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Original article

Anomalous origin of the right coronary artery evaluated with multidetector computed tomography and its clinical relevance

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ABSTRACT

Background: Anomalous origin of the right coronary artery (AORCA) is a rare congenital anomaly that may cause myocardial ischemia and sudden death.
Methods: We reviewed the clinicopathological records of three cases of AORCA, and compared these with two cases of sudden cardiac death with AORCA revealed by autopsy.
Results: We report three juvenile cases with an AORCA originating above the commissural junction between the left and right aortic sinuses, with interarterial and intramural compression. They presented with exertional symptoms and were diagnosed with an AORCA by multidetector computed tomography (MDCT), which successfully delineated the spatial resolution of the anomalous origin and course of the right coronary artery (RCA), in the operating room. All three underwent successful surgical unroofing of the RCA. Two cases of sudden cardiac death with AORCA revealed by autopsy showed a slit-like orifice, acute-angled take-off, and long intramural course of the RCA, resembling the RCAs of three juvenile cases.
Conclusions: It is crucial to be alert to the presentation of exertional symptoms, as sudden death may be the first manifestation of an anomalous coronary artery, such as those observed in these three cases. MDCT provided an excellent definition and spatial resolution of the unusual origin and intramural course of the RCA, facilitating the correct surgical remedy and resulting in a good outcome for the patients.

Introduction

The incidence of coronary arterial anomalies including anomalous origin of the right coronary artery (AORCA) in normal heart ranges from 0.2% of young patients undergoing echocardiography to 1.2% of patients at angiography [1–4]. Although most cases are clinically silent; anomalous aortic origin of a coronary artery has been associated with an increased risk of myocardial ischemia and infarction, congestive heart failure, and sudden cardiac death (SCD) [1,5]. Theories for the pathophysiology of AORCA associated with SCD have suggested an intramural course, the presence of a slit-like ostium, marked artery angulation, compression from the pulmonary artery, arterial spasm, and arrhythmia secondary to minor ischemic insults [3,6,7]. However, the relationship between the clinical course and the anatomical features of AORCA has not been completely elucidated.

Here we report three cases of AORCA. Multidetector computed tomography (MDCT) successfully demonstrated a small orifice, an acute-angled take-off of the right coronary artery (RCA) from the aorta, and an intramural course of the RCA within the aortic wall. MDCT provides greater spatial and temporal resolution than does conventional coronary angiography and thus may be more informative for determining the mechanisms by which myocardial ischemia is invoked in patients with AORCA.

Materials and methods

Subjects

We reviewed the clinicopathological records of three cases (Cases 1–3) of AORCA between January 2011 and December

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2014. The ethical committee of Toyama University approved this study, which proceeded in accordance with the ethical standards established in the 1964 Declaration of Helsinki.

Clinical analysis

The analysis covered demographic data (age, gender), cardiovascular risk factors, and the presence of clinical symptoms, including chest pain, at the time of the MDCT examination. The patients' medical histories were also evaluated retrospectively for the presence of arrhythmia, syncope, and myocardial infarction.

MDCT evaluation

The three patients underwent a CT examination using a 64-slice scanner and using a dual-source device with electrocardiogram (ECG)-controlled tube current modulation. The retrospective CT evaluation involved qualitative and quantitative analysis of high-risk anatomy features (take-off angle, orifice measurement, presence and length of intramural course, and type and length of the proximal course of the anomalous vessel). The take-off angle was measured in the plane parallel to the aortic annulus and was specified as the angle between the line parallel to the coronary sinus wall and the line parallel to the proximal course of the coronary artery. An acute take-off angle was defined as an angle $<45^{\circ}$ [8]. A slit-like orifice was determined if the value of the maximal orifice diameter exceeded twice the value of the orthogonal measurement. Anomalous vessels were screened for the presence of the intramural course (within the wall of aorta) and, if shown, its length was measured.

Clinical autopsy evaluation

Sudden death was defined as unexpected death as a result of natural causes occurring within 6 h of initial symptoms [8]. SCD was determined by a complete autopsy examination to exclude other possible causes of death, a toxicological examination, and an investigation of the scene of death.

