ANEURYSM OF THE AORTIC SINUSES WITH PSEUDO-COARCTATION OF THE AORTA

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Aortic sinus aneurysms are uncommon; association with pseudocoarctation has not been previously recognized. Morgan Jones and Langley (1949) in a classic paper reviewed forty-three autopsy proven cases, added four new ones, and divided them into congenital and acquired types: of the total, twenty-five were congenital. Venning (1951) reported three additional cases and Falholt and Thomsen (1953) in describing a case of unruptured congenital right aortic sinus aneurysm diagnosed by retrograde aortography increased the number of congenital sinus of Valsalva aneurysms to thirty-four. Recently, Besabe et al. (1954) reported a case of congenital aortic sinus aneurysm with rupture into the right atrium and embolic gangrene of a foot; at necropsy ulcerated bacterial endocarditis of a bicuspid aortic valve was found.

Sixteen cases of unruptured aortic sinus aneurysms have been diagnosed during life by angiocardiography at this centre. Seven were congenital, three associated with coarctation of the aorta (Dublicher et al., 1955), one other with coarctation of the aorta and subacute bacterial endocarditis (Steinberg and Finby, 1955), three with arachnodactyly (Steinberg and Geller, 1955), and nine acquired, due to syphilis (Merten et al., 1955). The seventeenth case of unperforated aortic sinus aneurysm herein reported is unique because of the association with pseudo-coarctation of the aorta.

CASE REPORT

A 26-year-old married sewing-machine operator referred by Dr. Raymond Miller was admitted with complaint of backache of two years duration. When 13 years old, she had aching bones and joints unaccompanied by fever for a week. The next year she fainted on two occasions and after examination was told she had "rheumatism of the heart." Her activities were restricted and she was well until two years ago when she was in a motor accident. Although the car in which she was a passenger overturned, she sustained only superficial injuries to the thorax; and since then has had much backache. There had been no dyspnea, orthopnea, or edema, and in addition to her household duties she had been steadily employed.

Physical examination showed a well developed and nourished woman in no distress. The only abnormal finding was a grade III systolic murmur heard all over the base of the heart, loudest along the left sternal border and transmitted to the neck and axilla. A loud systolic murmur was also heard along the course of the aorta posteriorly and at the epigastrium. The radial, abdominal, femoral, popliteal, and dorsalis pedis pulses were palpable. Blood pressure readings were: right arm, 126/90; left arm, 130/90; right leg, 140/120; and left leg, 132/110.

Laboratory data were as follows: hemoglobin, 12 g. per 100 ml.; erythrocytes, 4.6 m. /cu. mm.; hematocrit, 38 per cent; the differential blood count and smear were normal. The electrocardiogram showed normal sinus rhythm, rate 88 a minute, no deviation of the electrical axis, P-R II=0.16 sec. and QRS II=0.07 sec. The standard limb and unipolar leads were normal and the heart was in the vertical position.

The frontal teleroentgenogram (Fig. 1A) showed slight left ventricular prominence. A mass (arrow) was seen above what appeared to be a dilated aortic knob. Rib notching was not detected. The left anterior oblique view revealed bowing of the posterior wall of the trachea by a dilated descending aorta

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Fig. 1.—(A) Conventional frontal teleroentgenogram shows some prominence of the left ventricle. A mass (arrow) is present above what appears to be an aortic knuckle. (B) Left anterior oblique view shows anterior bowing of the posterior wall of the trachea by a dilated descending aorta. (C) Frontal oesophagram shows indentation of the oesophagus by a dilated descending aorta; a mass just above is indicated by arrow. No rib notchings are seen.
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Fig. 2.—(A) Frontal angiocardiogram at 1-5 seconds after the beginning of injection shows opacification of normal right cardiovascular structures. Note the indentation of the superior vena cava. (B) At 7 seconds, the left cardiovascular structures are opacified. Note the aneurysmal dilatation of the aortic sinuses, the dilated ascending aorta and aortic arch and left brachiocephalic vessel anomalies. The descending aorta is dilated but arterial collaterals are not present.

