Subvalvular and Apical Left Ventricular Aneurysms in the Bantu as a Source of Systemic Emboli


SUMMARY

Systemic emboli originating from apical and subvalvular left ventricular aneurysms are described in two Bantu patients. The infrequency of embolism from these aneurysms is probably related to the fact that such aneurysms usually have small ostia. The pertinent literature is reviewed and the similarity between these aneurysms, congenital epicardial cysts, and fibrous diverticula is discussed. It is postulated that these conditions share a common etiology, namely, a congenital weakness in the left ventricular wall in the form of a persistent primitive endothelial-lined channel. The high pressure in the left ventricle converts these channels into fibrous aneurysms with ostia lined by endocardium at the two sites where the ventricular wall is thinnest, namely, the apical and subvalvular areas.

Additional Indexing Words:
Diverticulum of heart  Epicardial cyst  Arterial occlusion

Systemic Emboli are rare in young people unless associated with rheumatic heart disease, and their occurrence suggests the presence of an unusual condition. The purpose of this communication is to report systemic emboli due to apical and subvalvular left ventricular aneurysms and to discuss the etiology of these aneurysms.

Subvalvular left ventricular aneurysms occurring in intimate relationship to the mitral and aortic valves have been described in Nigerians,1,2 the South African Bantu,3-7 the Indian,8 and the American Negro.9,10 Left ventricular aneurysms of uncertain etiology have been encountered in the South African Bantu not only in the subvalvular position but also at the apex of the ventricle.5,11 The subvalvular type is more common, and in only three of the 14 cases seen by us have the aneurysms been situated at the apex. Virtually all subvalvular and apical aneurysms have been described in Negroes, although a subvalvular aneurysm has been diagnosed antemortem by Pocock and associates12 in an 18-year-old white man.

Subvalvular aneurysms have a varied clinical presentation. Distortion of the mitral and aortic annuli produces incompetence of these valves while compression of a coronary artery may cause coronary insufficiency or occlusion.4 The occasional patient has been asymptomatic, but the majority have presented with either congestive cardiac failure, pulmonary edema, angina pectoris, or cardiac tamponade. Ventricular tachycardia13 and subacute bacterial endocarditis4 are uncommon modes of presentation.

In our experience, apical aneurysms are less likely to present in congestive cardiac
failure. This is probably related to the fact that their narrow ostia restricts the amount of regurgitant flow into the aneurysmal sac during ventricular systole. This flow, however, may be sufficient to produce ejection systolic and early diastolic murmurs. Rupture of an apical aneurysm has also been reported.

With the exception of one possible example of a subvalvular left ventricular aneurysm in which death was due to cerebral hemorrhage (in Abrahams and associates' case 4), the occurrence of systemic emboli from subvalvular and apical left ventricular aneurysms has not yet been reported in approximately 40 documented cases. In this paper, two cases of left ventricular aneurysms are described in which systemic emboli occurred.

Report of Cases

Case 1

A 13-year-old Bantu female had been hemiplegic on the right side for one year. This had been of sudden onset. Her speech was unaffected and she denied any other symptoms. An abnormal cardiac shadow was detected on a chest radiograph and she was referred to Baragwanath Hospital for investigation.

Examination revealed an intelligent, well-developed girl. She had spastic right hemiplegia and right facial paralysis. The pulse was 90 per minute and regular. All the peripheral pulses were palpable and equal. The blood pressure was 110/80 mm Hg. The jugular venous pressure was normal, the liver was not palpable, and there was no edema. There was a diffuse heave over the entire precordium. The maximum cardiac impulse was located in the fifth intercostal space, 1 inch outside the midclavicular line and had a heaving quality compatible with left ventricular hypertrophy. A third heart sound was audible at the apex but there were no murmurs.

X-rays of the chest (figs. 1 and 2) revealed a large protuberance in the region of the left ventricle containing many areas of calcification. An electrocardiogram showed sinus rhythm and a mean frontal plane axis of plus 30°. Deep Q waves, elevated S-T segments, and inverted T waves were present in leads aV, V, to V, and standard leads I and II.

A left ventricular cineangiogram demonstrated a large aneurysm (fig. 3) with a wide ostium situated at the apex of the ventricle. The mitral valve was competent.

A full blood count and Westergren sedimentation rate were normal. The Wassermann reaction was negative.

In view of the risk of further embolism and the possibility of rupture of the ventricle, surgery
Case 1. Single frame from forward cineangiogram (posteroanterior view) performed from main pulmonary artery, showing large apical aneurysm with a broad ostium arising from the apex of the left ventricle. Abbreviations: LV = left ventricle; AN = aneurysm.

Figure 3

Case 2. Left ventricular angiogram (retrograde Seldinger technique performed in left anterior oblique position). A small posterior submitral aneurysm has filled with dye. Abbreviation: AO = aorta; LV = left ventricle; AN = aneurysm.

