A Case of Kinking of the Aortic Arch

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KINKING of the aorta is a rare anomaly in which that part of the aortic arch that passes over into the descending aorta has a double kinking in the shape of a double S. The kink lies toward the front and somewhat to the right at the level of the ligamentum arteriosum, where aortic coarctation most frequently occurs. The superposed shadow of the kinking causes an opacity on the left of the mediastinal mass parasternally. The pulsating mass “above” the aortic knuckle represents the proximal part of the kinked aortic arch, and the apparent aortic knuckle is the distal portion of the arch. There is no apparent vascular obstruction or stenosis, but there is a very marked elongation of the kinked aortic arch.

The uncommon finding of an elongated thoracic aorta was first described by Rösler and White in 1931.1 They did not apply a new term for the malformation. The first clinical description was offered by Souders and associates in 1951.2 Their case had been treated as Hodgkin’s disease and because there was no response to x-ray therapy the patient was operated upon and the malformation of the aorta was found. They called the malformation “a subclinical form of coarctation.”

There followed 7 other publications3-9 under different titles: pseudocoarctation, aneurysm of the arch, etc., yet none of these terms is appropriate owing to ambiguity. DiGuglielmo and Guttadauro10 were the first to introduce the term “kinking of the aortic arch,” which is accurate, both anatomically and clinically, in that it describes a tortuosity and a curvature in the elongated aortic arch. To date a total of about 30 cases has been described in the world literature,11-13 9 of these in Europe.

Most cases were free from symptoms and were first noticed on radiographic examination. In 1 of DiGuglielmo’s cases it occurred in association with a ventricular septal defect.

All described cases exhibited far less tortuosity than our case, with scant clinical symptoms. A systolic murmur was heard in 2 cases above the aorta, and in 2 cases it was heard posteriorly. In only 1 case did it occur in the left intercostal region.

Another case of this unusual anomaly is reported here.

Case Report

A 21-year-old bricklayer suffered the onset of the present illness 4 months before admission to the University Hospitals, Zagreb, with the following symptoms: choking and a stabbing pain in the left chest from the left shoulderblade downward and toward the front; a sensation of a “current” in the left side of the neck and a feeling that “his head did not seem to belong to him,” especially when moving it to the right; easy fatigue; rapid palpitation; and a sensation of heat all over the body. These difficulties were much increased with excitement.

He was of normal development and average body structure. Pulsation of the right carotid artery was more pronounced than the left one. The blood pressure readings were in the left arm 135/95 to 150/100, in the right arm 145/95 to 165/100, in the left leg 160/130, and in the right leg 155/120. On palpation the apex beat was prominent, with a strong impulse from the left ventricle. A slight thrill was palpated above the aorta, and the second sound could be felt. On auscultation the sounds were loud. The first sound was split, the second component being a characteristic “systolic click.” The second sound above the aorta was accentuated and metallic, but the accentuated second sound disappeared 1 fingerbreadth from the sternal border. At the pulmonic area a weak systolic murmur was heard, which spread to the supraclavicular region and to the back in the interseapular area (fig. 1). No abnormal femoral or other arterial pulsation was palpated.

The electrocardiogram, circulation time, venous pressure, and oscillographic examination of arms and legs were within normal limits. The basal metabolic rate was +19 per cent.

X-ray of the lungs and heart (fig. 2) showed a semicircular opacity in the left parasternal area,
Auscultatory findings:

- systolic click
- accentuated second sound
- systolic murmur

Figure 1

Auscultatory findings.

seemingly located above the aortic arch and exhibiting marked arterial pulsation. No rib erosion was noticed.

Our patient did not exhibit clinical symptoms indicating the presence of an aortic ring. The esophogram presented a pattern contrary to that observed in aortic rings, and excluded this diagnosis. Coarctation of the aorta could not be excluded completely but could not be diagnosed in the absence of any positive findings. Hence the radiologist and we reached the conclusion that we had encountered a special malformation that could not be diagnosed without angio-cardiography. It disclosed a tortuosity or kinking of the aorta (figs. 3 and 4).

Discussion

In our patient the "streaming" sensation in the left side of the neck and the irradiating pain may be ascribed to the anomaly. The auscultatory findings were interpreted as follows: the systolic click indicates a vascular sound, the phenomenon is found in dilatation of the aorta or of the pulmonary artery. In calling it a click, we wish to emphasize that the sound is not associated with the closure of the mitral or tricuspid valves creating the first sound. The accentuated second sound above the aorta can be only partly explained by the hypertension, as it is heard very rarely indeed in such a mild rise of blood pressure (150/100) in a young subject. We believe that the aortic second sound was accentuated because the ascending aorta lay very close to the anterior thoracic wall because of the increased anterior curvature. In accordance with this possibility was the abrupt disappearance of this second sound exactly where the aorta curves backward, to the left of the sternum. The systolic murmur is the most difficult to explain, as, in theory, it ought not to be heard in an anomaly without either stenosis or shunt. The curvature acts hemodynamically as a sort of aneurysm, however, imposing resistance and turbulence at the sudden bends. From the embryologic aspect the anomaly develops on a similar basis as coarctation of the aorta, except that there is no stenosis.

With the exception of slight hypertension the patient usually does not experience marked disturbance. The prognosis, however, is a matter of conjecture. Pathophysiologically the anomaly appears similar to an aneurysm. Instead of the usual wide aortic arch there exist 3 curves, which sustain the full impact of the blood stream that pounds against the bends and produces an enlargement. As long as the wall remains elastic there is no impending danger of aneurysm; it becomes a menace.
only with the onset of atherosclerosis. The stretching of the media then creates a vicious circle. In the presence of an aneurysm the lateral pressure increases on the site of convexity and the artery widens, causing further damage to its wall. Hence aneurysms may appear in the parts most exposed to the blood stream, with possible pressure upon the surrounding organs and possibly even rupture.

The anomaly has not yet been submitted to surgery, and we do not consider it indicated in our case. The surgical risk would exceed by far the effect to be expected in cases of such slight discomfort to the patient.

Summary

A case of uncommon aortic anomaly has been described. The clinical signs as well as the differential diagnosis are stressed.

Summario in Interlingua

Es desriebite un caso de un incommun anormalitate aortie. Le signos clinie e le diagnose differential es presentate in detalio.

References

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Circulation. 1960;21:1147-1149
doi: 10.1161/01.CIR.21.6.1147

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/21/6/1147

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