Fig. 3.—(A) Left anterior oblique angiocardiogram at 1-5 seconds. The right cardiovascular structures are normal. Note the filling defect adjacent to the superior vena cava and outflow tract of the right ventricle. (B) At 7 seconds, the aneurysmal dilatation of the aortic sinuses, the dilated ascending aorta and deformed aortic arch and dilated descending aorta are demonstrated. The internal mammary arteries are of normal size.
Serial angiocardiography disclosed slight displacement of the terminal end of the superior vena cava at its entrance into the right atrium (Fig. 2A). A deformity of the superior part of the right atrium was also seen in Fig. 3. Fig. 2B and 3B showed that the filling defect adjacent to the superior vena cava was due to the aneurysmal dilatation of the aortic sinuses which measured 50 mm. in diameter. The ascending aorta (Fig. 2B and 3B) appears dilated and measured 40 mm. (average normal 38 mm.). The innominate artery was also dilated (20 mm.) and the transverse aorta abruptly narrowed and became rounded with a dilated (10 mm.) left subclavian branch at its upper pole. Below, the descending aorta became large and bulbous measuring 45 mm. in diameter. The dilated contour continued almost to the diaphragm. The internal mammary arteries were visualized and appeared normal in calibre. No collaterals were recognized (Fig. 3B).

Simultaneous direct brachial and femoral arterial pressure recordings were made and interpreted by Dr. Daniel S. Lukas. The readings were: brachial artery, 117/86 (mean 105); femoral artery, 119/86 (mean 102). A suggestive anacrotic limb and moderate rounding of the systolic phase of the brachial arterial pressure curve was present; but was not distinctive enough to warrant the diagnosis of aortic stenosis. The onset of systole in the femoral artery was 0.04 sec.

**DISCUSSION**

The aortic sinuses are three small dilatations in the wall of the aorta immediately above the valves. Each sinus lies just above the attachment of an aortic cusp. The right and left coronary arteries usually originate within two of the aortic sinuses; however, they may emerge immediately distal to the aortic sinuses. The aortic sinuses and their corresponding arterial valves are named according to the source of the coronary arteries. Thus, the right and left coronary arteries arise from the right and left aortic sinuses respectively. The remaining aortic sinus usually lies posteriorly and does not contain a coronary artery and according to the newest nomenclature is designated the non-coronary sinus.

The aortic sinuses are intracardiac and cannot be identified on conventional roentgenography. During angiocardiography they appear as dilatations at the root of the aorta immediately above the aortic valves and are best visualized in the left anterior oblique view (Dotter and Steinberg, 1951). Inconstant filling of the coronary arteries during angiocardiography does not always allow individual identification of the sinuses. However, in the left anterior oblique view the right coronary sinus is regularly anterior and just behind the sternum. The aortic sinuses are in close relation to all the cardiac chambers, particularly the right atrium and ventricle. The origin of the pulmonary artery, the interventricular septum, and left atrium are adjacent while the superior vena cava is more distant.

Anomalies of the aortic arch and descending aorta may simulate a superior mediastinal tumour. Sanders et al. (1951) reported three such cases: their first was found to have a short patent ductus arteriosus which was ligated but failed to affect the aortic deformity; the two others were suspected after conventional roentgenography and confirmed in one instance by angiocardiography.

Subclinical or pseudo forms of aortic coarctation became recognizable with the advent of angiocardiography (Dotter and Steinberg, 1951). A total of ten such cases have been encountered at this centre. Usually the patient is asymptomatic and is referred because of the finding of a pulsatile mediastinal swelling. On angiocardiography the nature of the aortic deformity is clearly delineated and easily differentiated from coarctation by the demonstration of an aortic lumen and the absence of arterial collaterals. In the case herein reported (Fig. 3B) although arterial collateral vessels were absent, the abrupt termination of the aortic arch, and the dilatation of the descending aorta favoured the diagnosis of coarctation. Indeed, the diagnosis of pseudo-coarctation depended upon the absence of any change in the blood pressure in the legs.

**SUMMARY**

A patient with aortic sinus aneurysm was found to have an associated pseudo-coarctation of the aorta. This is the eighth case of congenital aortic sinus aneurysm diagnosed during life by angiocardiography and the first associated with pseudo-coarctation.
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REFERENCES