International Journal of Cardiology xxx (2017) xxx–xxx

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Anomalous origin of coronary arteries from the "wrong" sinus in athletes: Diagnosis and management strategies

Vincenzo Palmieri [a,1,2], Salvatore Gervasi [a,1,2], Massimiliano Bianco [a,1], Roberta Cogliani [a,1], Barbara Poscolieri [a,1], Francesco Cuccaro [a,1], Riccardo Marano [b,1], Mario Mazzari [c,1], Cristina Basso [d,1,2], Paolo Zeppilli [a,*,1,2]

[a] Sports Medicine Unit, Catholic University of the Sacred Heart, Rome, Italy
[b] Department of Radiological Sciences, Institute of Radiology, Catholic University of the Sacred Heart, Rome, Italy
[c] Department of Cardiovascular Medicine, Catholic University of the Sacred Heart, Rome, Italy
[d] Department of Cardiac, Thoracic and Vascular Sciences, University of Padua Medical School, Padova, Italy

ARTICLE INFO

Article history:
Received 10 July 2017
Received in revised form 28 September 2017
Accepted 31 October 2017
Available online xxxx

Keywords:
Athletes
Congenital coronary anomalies
Sports eligibility
Surgery

ABSTRACT

Aims: Although anomalous origin of left (AOLCA) and right coronary artery (AORCA) from the wrong sinus may cause sudden death (SD) in athletes, early diagnosis and management of these anomalies are still challenging. We analysed clinical/instrumental profiles of athletes identified with AOLCA/AORCA focusing our attention on diagnosis, management and follow-up.

Methods and results: We report 23 athletes (17 males, mean age 27 ± 17 yrs.), 6 with AOLCA and 17 with AORCA. Diagnosis was made by trans-thoracic echocardiography (TTE) in 21/23(91%). Symptoms were present only in 10(41%). Only 3 had an abnormal rest-ECG and 9(39%) an abnormal stress test ECG (3 ST-depression, 4 ventricular arrhythmias, 1 supraventricular arrhythmias, 1 rate-dependent left-bundle-branch-block). Anatomy of the anomalous coronary artery showed no significant correlation with clinical presentation, except for a tendency to higher occurrence of proximal hypoplasia in symptomatic athletes (83% vs 40%, $p = 0.09$). All athletes were disqualified from competitive-sports and advised to avoid strenuous effort. Surgery was recommended to all athletes with AOLCA and 6 with AORCA, but only 6 underwent surgery. No major cardiac events or ischemic symptoms/signs occurred during a mean follow-up of 65 ± 70 months.

Conclusions: Early diagnosis of AOLCA/AORCA in athletes is feasible by TTE. Typical symptoms/signs of myocardial ischemia are present only in one third of cases thus underlying the need of a high index of clinical suspicion to achieve the diagnosis. After exercise restriction, none had major cardiac events or ischemia symptoms/signs recurrence. There was no correlation between anatomical characteristics and clinical presentation with the possible exception of coronary hypoplasia.

1. Introduction

Anomalous origin of a coronary artery is a rare congenital disease, with a prevalence in the general population between 0.17% and 1.3% [1–3]. It is usually a "benign", incidental finding, but in a minority of cases, like the origin of a coronary vessel from the pulmonary artery and the anomalous origin of left (AOLCA) or right (AORCA) coronary artery from the opposite, "wrong", sinus, it may lead to major cardiovascular events and sudden death (SD) [3–5].

Subjects with AOLCA and AORCA may complain of typical chest pain, but most of them have atypical symptoms like syncope/pre-syncope, chest "discomfort" (mild-moderate sense of chest tightness), dyspnoea, palpitations, or may be fully asymptomatic, being cardiac arrest the first manifestation of the disease [4–6]. Furthermore, in patients doomed to suffer SD, stress-test ECG in some cases shows ST-depression suggestive of myocardial ischemia, but more often is negative or shows non-specific findings such as ventricular arrhythmias and/or T-wave abnormalities [5]. Therefore, early diagnosis of AOLCA or AORCA, including malignant forms, may be very difficult, a crucial point for cardiologists and sports physicians, since these anomalies are the second cause of SD in young athletes in USA [7] and the third in Italy (Veneto Region) [8].

