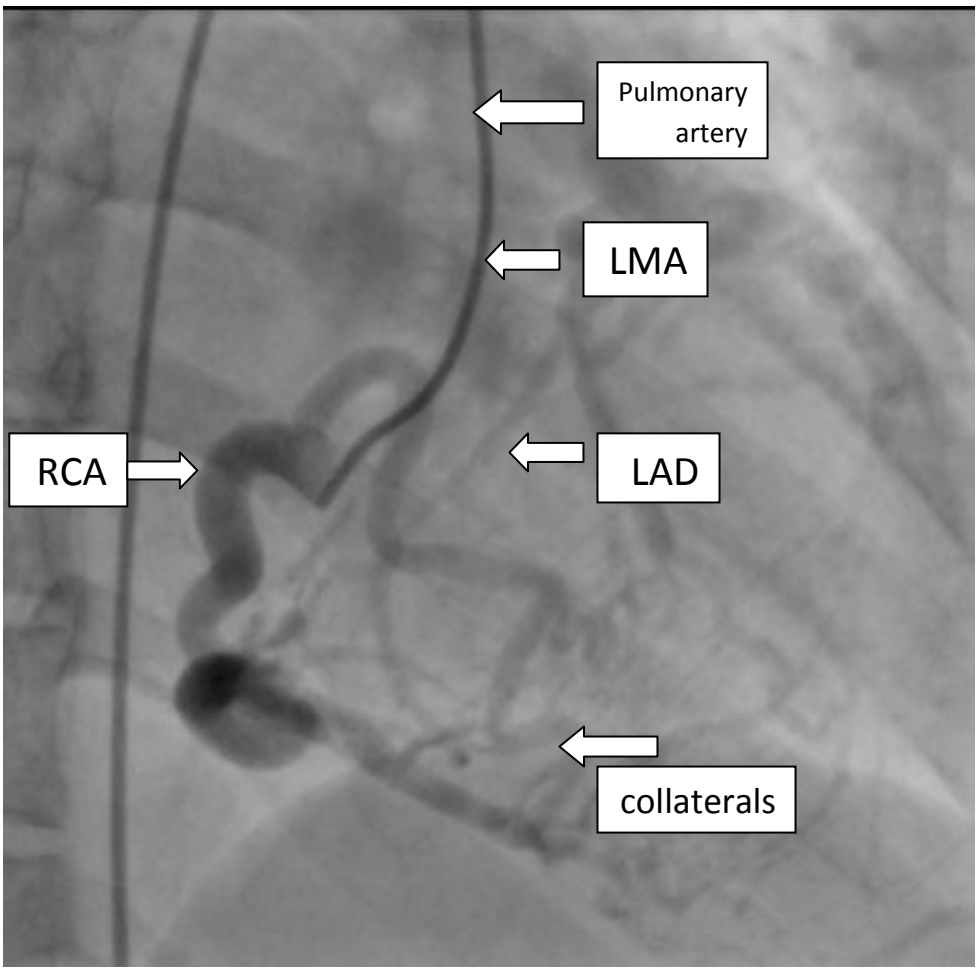

Fig 2. ***Right Coronary angiogram in LAO view: A retrograde flow seen via collaterals arising from dilated right coronary artery(RCA) to a large left descending artery (LAD) and left main artery (LMA) arised from pulmonary artery trunk. A significant left to right shunt filling the pulmonary artery via LAD and LMA.***

DISCUSSION

ALCAPA syndrome is very rarely presented late at adolescence and adulthood because of few patients can survive childhood without surgical repair.[4] Different presentations can be seen in adult type ALCAPA syndrome, of these; severe dilated cardiomyopathy (irreversible cardiac dysfunction), malignant arrhythmia (secondary to myocardial scar tissue), acute myocardial infarction, sudden cardiac death, severe mitral regurgitation.[5-10] In our case, she has an adulthood type, and presented with a preserved LV systolic function without evident functional or structural cardiac dysfunctions that prevent from a life-threatening cardiac event later in life. Abundant collaterals blood supply from RCA play an important role in some patients with ALCAPA to pass childhood with minor symptoms like shortness of breath, exercise intolerance and chest pain, and in most instances these symptoms are misdiagnosed (as in our case who misdiagnosed initially as a case of musculoskeletal chest pain). High index of suspicion and thorough physical examination play an important role for early diagnosis in children with exercise intolerance. ECG is usually unremarkable. Echocardiographic exam is an essential diagnostic tool for early diagnosis of ALCAPA syndrome by observation of a unique echocardiographic finding which is characteristic for ALCAPA syndrome as multiple turbulent flow seen in the interventricular septum because of abundant

collaterals and also higher coronary flow velocity in systole than diastole by using Pulse Width Doppler examination of right coronary artery and coronary collaterals.[17-19] So, echocardiography may be used as the first examination for diagnosis of ALCAPA syndrome.[20] In most cases of infantile type who were surgically corrected, have normalization of left ventricular systolic function and mitral valve insufficiency.[11,12] The long-survival at 20 years was estimated as 94.8%.[12] The prognosis of ALCAPA syndrome is usually determined by the degree of irreversible left ventricular systolic dysfunction and presence of myocardial scar tissue. Our patient had completely normal left ventricular systolic function two years after surgical repair, which is indicated the absence of irreversible LV systolic dysfunction at the time of diagnosis.

Conclusion: The diagnosis of ALCAPA syndrome in adolescence and adulthood needs high index of suspicion in patients with left ventricular dysfunction or evidence of ischemic heart disease.

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