Pseudocoarctation of the Aorta

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SUMMARY

In this report 5 patients between the ages of 5–14 years admitted to the Department of Pediatric Cardiology, Hacettepe University, and diagnosed as having pseudocoarctation of the aorta are presented. The clinical and angiographic findings of this rare abnormality of childhood are discussed, the importance of differential diagnosis of this entity from coarctation of the aorta is emphasized, and the literature reviewed.

Additional Indexing Words:
Pseudocoarctation of the aorta Kinking of the aorta

PSEUDOCOARCTATION of the aorta is a rare condition resulting from abnormal elongation of the aortic arch with redundancy and kinking of the aorta at the level of the ligamentum arteriosum.[1)–3)] The anomaly is believed to be due to failure of normal embryologic compression.[2)] Pseudocoarctation may be an isolated anomaly or it may be associated with other congenital cardiac defects.[4)] Five patients with a diagnosis of pseudocoarctation of the aorta made by the Department of Pediatric Cardiology, Hacettepe University are presented, since this anomaly is usually regarded as being rare in childhood.

CASE REPORT

Between the years 1979–1989 5 patients were diagnosed as having pseudocoarctation of the aorta by the Department of Pediatric Cardiology, Hacettepe University. All the patients were male and aged between 5–14 years. The cases were asymptomatic and they had been referred to our hospital for further evaluation of cardiac murmurs.

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Table I. Summary of the Clinical and Laboratory Findings

Case	Age	Blood pressure Arm-Leg (mmHg)	Cardiac catheterization	Angiocardiography	Associated cardiac defects
1. A. D.	14	100/70- 90/70	120 mmHg pressure gradient between left ventricle and aorta	Dilated ascending aorta and pseudocoarctation at isthmus	Valvular aortic stenosis and mild dilatation of the left coronary artery
2. T. K.	7	100/70- 90/70	Normal	〃	Bicuspid aortic valve
3. D. S.	5	130/90-120/90	Patent foramen ovale	〃	None
4. Z. O.	8	100/50- 90/40	Patent foramen ovale	〃	None
5. M.D.	10	90/40- 90/40	Small VSD (Qp/Qs=1. 2)	Dilated ascending aorta and pseudocoarctation at isthmus, minimal left to right shunt at the level of ventricle	Small VSD

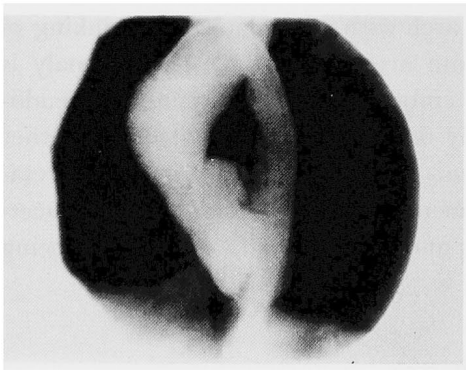

Fig. 1.

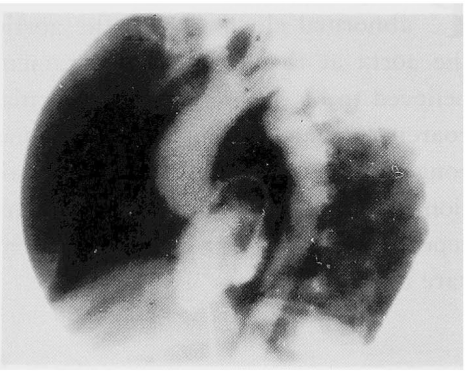

Fig. 2.

Fig. 1. Left ventriculography in left anterior oblique view of the second case shows a pseudocoarctation of the aorta and dilated ascending aorta.

Fig. 2. Left ventriculography in left anterior oblique view of the third case shows an elongated aortic arch with kinking and aneurysm formation of the ascending aorta.

On physical examination, blood pressures in the upper and lower extremities were within normal limits. Femoral artery pulses were present. Heart sounds were normal and in 4 patients 2–3/6° systolic ejection murmurs were heard in the left 2nd to 4th intercostal spaces while in 1 patient a thrill in the jugular notch and a systolic ejection murmur in the right 2nd intercostal space were heard. The electrocardiograms revealed normal QRS axes in the frontal plane with left ventricular dominance in 3 patients and left ventricular hypertrophy together with ST segment depression and T

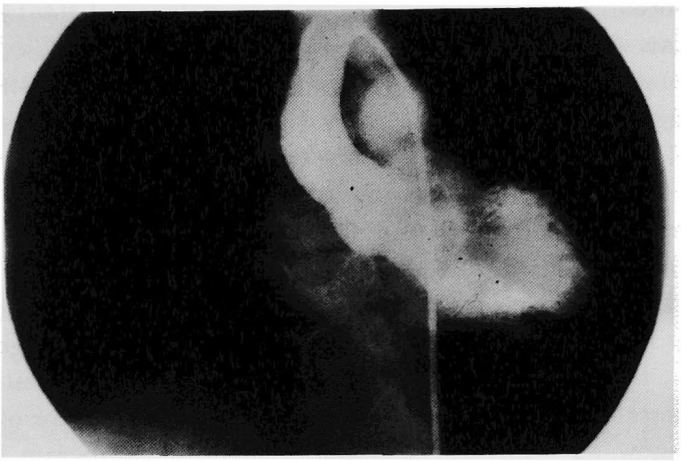

Fig. 3. Left ventriculography in antero-posterior view of the fifth case shows an elongated aortic arch with kinking and aneurysm formation.