Trans-thoracic echocardiography (TTE) is considered a reliable non-invasive tool in detecting such anomalies, although its sensitivity/specificity can be influenced by the acoustic window and operator's experience [9–11]. Confirmation of diagnosis can be easily obtained by Magnetic Resonance Imaging (MRI) or Coronary Computed Tomography (CCT) [12–14].

Another challenge is risk-stratification and decision about medical or surgical management and competitive-sports eligibility [15–19]. To

* Corresponding author at: Sports Medicine Unit, Catholic University of the Sacred Heart, Largo Francesco Vito 1, 00168 Rome, Italy.
E-mail address: paolo.zeppilli@unicatt.it (P. Zeppilli).
[1] This author takes responsibility for all aspects of the reliability and freedom from bias of the data presented and their discussed interpretation.
[2] These authors equally contributed.

https://doi.org/10.1016/j.ijcard.2017.10.117
0167-5273/

date, most authors agree in exercise restriction and surgical indication in patients with AOLCA and in those with AORCA when symptomatic and/or with evidence of ischemia at provocative tests. On the other side, there is still uncertainty about management of asymptomatic subjects with AORCA and negative provocative tests. Some authors underlined that some anatomical features of the anomalous artery, such as ostium morphology, inter-arterial course, etc., should be taken into account in evaluating the risk and the need of surgical treatment [5,20,21]. Unfortunately, none of these "anatomical criteria" is precisely codified to date. New parameters, as fractional flow reserve (FFR) with coronary angiography (COR) [22] or, as recently proposed, by CCT [23], can open new perspectives.

The final problem is returning to competitive-sports after surgery. This possibility seems to be strongly influenced by type of intervention, its efficacy in eliminating all mechanisms potentially responsible for ischemia [24–27] and it requires a correct methodology of evaluation.

In this report, we present 23 athletes, many of them very young, with AOLCA/AORCA, discussing our investigation protocol and decisions about competitive-sports eligibility, and trying to delineate possible management strategies of this potentially lethal disease.

2. Material and methods

We conducted a retrospective analysis among all athletes (subjects practising sports at competitive or professional level) referred to our Sports Cardiology Unit from May 1986 to May 2017, where we visit about 2000 athletes and perform about 350 TTE per year (roughly 10,000 TTE in thirty years). We identified 39 cases of coronary anomalies: 6 AOLCA, 17 AORCA, 1 single coronary artery, 15 anomalous origin of the circumflex artery from the right sinus/coronary artery. To the purpose of the study, only subjects with AOLCA or AORCA ($n = 23$) were selected. Four of them had been diagnosed elsewhere, and sent to us for a consultation. Nineteen had been firstly diagnosed in our Centre, where a systematic search of ostia and first tracts of coronary arteries is carried out during TTE, either performed for routine examination (professional athletes), or to clarify symptoms and/or ECG anomalies observed during basal pre-participation screening for competitive-sports (which is mandatory in Italy and includes rest- and stress test-ECG). When suspecting a coronary anomaly, TTE was repeated by a second experienced operator (P.Z. or V.P.) and, if the suspicion was confirmed, further investigation, COR in the past or, more recently, MRI/CCT, was performed. TTE was performed using three machines: a Siemens Sonoline CD, an Acuson-CV70 (Siemens Medical Solutions USA, Inc., WA, USA) and, more recently, with a Toshiba Artida U.S. (Toshiba Medical Systems, Inc. Tustin, CA).

We focused our attention on symptoms occurred during or immediately after exertion, rest and stress-test ECGs, anatomical details obtained by TTE and CCT/MRI, indications for surgical counselling, decisions about competitive-sports eligibility (pre- and eventually post-surgery) and follow-up data.

The whole study was conducted according the GCP and the Helsinki's declaration. Written consent was obtained from all subjects, and the study design was approved from the Ethics Committee of the Catholic University of Rome. For multiple categories, we performed a statistical analysis using Chi-Square Test or Fisher's Exact Test.

