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CASE REPORT

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The co-existence of an aberrant origin of the right subclavian artery and a coronary myocardial bridge

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We encountered the co-existence of an aberrant origin of the right subclavian artery and a myocardial bridge on the left anterior descending coronary artery in the cadaver of an 80-year-old Japanese woman during the course of educational dissection at Nagoya City University Medical School. We document the precise gross anatomical findings with some morphometric measurements. Neither an aberrant origin of the right subclavian artery nor the cardial myocardial bridge is a very rare anomaly, but a case of both anomalies being found in the same body is very rare. We believe this is the first report of the simultaneous occurrence of these two anomalies.

Key words: aberrant origin, right subclavian artery, myocardial bridge, coronary artery

INTRODUCTION

An aberrant origin of the right subclavian artery is an anomaly which occurs during the embryological development of the aortic arch [10, 15, 28]. The incidence of this anomaly has been estimated to be 0.5 to 0.8% in the general population [3, 11].

Myocardial bridge indicates the conditions in which a coronary artery is engulfed by myocardial fibers for a limited segment. The incidence of myocardial bridge varies in previous reports from 5.4% [35] to 85.7% [13] depending on the methods of observation used [4, 14, 54]. In this study, we review one case of the co-existence of an aberrant origin of the right subclavian artery and a coronary myocardial bridge in the same cadaver.

CASE REPORT

Two distinct anomalies of the circulatory system, the aberrant origin of the right subclavian artery and a left anterior descending coronary artery having a short bridging segment of myocardial muscle, were observed during routine educational dissection at Nagoya City University Medical School in a female cadaver who died of stomach cancer at the age 80. After classical dissection a detailed dissection was added in order to make the features completely visible. Photographs were taken after morphometric measurements.

The present study was conducted within the parameters of the written permissions we received from the donor and her surviving relatives. The protocol for the present research did not include any specific issues that needed to be approved by the Ethics Committees of our institution. The present work conformed to the provisions of the Declaration of Helsinki in 1995 (as revised in Edinburgh in 2000).

RESULTS

The aortic arch gave origin to the following four branches in order of appearance from right to left: 1) the right common carotid artery, 2) the left com-

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mon carotid artery, 3) the left subclavian artery and 4) the right subclavian artery. These branches arose independently and a small bicarotid trunk was present (Fig. 1). The right subclavian artery branched about 20.1 mm distal to the left subclavian artery and about 28.5 mm dorsal to it. The right subclavian artery arose from the posterior surface of the distal portion of the aortic arch at the level of the fourth thoracic vertebra. It had a small aortic diverticulum at its origin. At its origin the external diameter of the abnormal artery was 17.2 mm, although the diameter decreased distally to 10.1 mm at a position inferior to the right clavicle. The external diameters of the first three branches of the aortic arch were 11.3 mm, 11.0 mm and 10.7 mm in order of appearance. The anomalous artery passed obliquely toward the right side between the oesophagus and the vertebral column and gave off the right vertebral artery, after which it followed the usual course (Fig. 2). The right vertebral artery entered the transverse foramen of the 6th cervical vertebra in the same manner as the left vertebral artery. The other stems of the aortic arch followed normal arrangements. The aortic arch branching pattern of this case belongs to Type G of the Adachi-Williams-Nakagawa classification (Fig. 3, 4). The right vagus nerve descended into the thorax anterior to the anomalous vessel. Its inferior laryngeal branch passed directly in the neck to reach the larynx without looping around the right subclavian artery. The course of the left vagus nerve and that of the left recurrent laryngeal nerve were found to be normal. The course and termination of the thoracic duct were normal, and the sympathetic trunk and the *ligamentum arteriosum* were also normal.

The middle segment of the left anterior descending coronary artery of this heart had a short bridging segment of myocardial muscle, while the other parts of the coronary arteries were located epicardially. The myocardial bridge existed 17.0 mm distal from the origin of the left anterior descending coronary artery. The length of the short segment of muscle by which a part of the otherwise epicardially located coronary artery was covered was 28.4 mm. The thickness of the bridging muscle band was 2.0 mm (Fig. 5).