Figure 5

was recommended. Operation was performed under cardiopulmonary bypass (Mr. L. du Plessis). A large pulsatile aneurysm was found at the apex of the left ventricle extending onto the left leaf of the diaphragm. Adhesions were present between the pericardium and the aneurysm. The aneurysm was excised and the ostium, which was lined by endocardium, was closed with a Teflon patch. The postoperative period was uneventful. On macroscopic examination, the aneurysmal sac was lined with a mural thrombus. Histological examination of the sac showed it to consist of dense, avascular, nonspecific fibrous tissue; there was no evidence of any inflammatory reaction.

Case 2

A 35-year-old Bantu male was referred for investigation of an abnormal cardiac silhouette which had been detected at a routine radiographic examination. He denied any symptoms. Examination revealed a well-built man, comfortable at rest. The pulse was 80 per minute and regular. The left radial and brachial pulses were weaker than the right and were slow-rising in character. All the other pulses were of good
LEFT VENTRICULAR ANEURYSMS

Figure 6

Case 2. Angiogram performed at the origin of the left subclavian artery (Subcl. A). Occlusion of this vessel just distal to the vertebral artery (Vert. A) is demonstrated (see arrow).

volume. The blood pressure was 180/125 mm Hg in the right arm and 150/100 mm Hg in the left arm. The apex beat was in the fifth interspace on the midaclavicular line and had a heavy component with left ventricular hypertrophy. An atrial sound and a third heart sound were audible at the apex. The aortic component of the second heart sound was increased in intensity. The fundi showed grade 2 hypertensive retinopathy. The electrocardiogram revealed sinus rhythm and a mean frontal plane axis of −15°. The T waves were inverted in leads V5, V6, aVL, and standard leads I and II. In addition, abnormal Q waves were present in standard lead I and aVL.

X-rays of the chest (fig. 4) showed prominence of the ascending aorta and a large bulge which contained circular areas of calcification, on the left cardiac border. A left ventricular angiogram in the left anterior oblique position demonstrated a small aneurysm with a narrow ostium in the posterior subvalvular position (fig. 5). The calcified mass itself failed to opacify. There was no mitral or aortic regurgitation. The left subclavian artery was occluded just distal to the origin of the vertebral artery (fig. 6).

Since the larger submitral aneurysm was almost certainly thrombosed, it was decided to treat the patient conservatively with anticoagulants and antihypertensive drugs.

Discussion

It is clear that the hemiplegia in case 1, and the occlusion of the left subclavian artery in case 2, were due to embolism. The systemic hypertension in case 2 may also have resulted from renal emboli.

According to Schlichter and associates, thromboembolic phenomena occur in 51.9% of left ventricular aneurysms secondary to myocardial infarction. It is therefore surprising that systemic embolism has not been previously reported with subvalvular or apical aneurysms, especially as they often contain mural thrombi. It is possible that, as experience with these aneurysms increases, other cases will be reported; however, the apparent rarity of emboli may be related to the fact that the ostia of these aneurysms are usually small. A large ostium would presumably facilitate the discharge of an embolus; it is thus noteworthy that the aneurysm in case 1 was exceptional in that it had a wide ostium.

In most instances, left ventricular aneurysms are a consequence of myocardial infarction. However, syphilis, tuberculosis, rheumatic myocardial necrosis, trauma, mycotic infection, polyarteritis nodosa, Loeffler's parietal endocarditis, and anomalous origin of the coronary artery from the pulmonary trunk are other causes which have been documented. There has been no evidence to incriminate any of these conditions as likely etiological factors for subvalvular or apical aneurysms. However, since their ostia are lined by endocardium, a congenital etiology is possible. In view thereof, a consideration of the pathology of so-called congenital diverticula and congenital epicardial cysts is relevant to the discussion of the etiology of these aneurysms.

Two types of congenital diverticula of the left ventricle have been described, namely,
muscular and fibrous. The muscular type arises from the apex of the ventricle. They are frequently accompanied by midline abnormalities such as defects in the pericardium, diaphragm, and anterior abdominal wall, and may therefore present as a pulsatile tumor at the umbilicus.22-25 In addition, they are often associated with cyanotic congenital heart disease.26, 27 It is thus unlikely that subvalvular and apical aneurysms are related to this type of diverticulum.

The fibrous type of congenital diverticulum of the left ventricle occurs either in the apical or the subvalvular position. Drennan and Van der Vliet28 reported the case of a 4-year-old African female who died suddenly following rupture of a small “cyst-like diverticulum” situated at the apex of the left ventricle. At the site of the perforation, the endocardium was continuous with the fibrous wall of the cyst. These authors also found that in 15 postmortem specimens of normal hearts, the average thickness of the left ventricular wall was 13 mm, whereas that of the apex was only 2 mm. They postulated that a congenital weakness in the apical area could give rise to an aneurysmal protrusion. Swyer and associates,29 described a Caucasian infant who died of a hemopericardium 38 hours after birth due to rupture of a small “diverticulum.” The sac was composed of fibrous tissue and contained thrombus. The description of this diverticulum, in which the endothelial-lined ostium was situated behind the anterior cusp of the mitral valve, was compatible in all respects with a submittal aneurysm. Swyer and associates,29 postulated a defect in the muscular attachment of the left ventricle to the mitral ring, and that subsequent “outpouching” and aneurysm formation resulted from the high pressure in the left ventricle.