3. Results

3.1. Diagnosis, prevalence and follow-up

Our series consist of 23 cases (Table 1), 6 AOLCA (Table 1A, 3 males, mean age at diagnosis 19 ± 14 yrs.) and 17 AORCA (Table 1B, 14 males, mean age at diagnosis 29 ± 17 yrs.). Ten athletes were symptomatic (43%), 13 (57%) asymptomatic (see below for details). In 21 out of 23 (91%), suspicion or diagnosis had been made by TTE. The remaining two subjects were: a 50-years old runner with AORCA discovered by COR, performed for an inferior-STEMI episode after exertion in absence of atherosclerotic disease, sent to us for counselling; a 13-years old soccer player, asymptomatic, sent to us after an incidental diagnosis of AORCA at a MRI performed as control of previous Kawasaki's Disease.

In 18/21 (6/6 AOLCA and 12/15 AORCA) TTE was performed as second level examination due to either symptoms or ECG signs. The remaining 3 AORCA underwent TTE because one was a professional soccer player, one was included in a specific screening program inclusive of TTE and one had hypertension. A single case of AORCA was incidentally identified among 227 professional athletes who consecutively underwent routine TTE examination (0.44%).

Twenty out of 21 cases (95%) identified by TTE, were further investigated by COR (3 cases), CCT or MRI. One subject, with an unequivocal TTE evidence of AORCA, refused further investigations.

Follow-up was available in 20 cases, 5 AOLCA and 15 AORCA (mean 65 ± 70 months).

3.2. Clinical, anatomical and instrumental findings in relation to type of anomaly

AOLCA (Table 1A). In all six cases, the left coronary trunk arose from the right sinus of Valsalva and had full inter-arterial (INT) course. An intramural proximal segment was documented in two cases (33%). Symptoms during or immediately after exertion were present in 3 (50%): one subject had chest pain and syncope, one chest "discomfort" and presyncope, and one dyspnea and chest "discomfort" (Fig. 1). Rest-ECG was normal in 5 (83%) and showed lateral T-wave inversion in one (with evidence of mild apical hypertrophic cardiomyopathy). Five subjects underwent stress test ECG, which was normal in 3 (60%), showed a rate-dependent left-bundle-branch-block (LBBB) in one, and ST-segment depression >1 mm in one.

AORCA (Table 1B). Ten out of the 17 athletes (59%) were *asymptomatic*. Among them, 8 (80%) had normal rest-ECG, 2 (20%) showed a left-anterior-fascicular-block. Stress test ECG was normal in 4 (40%), showed isolated supraventricular premature beats (SVPB) in 1 (10%),

Table 1A
Clinical data of athletes with anomalous origin of the left coronary artery from the right sinus of Valsalva (AOLCA).

N	Age	Sex	Sport	TTE+	Further investigation	Course	Anatomical details	Symptoms*	Rest-ECG	Stress-test ECG	Management	Type of surgery	Follow up (months)
1	47	M	Running	Yes	COR/MRI	INT	Extramural	NO	Normal	Rate-dep.LBBB	BB;ER Refused Surgery		120
2	14	F	Dancing	Yes	CCT	INT	Intramural segment	NO	Normal	ST-depression	BB;ER Refused surgery		246
3	16	M	Soccer	Yes	COR	INT	Not available	No	Lateral-TWI	Normal	BB;ER Refused surgery		NA
4	11	M	Swimming	Yes	COR/CCT	INT	Intramural segment	Chest pain, syncope	Normal	Normal	Surgery	Unroofing	143
5	11	F	Dancing	Yes	COR/CCT	INT	Extramural; Intramyocardic segment	Chest discomfort, presyncope	Normal	Normal	Surgery	Pulmonary traslocation	91
6	15	F	Figure-skating	Yes	CCT	INT	Extramural	Dyspnea, chest discomfort	Normal	Not performed	Surgery	Unroofing	6

Please cite this article as: V. Palmieri, et al., Anomalous origin of coronary arteries from the "wrong" sinus in athletes: Diagnosis and management strategies, Int J Cardiol (2017), https://doi.org/10.1016/j.ijcard.2017.10.117