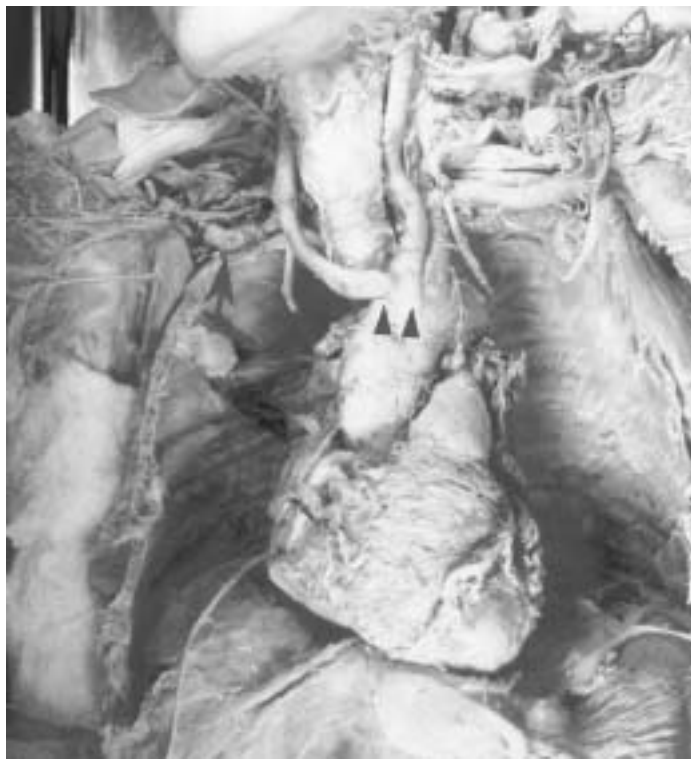
DISCUSSION

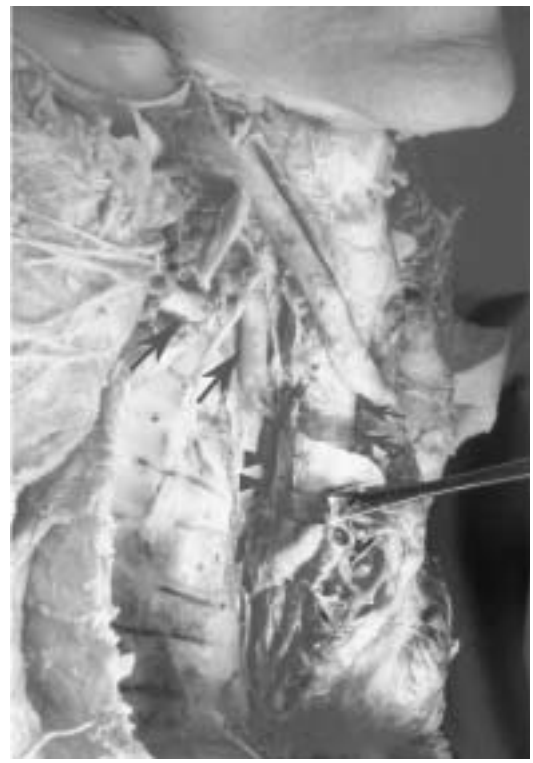
The aberrant right subclavian artery arises from a point distal to the left subclavian artery and most frequently passes behind the oesophagus to the right arm. Rarely, it passes between the oesophagus and the trachea and is seldom anterior to both structures [8, 15, 18]. Therefore, the anomalous artery in the present study took the most common course.

