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Diagnostic techniques in a case of patent ductus arteriosus: Is the computerized tomography angiography gold-standard method for the diagnosis of patent ductus arteriosus?

*Bir patent duktus arteriyozus vakasında tanısal tetkikler: Bilgisayarlı tomografi anjiyografisi
patent duktus arteriyozus tanısında altın standart yöntem midir?*

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Introduction

The patent ductus arteriosus (PDA) is the patency of ductus arteriosus connecting the descending aorta just distal to the left subclavian artery to the pulmonary trunk after fetal life. In the normal full-term newborn, the ductus arteriosus functionally closes shortly after birth due to increase in partial pressure of oxygen and decreased synthesis of prostaglandins (1). Anatomical closure completes after few weeks from birth, by intimal proliferation and fibrosis (2). Occasionally, ductus fails to close and can be seen at older ages uncommonly (3).

Case report

A 42-year-old woman applied to general health provider because of nonspecific upper respiratory tract infection. On routine physical examination, low-grade continuous murmur, peaking in late systole was heard incidentally in the second left intercostal space. Then the patient was referred to our clinic for further examination. Physical findings other than the low-grade murmur were unremarkable. Electrocardiogram, chest X-ray and laboratory tests were normal. Transthoracic echocardiography demonstrated an abnormal turbulent flow from the descending aorta to the pulmonary trunk in the suprasternal view. The left atrial and left ventricular dimensions and pulmonary arterial pressure were all normal. In order to determine the accurate anatomical localization, computerized tomography angiography (CTA)

was performed. It was reported firstly that an abnormal connection was present between the ascending aorta and the pulmonary artery in the axial 2-dimensional view (Fig. 1). Because of this type of connection is seen uncommonly at older ages and the patient was asymptomatic, selective aortic angiography was performed and the small PDA was seen between the descending aorta just distal to the left subclavian artery origin and the left pulmonary artery on lateral projection with 30 degrees cranial angulation (Fig. 2). The coronary angiogram was normal. After that, the patient was consulted by radiology department for reexamination of previous images and then, 3-dimensional reconstructed views showed the exact location of the communication between the descending aorta after the origin of subclavian artery and the left pulmonary artery (Fig. 3).

Discussion

The patent ductus arteriosus usually closes after birth and if it remains open, this may cause heart failure and pulmonary hypertension resulting from a large left-to-right shunt and rarely ductus arteriosus endarteritis (4, 5). In its simple isolated form, the patient with PDA is often asymptomatic. The age of patient at diagnosis varies from infancy to old age. The diagnosis can be made clinically and confirmed by echocardiography, CTA, magnetic resonance imaging or selective catheterization (6).

Transthoracic echocardiography is the initial diagnostic technique for the evaluation of patients with suspected congenital cardiac diseases. However, use of this modality is not sufficient due to several

limitations in the examination of great vessels, intracardiac anomalies, pulmonary veins and coronary arteries. Echocardiographic detection of PDA is based on the presence of left-sided cardiac chamber dilatation and the turbulent flow in the proximal pulmonary artery. Because

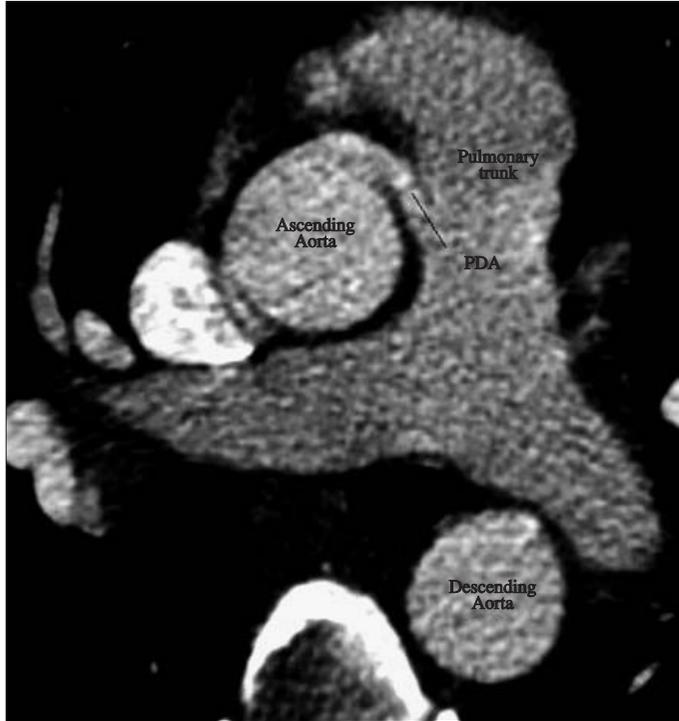


Figure 1. Computed tomography angiography 2-dimensional view of a connection between the ascending aorta and the pulmonary trunk (PDA)

