Cardiology in the Young (2014), **24**, 552–554
doi:10.1017/S1047951113000875

Brief Report

Clinically asymptomatic myocardial bridging in a child with familial subaortic stenosis

Selman Gokalp,[1] Funda Oztunc[2]

[1]*Department of Pediatric Cardiology, Trakya University Medical Faculty, Edirne;* [2]*Department of Pediatric Cardiology, Istanbul University Cerrahpasa Medical Faculty, Istanbul, Turkey*

Abstract Myocardial bridging is usually seen in the setting of hypertrophic cardiomyopathy or left ventricular hypertrophy. It is rarely reported in an asymptomatic patient with an otherwise structurally normal heart. Familial subaortic stenosis is also a rare entity, and its mode of inheritance is still unknown. Here, we described the case of a 13-year-old asymptomatic girl with a positive family history of sudden cardiac death and subaortic stenosis who was diagnosed with severe myocardial bridging concomitant with familial subaortic stenosis.

Keywords: Myocardial bridging; familial subaortic stenosis; children

Received: 15 March 2013; Accepted: 24 April 2013; First published online: 28 June 2013

A SEGMENT OF THE EPICARDIAL CORONARY ARTERY embedded in the overlying myocardial tissue is known as myocardial bridging. Myocardial bridging is often an incidental finding in adults, whereas childhood myocardial bridging is almost always associated with hypertrophic cardiomyopathy or left ventricular hypertrophy.[1] Moreover, isolated myocardial bridgings have been found at autopsy in adolescents who had a cardiac arrest during strenuous physical activity.[2] However, after new diagnostic modalities such as quantitative coronary angiography, intravascular ultrasound, and intracoronary Doppler flow velocity measurements became available, the number of newly diagnosed asymptomatic patients with myocardial bridging also increased.[3]

Discrete membranous subaortic stenosis can never be seen at birth, but different members of the same family might be affected. Although the genetic basis of discrete subaortic stenosis is still unknown, familial incidences suggest a multi-factorial model of inheritance in association with environmental factors. Apart from isolated cases, subaortic stenosis might also be associated with other anomalies.[4]

Correspondence to: Dr Selman Gokalp, Trakya Universitesi Tip Fakultesi Cocuk Kardiyoloji Bilim Dali, Balkan Yerleşkesi, Edirne, Turkey. Tel: +90 533 5151474; Fax: +90 284 2352338; E-mail: sgokalp@hotmail.com

Here, we describe the case of a 13-year-old asymptomatic girl who was diagnosed with myocardial bridging during her follow-up for discrete subaortic stenosis.

Case report

A 13-year-old asymptomatic girl was being followed up for the diagnosis of mild discrete subaortic stenosis. Her family history revealed an 11-year-old brother who was being followed up for mild discrete subaortic stenosis and died suddenly during exercise. He was also asymptomatic and had no signs of ischaemia on follow-up visits. Her 16-year-old brother is also being followed up for mild subaortic stenosis and is asymptomatic.

Her physical examination revealed a 2/6 sytolic murmur, best heard at the second and third left intercostal area, with no additional pathological findings. The electrocardiographies on follow-up were normal, without any ischaemic findings, but negative T waves in lead V5–V6 were determined recently. Transthoracic echocardiography revealed mild discrete subaortic stenosis with a left ventricular–aortic systolic pressure gradient of 25 mmHg. The 24-hour electrocardiography evaluation was also normal. An exercise stress test, which was normal

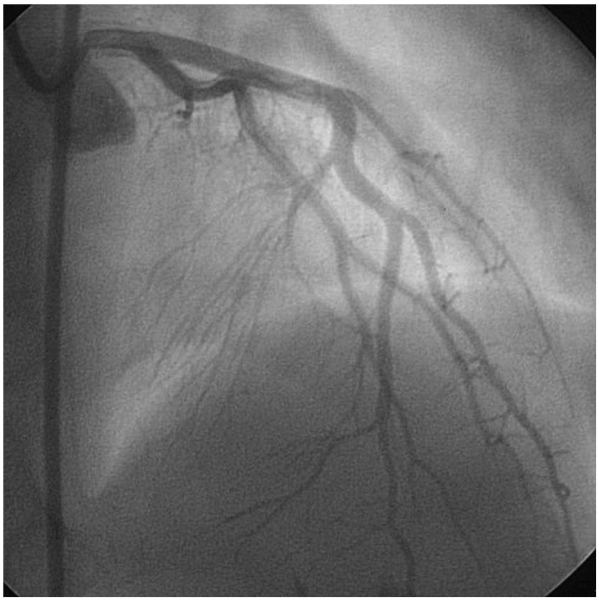

Figure 1.
The left anterior descending coronary artery before systolic compression.