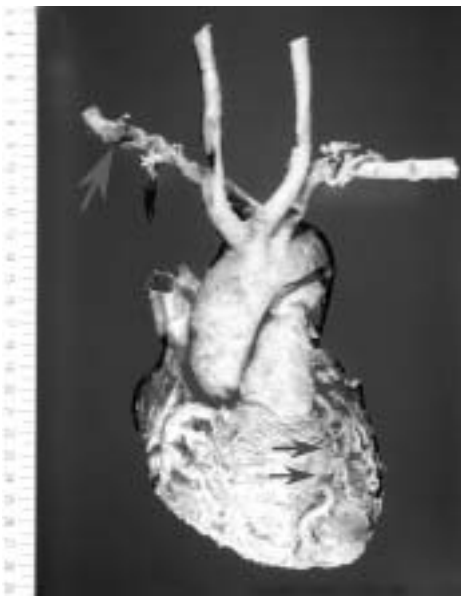
A retro-oesophageal right subclavian artery arising from the proximal portion of the descending thoracic aorta is usually asymptomatic and insignificant clinically. However, from the relation of the anomalous vessel to surrounding structures, symptoms such as inequality of the upper extremity pulses, dysphagia, dyspnoea, enlargement of thoracic ducts and tracheo-oesophageal fistula may sometimes occur [5, 49]. It is also noteworthy that this anomaly can result in subclavian artery aneurysm, and that almost all cases of distal arterial embolisation from an anomalous right subclavian artery have been associated with a right subclavian artery aneurysm [3, 7]. In addition, the risk of iatrogenic injury to the subclavian artery could increase. Therefore, an aberrant subclavian artery can sometimes be a challenging problem for surgical correction.

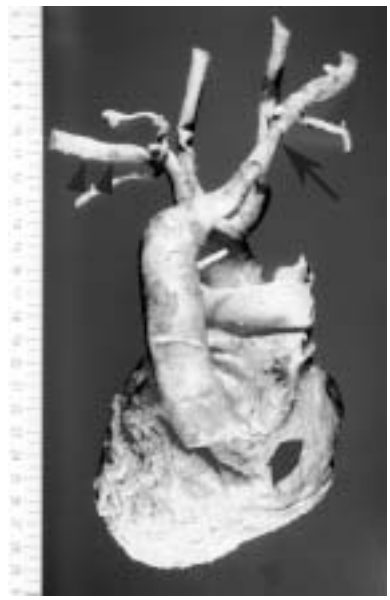
In 1899 Holzapfel classified anomalous cases of the right subclavian artery into 10 types, and the present case belongs to Type 5 [18]. According to the Adachi-Williams-Nakagawa classification, the present case belongs to Type G [1, 31, 51]. The incidence of the Adachi Type G variation in Japanese adults has been reported to be from 0.15 [56] to 1.6% [19] with an average of about 0.5% [43, 55]. In the previous literature for the cases of Adachi Type G referred to in the Japanese, 132 reports in adults, foetuses and neonates have been presented in the anatomical field, comprising 101 reports for adults, including two cadavers whose ages are unknown and 32 for foetuses and neonates [6, 16, 23–26, 29, 32, 33, 36–38, 40–46, 57]. The present case is, therefore, the 134th of Adachi's type G (including Williams-Nakagawa's type H and CG) reported in Japan.

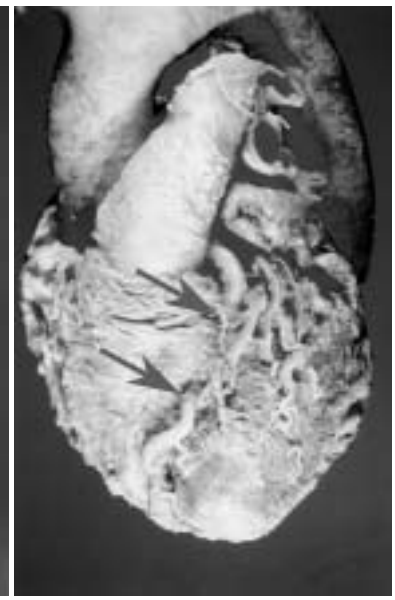
Coronary arteries and their main branches are normally located epicardially on various parts of the heart; however a coronary artery sometimes becomes engulfed by myocardial fibres for a limited segment [4, 35, 54]. Several terms have been used to designate this phenomenon, name, including myocardial bridge, intramural coronary artery, mural coronary artery and coronary artery overbridging. The first article entirely devoted to myocardial bridges in man was published in 1951 by Geiringer who used the term "mural coronary". He found that 25% of the left anterior descending coronary arteries of human hearts have a bridging segment [14]. In 1956 Edwards reported such bridging segments in 15 out of