Fibrous diverticula are probably related to “congenital epicardial cysts.” Patterson and Spink30 reported a 2-week-old infant (race not stated) who died following rupture of such a cyst which was situated in the left atroventricular groove. The wall had an inner endothelial coat, a middle coat consisting largely of fibrous tissue but with sparse non-striated muscle, and an outer coat continuous with the epicardium. The cyst communicated with the cavity of the left ventricle through endothelial-lined channels which were in direct continuity with the intertrabecular spaces of the ventricular cavity. Patterson and Spink30 considered that the primary defect was this system of endothelial channels in the myocardium which were probably remnants of the embryonic meshwork of endothelial-lined spaces that initially permeate the full thickness of the normally developing myocardium. These channels had reacted to the high pressure in the ventricle by forming an epicardial cyst, which subsequently ruptured.

A similar case, occurring in a 62-year-old white American male, has been reported by Lovitt and Lutz.31 In this instance a small “aneurysm” whose walls showed sclerotic changes was situated near the tip of the left ventricle. It is significant that the cavity communicated with the lumen of the left ventricle through an endothelial-lined channel.

No other abnormalities were associated with these congenital “diverticula” and “cysts.” These lesions therefore closely resemble apical and subvalvular aneurysms in anatomic position and histological structure.4, 5, 11 Indeed, apart from size and the presence of fibrosis and calcification, which are presumably related to the age of the lesions, the only differences between them seem to be those of nomenclature. The muscular tissue found in the subvalvular diverticulum described by Swyer and associates29 and the epicardial cyst described by Patterson and Spink30 might have been replaced by fibrous tissue had the infants survived long enough.

It appears to us, therefore, that congenital fibrous diverticula and epicardial cysts are basically the same as apical and subvalvular aneurysms. It is noteworthy that these lesions occur at the two areas where the ventricular wall is thinnest—the apical and subvalvular areas. A persistent endothelial-lined channel in these positions could then develop into
an aneurysm under the influence of the high pressure in the left ventricle. The posterior submitral area has been shown to retain these endothelial clefts even in full-term fetus. Presumably the stress of this pressure eventually converts the endothelial-lined sac into fibrous tissue so that only the ostium is lined by endocardium. If this similar etiology for apical and subvalvular aneurysm is accepted, the differences in their clinical manifestations would then be related to their anatomic positions, the subvalvular type usually having a more distinctive presentation because of their tendency to involve the mitral or aortic annuli or both and the coronary arteries.

Acknowledgment
We are indebted to Mr. W. Grundill and Mr. G. Katz for referring the patients to us. We wish to thank Dr. W. H. Kenny, Superintendent, Baragwanath Hospital, and Dr. H. Van Wyk, Superintendent, Johannesburg General Hospital, for permission to publish.

References

Circulation, Volume XXXV, June 1967
Atrial Fibrillation—Eighteen Centuries Ago

"Everybody knows what happened to Antipater who practiced medicine with great renown in Rome. As a man less than 60, but more than 50 years of age, Antipater suffered from a short fever of supposedly known cause. It happened that he felt his pulse after the fever's decline in order to know what to do about himself. Antipater was at first shocked when he found a complete irregularity of his arterial pulse. But since he was sure that he no longer had any fever he went to the bath because he felt fatigued from suffering and sleeplessness. He then submitted himself to a very light diet for three days. Since he had no more fever he devoted himself, as before, to his usual everyday duties. But he always checked his arterial pulse at the wrist and was astonished because of the persistence of the anomaly of the pulse.

"Antipater met me one day and stretched his arm out and, laughing, asked me to feel his pulse. Smiling in my turn I said: 'What is the riddle you want to solve?' Feeling his pulse I found a complete irregularity, not only in the order of the pulses which we call an irregularity of the sequence, but also one in the filling of the arteries. I was surprised that anyone could live with a pulse like that and asked him if he had any difficulties in breathing. He answered that he did not feel any difficulties. I observed him very frequently for six months to see if any change occurred, by feeling his radial pulse. When he asked me what the condition of his body ('diathesis' in Greek) was and how it could bring about such a pulse without fever I answered him that I had described in my 'Book on Pulses' a similar anomaly.

"I had assumed that this irregularity of the pulse came about by a narrow passage in the great pulmonary veins."*—RUDOLPH E. SIEGEL: Galen's Description of Mitral Stenosis: The Earliest Clinical Record of Rheumatic Heart Disease. Bull Cleveland Med Library 8: 16, 1961.

*The last sentence of this quotation can be better understood when we remember that the ancients considered the right atrium and auricle as a part of the vena cava, but the left atrium and auricle as part of the pulmonary vein.—Personal communication from R. E. Siegel.
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Circulation. 1967;35:1156-1162
doi: 10.1161/01.CIR.35.6.1156

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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