wave inversion in 1 patient. The roentgenograms showed a prominent aortic knob in 3 patients and mild cardiomegaly in 1 patient. The echocardiographic examinations revealed a bicuspid aortic valve in 1 patient. In another case a pressure gradient of 20 mmHg was found at the distal portion of the pseudocoarctation. Angiocardiograms of the cases showed a dilated, elongated and kinked ascending aorta with no segmental coarctation. In the patient with a bicuspid aortic valve, left cardiac catheterization revealed no pressure gradient across the kinked area. The aortogram of this patient showed no segmental coarctation. In addition there was no evidence of rib notching or collateral circulation. Associated with the pseudocoarctation in one case was a small ventricular septal defect. Another case had valvular aortic stenosis and a dilated left coronary artery. Surgery is planned for the patient with aortic stenosis, and clinical follow-up is planned for the other cases (Table I, Figs. 1, 2 and 3).

Discussion

Pseudocoarctation of the aorta was first defined by Rosler and White in 2 adult cases.[5] Sauders and associates[6] described clinical findings of pseudocoarctation in 1951 and Dotter and associates[7] defined angiographic findings of this anomaly in the same year. Dungan and associates[8] showed that the systolic pressure gradient across the pseudocoarctation was either normal or less than 25 mmHg.

The absence of systemic hypertension, collateral circulation in the thoracic aorta and positive pulsation of the femoral artery should differen-

tiate pseudocoarctation from true coarctation of the aorta.[8),9)] However in some patients the femoral artery pulse may be weak. These weak pulsations may be explained by the elongated aorta characterized by a sharp anterior angulation of the aortic arch at the level of the ligamentum anteriosum which causes slow blood flow.[3),8)]

Cases of pseudocoarctation are seen with equal frequency in males[8)] and females and all age groups and they are asymptomatic as in our patients. A systolic murmur at the base with radiation to the neck was heard in all cases. This murmur has been attributed to a change from laminar to turbulent flow in the region of the anomaly. Because of the elongation of the aortic arch, the chest x ray shows the appearance of a mediastinal mass.[4)] The esophageal barium x ray in the left lateral or left anterior oblique position may show an "S" or "3" shape.[10),11)] An aortogram is often required to show the kinked portion of the aorta.[1)]

Pseudocoarctation of the aorta is usually an isolated anomaly. However it may be associated with patent ductus arteriosus, ventricular septal defect, aortic stenosis, subaortic stenosis, transposition of the great arteries, atrial septal defect, anomalies of the aortic arch, aneurysm of the sinus of Valsalva, corrected transposition and bicuspid aortic valve.[3),4),8),9),12)] The incidence and distribution of associated cardiac anomalies are parallel with those seen in true coarctation.[9)] Three of our patients had different types of associated cardiac anomalies. One had a ventricular septal defect, the second had valvular aortic stenosis and the third patient had a bicuspid aortic valve. Three patients with a bicuspid aortic valve lesion associated with pseudocoarctation of the aortic arch were presented by Angelini and his associates in 1985. They believed that pseudocoarctation cases with associated bicuspid aortic valve disease become symptomatic, usually in later life and require surgical repair of the bicuspid aortic cusp.[9)] Keller and Cheitlin[13)] described a family with coarctation of the aorta associated with pseudocoarctation. Sometimes pseudocoarctation is part of a syndrome of complex malformations, e.g. Turner's, Noonan's and Hurler's syndromes.[11)] Winer and associates reported a proximal mid-aortic coarctation associated with pseudocoarctation.[2)]

In the differential diagnosis, aortitis syndrome (Takayasu's disease) must be taken into consideration in children. In these patients arteriography shows narrowing and/or dilatation of the affected arteries.[14)] These findings were not seen in our cases and they had no acute symptoms of aortitis syndrome. Their blood pressures in the upper and lower extremities were within normal limits and peripheral arterial pulses were present.

Aneurysm formation and mediastinal mass are important alternatives

in the differential diagnosis of pseudocoarctation.[4),6),8),11),12),15)] Pattinson and Grainger[16)] reported a case who had an unnecessary exploratory thoracotomy because of an abnormal left superior mediastinal opacity on the posteroanterior chest film. Surgery is not necessary in pseudocoarctation of the aorta. However an aneurysm may occur in the elongated and kinked segment of the aortic knob, and for this reason these cases should be examined regularly.[12),17)] One patient was reported to have died preoperatively from rupture of a dissecting aneurysm located distal to the pseudocoarctation.[18)] When an aneurysm occurs, surgical resection is the treatment of choice.[12)]

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