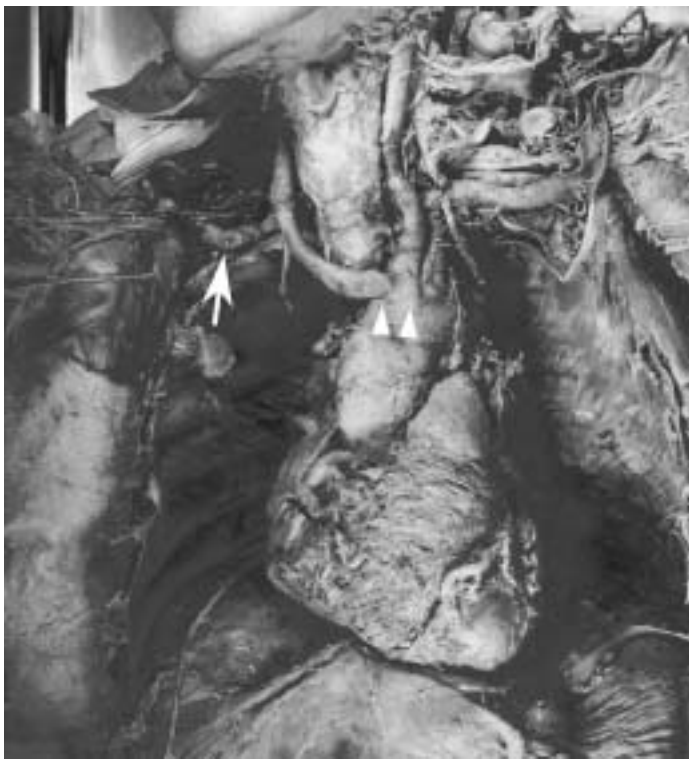


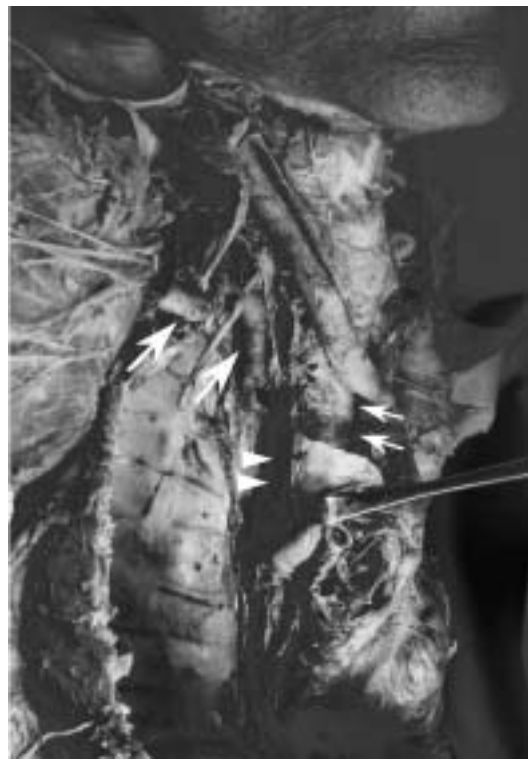


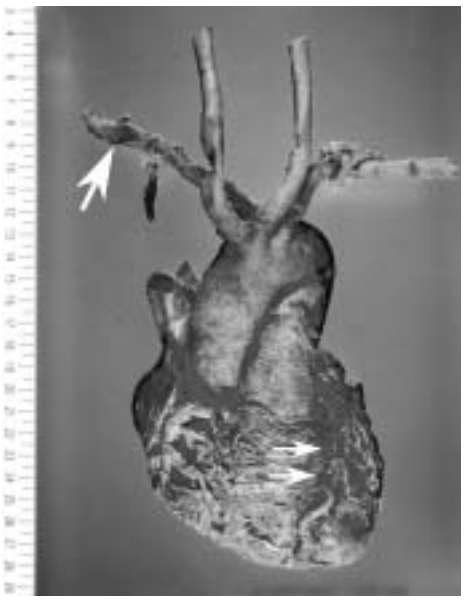


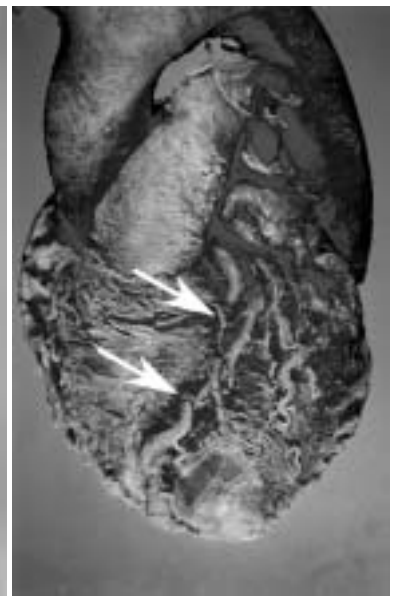


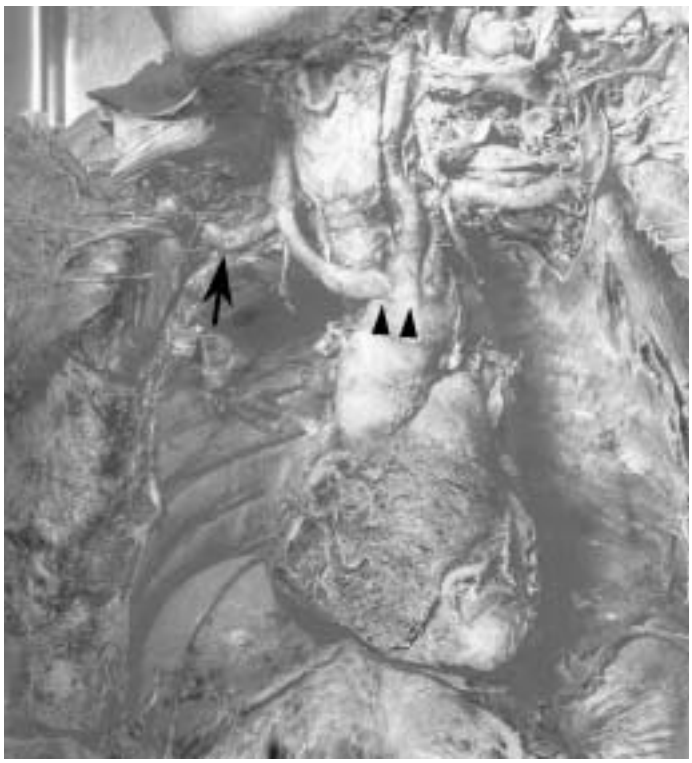


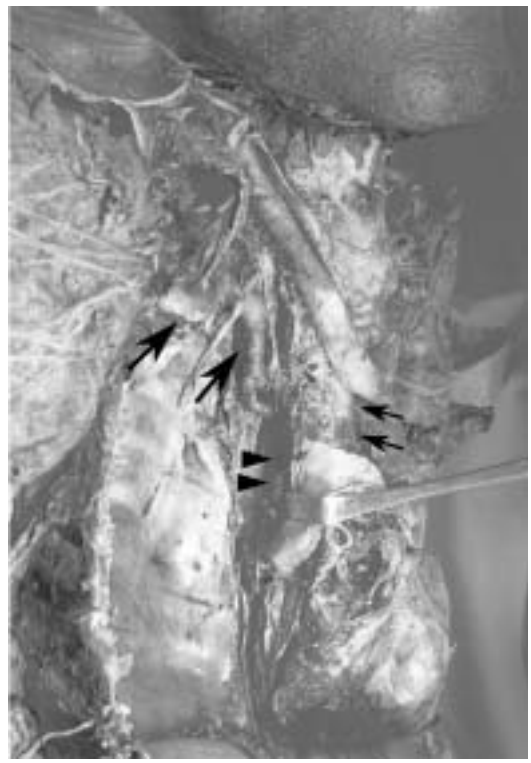












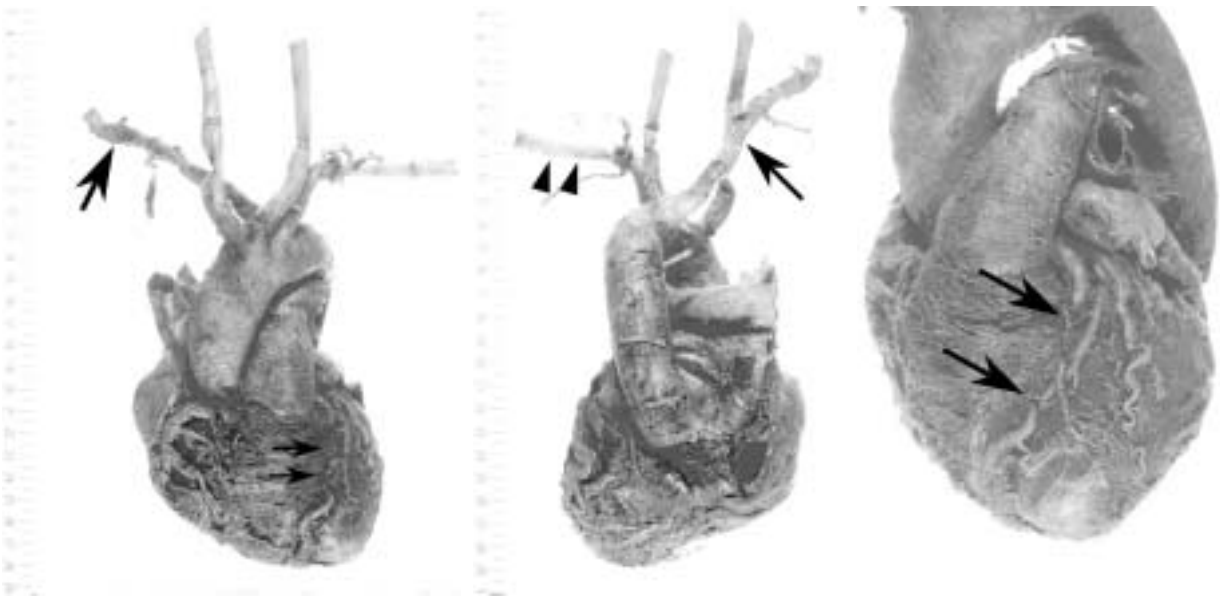


Figure 1. Anterior view. The so-called bicarotid trunk arises from the aortic arch (arrow heads). The origin of the right subclavian artery (arrow) is not visible in this view.

Figure 2. Lateral view from the right side. The right subclavian artery (arrows) passes behind the oesophagus (arrow heads) and the trachea (small arrows) to the right arm.

Figure 3. Anterior view. The right subclavian artery (arrow) passes obliquely toward the right side behind the aortic arch. The aortic arch branching pattern of this case belongs to the Type G of the Adachi-Williams-Nakagawa classification. The myocardial bridge exists on the left anterior descending coronary artery (small arrows).

Figure 4. Posterior view. The right subclavian artery (arrow) arises distal to the left subclavian artery (arrow head) as the last branch of the aortic arch.

Figure 5. Anterolateral (left side) view. The myocardial bridge exists on the left anterior descending coronary artery (arrows).

276 autopsies; among these 15 cases, 13 of the segments were located on the left anterior descending coronary arteries [13]. Polacek was the first to use the term "myocardial bridge" to describe a short bridging segment of myocardial muscle band [35].