Clinical autopsy evaluation was carried out for two cases of SCD [9]. One of these (Case 4) was a 75-year-old man with a history of mild hypertension, who had been administered a calcium blocker for 10 years. He collapsed while walking back to his home and was immediately transferred to a hospital but resuscitation was unsuccessful. There was no traumatic injury to his body and toxicological screening of sampled blood was negative. The cause of death was considered to be SCD. The other case (Case 5) was a 32-year-old man without any clinical history, who was found dead in a sauna. Autopsy did not show any trauma to his body. Case 5 was found dead in a sauna after abnormal use of methamphetamine. The blood content of methamphetamine was 1.59 μg/ml, which was below the lethal level of widely used criteria (10 μg/ml). His cause of death was also considered to be SCD under the unusual administration of methamphetamine.

The hearts of these two cases were examined in a manner similar to that described in our previous report [9,10]. Subsequently, coronary arteries were dissected to define the angulation of the first 1.5 cm of the coronary artery relative to the aortic wall. The right and left ventricles were cut at 1-cm intervals parallel to the levels of the papillary muscle from the apex. The hearts were then opened according to the direction of blood flow and any abnormalities of the myocardium, endocardium, or valves were noted. The location and shape of the coronary ostium were also recorded and the epicardial coronary arteries were cut transversely at 5-mm intervals and decalcified as required. The anatomical definitions of normal and anomalous coronary arteries were based on the criteria described by Angelini [11]. The definitions of acute-angle take-off and an ostial valve-like ridge were based on the criteria proposed by Taylor et al. [12]. Significant atherosclerosis was defined as narrowing to 50% of the normal cross-sectional area [1]. Sections at the level of the papillary muscle and the apex of heart muscle were examined histologically in detail. An intramural coronary artery is defined histologically as the coronary artery being contained completely within the aortic wall, sharing the same media with the aorta without interposed adventitia [1].

Results

Clinical profile of the three cases

The clinical summary and pathological findings are summarized in Table 1. Case 1 was a 15-year-old boy who presented with syncope on exercise. Cases 2 and 3 were of a 15-year-old boy and a 13-year-old boy, respectively, who were referred to our hospital because of episodes of chest pain during exercise. In all three cases, the symptoms had been noted a year earlier.

Clinical data

The laboratory data for these three cases, including troponin T and creatine kinase-MB levels, were all normal (Table 1). Results from a 12-lead ECG and a 24-hour Holter monitor were unremarkable. Myocardial perfusion single-photon emission CTs were normal. An echocardiogram revealed normal ventricular function, although the coronary arterial origins were not clearly visualized.

The three patients had surgically confirmed anomalous coronary arteries with an intramural segment. All of these patients underwent the surgical procedure known as unroofing [13]. This procedure consists of incising the internal aortic wall between the aortic lumen and the coronary artery lumen along the entire length of the intramural segment without the use of reimplantation or bypass grafting [14,15], resulting in an enlargement of the neo-ostium and of the slit-like orifice, the acute axial angle of origin, and the intramural nature of the anomalous artery.

During three years of post-surgery follow-up, these patients have remained well without any cardiovascular symptomatology.

MDCT data

MDCT demonstrated in each patient that the RCA origin arose anomalously and had an aberrant intramural course (Figs. 1 and 2).

Table 1
Clinical findings.

Case	Age/gender	Coronary artery	Symptoms	Laboratory data	Exercise test	Stress echo	Myocardial perfusion scan	Coronary angiography
1	15/M	AORCA	Chest pain syncope	Normal	Normal	n/a	Normal	Normal
2	15/M	AORCA	Chest pain	Normal	Normal	n/a	Normal	n/a
3	13/M	AORCA	Chest pain	Normal	Normal	Normal	n/a	n/a
4	75/M	AORCA	SCD	n/a	n/a	n/a	n/a	n/a
5	32/M	AORCA	SCD	n/a	n/a	n/a	n/a	n/a

AORCA, anomalous origin of the right coronary artery; SCD, sudden cardiac death.