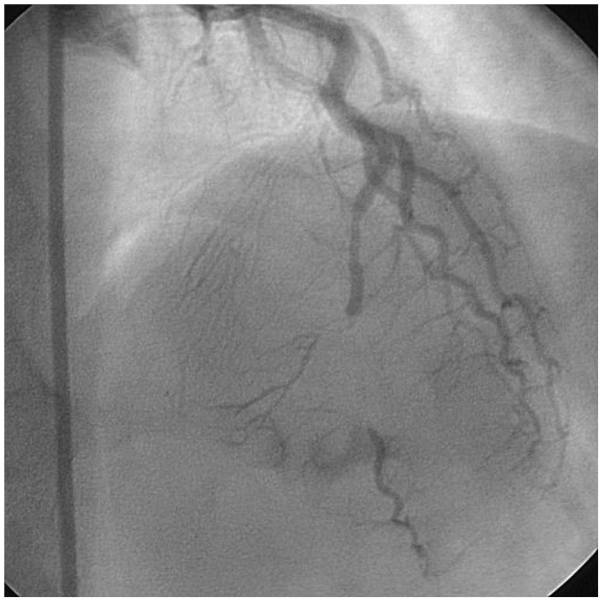

Figure 2.
Systolic compression of the left anterior descending coronary artery.

3 years ago, revealed a 1 mm ST segment depression without any symptom on recent examination.

Owing to a family history of sudden cardiac death and ischaemic findings on the exercise test, a computed tomography angiography of the heart was performed. It revealed that a segment in the middle part of the left anterior descending artery was embedded in the myocardium. On the basis of these findings, the patient underwent cardiac catheter angiography. After the diagonal 2 division of the left anterior descending artery, a short segment of myocardial bridging with a 70–80% narrowing was observed. Just 1 cm distal to this segment, another myocardial bridging of 20 mm length with a positive milking effect of 100% systolic compression was also seen (Figs 1 and 2; Supplementary videos: 1, 2). The myocardial fractional flow reserve was 0.80 with 120 mcg adenosine. The systolic gradient between the left ventricle outlet and the ascending aorta was 15 mmHg. To reveal the ischaemic effect of myocardial bridging, myocardial perfusion scintigraphy was performed. The scan revealed anterior and anteroseptal ischaemia at the apical part of the left ventricle.

β-Blocker therapy was initiated. Thereafter, she was operated upon successfully for myocardial bridging and discrete subaortic stenosis. The postoperative echocardiogram was normal, and the exercise stress test showed no signs of ischaemia.

Discussion

Coronary arteries and their major branches lie on the surface of the heart. When a band of myocardial muscle fibres overlies a segment of the coronary artery, which results in mechanical stenosis secondary to systolic compression, a condition termed as myocardial bridging occurs. The incidence of myocardial bridging in the general population is between <5% and 86% in different series, according to the method used to determine the bridging.[5] Such a wide discrepancy suggests that only a minority of patients with myocardial bridging are in fact at risk of clinical symptoms and cardiac events.

Myocardial bridging is usually seen in the setting of hypertrophic cardiomyopathy or left ventricular hypertrophy and is rarely reported in an otherwise structurally normal heart. The incidence of myocardial bridging among children with hypertrophic cardiomyopathy on coronary angiography varies between 28% and 40%.[1,6] As selective coronary angiography is not a routine procedure for children, the exact incidence of myocardial bridging among healthy children and patients, other than those having hypertrophic cardiomyopathy, is not known. Yet, symptomatic myocardial bridging probably occurs in a small subgroup of children, in whom ischaemic symptoms can be linked directly to their myocardial bridging. Sharma reported symptomatic myocardial bridgings in children. All patients included his study had significant symptoms of chest pain or syncope with exertion or cardiac arrest, supported by the evidence of coronary ischaemia linked to myocardial bridging, which ultimately resolved after mechanical decompression in short-term follow-up. Further, all his patients had hypertrophic cardiomyopathy or left ventricle hypertrophy, in contradistinction to our patient.[7]

https://doi.org/10.1017/S1047951113000875 Published online by Cambridge University Press

Myocardial bridgings are most commonly located over the middle segment of the left anterior descending coronary artery and may occasionally involve the diagonal branches, the posterior descending right coronary artery, and the marginal branch of the circumflex artery.[3] In our patient, two different parts of the left anterior descending artery followed an intramural course, causing a 70–80% and 100% narrowing during systole, respectively. Over 90% systolic compression of the tunnelled coronary segment with an ongoing diastolic compression of at least 50% characterises a significant myocardial bridging, as was seen in our case.[5] It is surprising that our patient never displayed symptoms, even during tremendous exercise.