PDA - patent ductus arteriosus

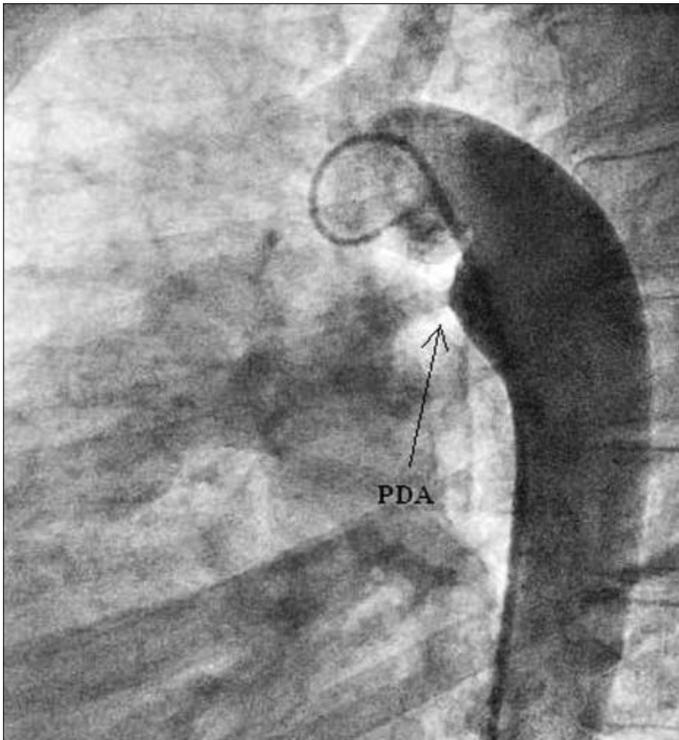


Figure 2. Aortography image of PDA between the descending aorta and the pulmonary trunk (arrow)

PDA - patent ductus arteriosus

asymptomatic PDAs are usually small, they can easily be missed during routine echocardiographic examination (7). Transesophageal echocardiography is highly sensitive and specific in the detection of PDA in adolescents and adults, but it is invasive and necessitates local or sometimes general anesthesia (8).

Computerized tomography angiography is an examination using X-rays to visualize blood flow in arterial and venous vessels throughout the body. It is a promising noninvasive technique for the evaluation of the congenital cardiac anomalies and complex cases with concomitant abnormalities can be accurately examined by using multiplanar reconstruction images. Computerized tomography angiography provides detailed information about the precise location and size of PDA, presence of calcification and relationship to adjacent anatomic structures (7).

The CTA images of blood vessels anywhere in the body may be misevaluated if the patient moves during the examination or the heart is not functioning normally. Computerized tomography angiography is not yet able to show small, twisted vessels precisely in rapidly moving organs. It is a common practice to use the β -blocking agents to maintain heart rates at a level below 80 beats per minute during image acquisition. The pulsation of great vessels may lead to misdiagnosis about the exact location of these types of congenital anomalies. Besides the motion artifacts, other factors, such as the presence of high attenuation structures like the calcified ductus, can interfere with the accurate evaluation of the lumen and the ductus may appear oversized (9).

Conclusion

In conclusion, the asymptomatic PDAs are often small and they can be inadvertently missed unless special attention is directed to this region during routine examination or evaluation for other cause.



Figure 3. Three-dimensional and colored computed tomography angiography view of the PDA (arrow).

Ao - aorta, LPA - left pulmonary artery, PDA - patent ductus arteriosus

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Surgical repair of supra-avalvular aortic stenosis in Williams syndrome

Williams sendromlu bir olguda supra-avalvüler aort stenozunun cerrahi tedavisi

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Introduction

Williams syndrome is a rare genetic condition which occurs in 1/7500 births, and causes cardiovascular, developmental and intellectual problems. The syndrome is characterized by varying degrees of left ventricular outflow tract obstruction beginning superior to the aortic valve, hypercalcemia, elfin faces, and mental retardation (1).

Case report

In April 2007, a 17-year-old boy was referred to our clinic because of a cardiac murmur. He had moderate mental retardation documented with WISC-R test. Aortography, coronary angiography, echocardiography and 3-D computed tomography were performed before the operation, and all demonstrated a typical hour-glass type stenosis just above the aortic valve (Fig. 1-2). Computed tomography revealed minimal stenosis of left pulmonary artery (Fig. 3). The preoperative peak systolic pressure gradient between the left ventricle and ascending aorta was 110 mmHg. There was no coronary stenosis, other cardiac anomalies or renal artery stenosis.

We performed successful surgical repair of extended aortoplasty using Doty's method (two sinus reconstructions) for congenital supra-avalvular aortic stenosis. After standard cardiopulmonary bypass, bi-sinusal inverted Y incision was made between noncoronary cusp and right coronary cusp; and the ridge was removed with sharp dissection above the left coronary sinus. Resulting defect on the aorta was then repaired with Dacron patch (Fig. 4). The systolic pressure gradient between the left ventricle and ascending aorta improved to

normal postoperatively. Peroperative gradient between the main pulmonary artery and pulmonary artery was 22 mmHg on the right and 10 mmHg on the left and did not require surgical correction. The patient was extubated 7 hours following surgery, and was discharged at the 5th postoperative day. Six months after the procedure, the patient was fine and had no complaints.

Discussion

Supra-avalvular aortic stenosis is an unusual form of obstruction of the left ventricle outflow tract, which occurs in 3% to 6% of cases of various types of aortic obstruction. It may be due to the presence of a discrete fibrous membrane, an hourglass narrowing, or a diffuse narrowing. The anatomical variants of supra-avalvular aortic stenosis affect post surgery results and survival rates, e.g. the hourglass type of obstruction is the most common, whereas the diffuse type of stenosis mostly results in death (2). The syndrome was first described in 1930 by Mencarelli, and its association with other facial defects and psychomotor retardation was established in 1961 (3). In 1965, Logan described patients who had a familial form of supra-avalvular aortic stenosis with normal face and normal mental development, which may delay diagnosis (4, 6). Supra-avalvular aortic stenosis is a familial autosomal dominant disorder with variable expression and affects both sexes equally. The vascular pathology of familial supra-avalvular aortic stenosis and Williams syndrome results from mutations involving the elastin gene on chromosome 7q11.23 (5).

In 1961, McGoon et al. proposed widening the aorta diameter with a synthetic graft as a treatment option. In 1977, Doty used his