Since 1951 more than 200 papers, based on various research methods such as autopsy, scintigraphy and angiography, have been published on the occurrence and significance of myocardial bridges [4, 21, 39, 54]. The most frequent site of a myocardial bridge is the middle segment of the left anterior descending coronary arteries and a typical myocardial bridge is 10 to 30 mm long and 2 to 4 mm thick [4, 54]. Therefore, the situation and size of the myocardial bridge in this study is typical for this anomaly.

Although a myocardial bridge is generally asymptomatic, there have been reports of its association with infarction, vasospasm, myocardial stunning and sudden death [2, 4, 22, 27, 30, 34]. For such a symptomatic myocardial bridge, intracoronary stent implantations or surgical treatments such as supra-arterial myotomy might be recommended [9, 54].

In observations on the vasculature of staged human embryos, the subclavian arteries originate from the 7th intersegmental arteries. The right 7th intersegmental artery forms a confluence with the right 4th arch. Between the late 5th and the 7th week of embryonic age this remodelling event is completed and results in the normal anatomy of the right subclavian artery. The embryological change leading to an aberrant origin of the right subclavian artery is the disturbance of the connection between the 7th intersegmental artery and the right 4th arch. Instead of the normal development, the right 7th intersegmental artery becomes attached to the distal descending aorta [10, 15, 28].

On the other hand, the first evidence of coronary vessel development is the appearance of structures such as blood islands just under the epicardium in the sulci of the developing heart in the 5th week of embryonic age. During the 5th, 6th and 7th weeks of embryonic age the capillary plexuses developing from these foci form connections both with coronary veins sprouting from the coronary sinus and with coronary arteries growing from the aorta [12, 17, 20]. Thus it is probable that the teratogenic event causing both an aberrant origin of the right subclavian artery and a myocardial bridge on the left anterior descending coronary artery occurs at the same embryonic age, between the 5th and 7th weeks.

In the present study, two different variations occurred together in the same person. Of these, an aberrant origin of the right subclavian artery is outside the pericardium and a coronary myocardial bridge is inside the pericardium. Recent studies surrounding basic fibroblast growth factor (bFGF) suggest that these anomalies may not be pure coincidence. Chromosome 22q11.2 deletion (del22q11) causes most cases of DiGeorge syndrome, with an incidence of 1 in 4,000–5,000 live births. This syndrome involves thymic defects, cardiac abnormalities and cleft palate facial anomalies, and it is also known as one of the most common genetic causes of aortic arch abnormalities. Genetic manipulation in mice [50, 52] and mutational analysis in DiGeorge syndrome human patients [53] have shown that Tbx1, which encodes a T-box transcription factor, has a key role in the pathogenesis of this syndrome. In experimental mouse models, Tbx1 haplo-insufficiency causes early growth and remodelling defects of the 4th pharyngeal arch arteries (PAAs) during the embryonic periods. This developmental failure of the right 4th PAA causes an aberrant origin of the right subclavian artery, which is the most common abnormality observed in compound heterozygous mutant mice. Although little is known about specific target genes for T-box proteins, bFGF signalling has been shown to interact with the function of some T-box genes. In addition, it is noteworthy that bFGF with vascular endothelial growth factor (VEGF) can enhance the formation of coronary vessels in the chick embryo. The early vascularisation of the embryonic heart is enhanced after injections of bFGF and VEGF into the vitelline vein before the onset of myocardial vasculogenesis [47], while the administration of neutralising antibodies to bFGF and VEGF clearly altered the arteriolar hierarchy of the heart [48].

This accumulated knowledge suggests that the co-existence of an aberrant origin of the right subclavian artery and a coronary myocardial bridge in the same person might have some significance both in the clinical and basic medical sciences.

To our knowledge, the co-existence of the aberrant origin of the right subclavian artery and a coronary myocardial bridge has never been cited in the previous literature in the anatomical fields. Recently new non-invasive imaging techniques, such as computed tomography and magnetic resonance imaging angiography, have made it possible to demonstrate anatomical abnormalities which have been difficult to detect. The possibility of these two anomalies should always be kept in mind during the clinical diagnoses and treatments of the heart and major vessels.

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