Discrete subaortic stenosis can be seen in different members of the same family. The genetic basis of this disease is still indefinite. Some authors propose an autosomal recessive condition, whereas others propose an autosomal dominant inheritance pattern.[8] A multifactorial model of inheritance, in association with environmental factors, may be involved in the pathogenesis. A familial incidence of discrete subaortic stenosis, although very rare, signifies the screening of family members.[9] In our case, three members of the same family had mild subaortic stenosis.

Myocardial bridging is usually a benign condition with a long-term survival but can also cause myocardial ischaemia, myocardial infarction, exercise-induced tachycardia, conduction disturbances, and sudden death. Our patient was symptom-free in her daily life but had ischaemic findings during the exercise stress test. We estimate that her brother who died suddenly during exercise also had myocardial bridging with discrete subaortic stenosis. Therefore, we recommended her elderly brother to undergo coronary angiography, although he is asymptomatic.

In patients with myocardial bridging, medical or surgical management remains controversial. To date, however, there is no convincing evidence that surgery improves morbidity or mortality in patients with bridging.[10] Nitrates, β-blockers, and calcium channel blockers could be used for symptomatic relief. Surgery could be the choice for severe cases. Although our patient was asymptomatic but had a myocardial bridging segment of 100% narrowing, surgical decompression was performed. After surgery, she remained asymptomatic, and the signs of ischaemia on the exercise test resolved.

Our case is different from those reported in the literature, as our patient had neither left ventricular hypertrophy nor hypertrophic cardiomyopathy on echocardiography but had severe myocardial bridging on angiography. This case is also unique as the myocardial bridging was concomitant with familial subaortic stenosis.

Acknowledgements

We are grateful to Professor Dr Tevfik Gurmen for his kind contribution to the coronary angiography evaluation of the patient.

Financial Support

This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest

None.

Ethical Standards

Informed consent was obtained from parents of the patients before performing catheter angiography, and the study was reviewed and approved by the Ethical Committee of Cerrahpasa Medical Faculty, Istanbul University.

Supplementary materials

For supplementary material referred to in this article, please visit http://dx.doi.org/10.1017/S1047951113000875

References

1. Yetman AT, McCrindle BW, McDonald C, Freedom RM, Gow R. Myocardial bridging in children with hypertrophic cardiomyopathy. A risk factor for sudden death. N Eng J Med 1998; 339: 1201–1209.
2. Corrado D, Thiene G, Cocco P, Frescura C. Non-atherosclerotic coronary artery disease and sudden death in the young. Br Heart J 1992; 68: 601–607.
3. Bourassa MG, Butriaru A, Lesperance J, Tardif JC. Symptomatic myocardial bridges: overview of ischemic mechanism and current diagnostic and treatment strategies. JACC 2003; 41: 351–359.
4. Abdallah H, Toomey K, O'Riordan AC, Davidson A, Marks LA. Familial occurrence of discrete subaortic membrane. Pediatr Cardiol 1994; 15: 198–200.
5. Mohlenkamp S, Hort Wge J, Erbel R. Update on myocardial bridging. Circulation 2002; 106: 2616–2622.
6. Mohiddin SA, Begley D, Shih J, Fananapazir L. Myocardial bridging does not predict sudden death in children with hypertrophic cardiomyopathy but is associated with more severe cardiac disease. JAAC 2000; 36: 2270–2278.
7. Sharma J, Hellenbrand W, Kleinman C, Mosca R. Symptomatic myocardial bridges in children: a case report with view of literature. Cardiol Young 2011; 21: 490–494.
8. Petsas AA, Anastassiades LC, Constantinou EC, Antonopoulos AG. Familial discrete subaortic stenosis. Clin Cardiol 1998; 21: 63–65.
9. Fatimi SH, Ahman U, Javed MA, Shamim S, Ahmad R. Familial membranous subaortic stenosis: review of familial inheritance patterns and a case report. J Thorac Cardiovasc Surg 2006; 132: 1484–1486.
10. Downar J, Williams WG, McDonald C, Wigle ED, McCrindle BW. Outcomes after "unroofing" of a myocardial bridge of the left anterior descending coronary artery in children with hypertrophic cardiomyopathy. Pediatr Cardiol 2004; 25: 390–393.