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Fatal Cases of Congenital Coronary Aneurysms in Young Persons

J. SIMONSEN

University Institute of Forensic Medicine, Department of Forensic Pathology, and University Institute of Pathological Anatomy, Copenhagen (Denmark)

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Summary. The literature of coronary aneurysms is reviewed. Such lesions are rare and amongst the approximately 100 cases published only 16 fulfil the criteria of being congenital in origin.

This paper deals with 3 cases of congenital coronary aneurysms in young persons — 33, 14 and 21 years of age — of which two showed multiple aneurysms. In all cases the cause of death was due to compromised coronary circulation due to the coronary aneurysm.

Differential diagnostic difficulties against arteriosclerotic aneurysms are elucidated. Low age in connection with histologic normal vessels, multiplicity and the presence of additional congenital malformations support the congenital etiology.

Zusammenfassung. Die Literatur über Coronaraneurysmen wird durchgearbeitet. Es werden hier gut 100 Fälle erwähnt, aber nur in 16 Fällen ist das Leiden als angeboren anzusehen.

Es wird über 3 Fälle von angeborenen Coronaraneurysmen bei jüngeren Personen (33, 14 und 21 Jahre alt) berichtet, von denen 2 multiple Aneurysmen aufweisen. In sämtlichen Fällen war der Tod auf erschwerten Coronarkreislauf zurückzuführen. Es wird betont, daß das Leiden außerordentlich selten vorkommt.

Die differentialdiagnostischen Schwierigkeiten den arteriosklerotischen Coronaraneurysmen gegenüber werden hervorgehoben. Junges Alter in Verbindung mit im übrigen histologisch normalen Gefäßen, Multiplizität sowie das Vorhandensein von anderen angeborenen Mißbildungen sind Faktoren, die für eine kongenitale Ätiologie sprechen.

Key words: Coronary Aneurysm, congenital — Coronary Aneurysm, arteriosclerotic — Coronary Aneurysm, mycotic-embolic — Coronary Aneurysm, dissecting — Coranary Aneurysm, Rupture of — Coronary Aneurysm, Occlusion of.

It is generally accepted that Morgagni was the first who briefly mentioned aneurysms of the coronary arteries in 1761. Bougon described in 1812 details of a 40 year old man who died suddenly from a haemopericardium due to a ruptured, calcified aneurysm of the right coronary. This patient also had a calcified aortic stenosis.

During the next century not much attention was paid to this disease. In 1929 Packard and Wechsler published 28 cases collected from the literature and added one themselves. They emphasized that the condition was often symptomless until it suddenly caused sudden death. The etiology was discussed intensely but the possibility of congenital origin was not mentioned. In the following years single cases were still published with the result that Scott in 1948 was able to collect from the literature 46 cases. He added one himself. The localization of the aneu-

rysms in the 27 cases was on the left coronary artery, in 11 on the right, in 3 cases the localization was unknown and in the remaining 6 cases there were multiple aneurysms. Approximately one third or exactly 15 cases were classified as congenital. 13 cases were mycotic-embolic, 6 arteriosclerotic and 6 syphilitic. 4 cases were not classified, 2 were part of generalized periarteritis nodosa and the last was interpreted as rheumatic.

Among the congenital aneurysms 14 were males compared to 1 female. Scott pointed out also that the cause of death before 1929 in the majority of cases was rupture of the aneurysms while the dominating lesion after 1929 was occlusion of the vessels.

In 1958 Munkner *et al.* reported a case diagnosed during life. This was a congenital coronary aneurysm in a 6 year old child diagnosed by coronary angiography. In this case which is believed to be the first diagnosed *in vivo* there was in addition an arteriovenous fistula.

Daoud *et al.* presented in 1963 10 cases of their own and added to their survey 79 more cases from the literature. In this total of 89 cases, there were 66 men and 23 women, a ratio of almost 3:1. Mycotic-embolic aneurysms were dominating in younger persons and arteriosclerotic in older persons while there was no age peak for the congenital aneurysms. The etiology of 52% of all these aneurysms was arteriosclerotic; 17% was congenital; 11% each was mycotic-embolic and dissecting and 4% was luetic. 5 case reports did not provide an etiologic diagnosis. 18 cases showed multiple aneurysms of which 12 were arteriosclerotic, 3 congenital and 1 mycotic-embolic. 2 case reports in this category did not provide an etiologic suggestion.

Cases among children are recorded by Forbes and Bradley (1960) who presented a 9 year old boy with a coronary aneurysm. The microscopical examination did not allow the authors to form an opinion concerning the etiology. In the first case presented by Bertelsen and Lindahl (1964) — an 8 year old girl — bilateral aneurysms developed on basis of periarteritis nodosa. The cause of death was haemopericardium due to rupture of the aneurysm in the left coronary artery. In the second case — an 81 year old man who died of uremia — the aneurysm was localized in the right coronary artery and had developed from arteriosclerosis.

In 1968 Sayegh *et al.* published a case which is believed to be number 100. It was a 43 year old man suffering from typical coronary attacks. Multiple coronary aneurysms were diagnosed by cinecoronary angiography. The case was believed to be the second diagnosed during life.

Recently Benson (1970) published a case of dissecting aneurysms in both the right and left coronary artery in a 62 year old man who died suddenly. In an extensive review of the literature the author found a total of 23 recorded cases of dissecting coronary aneurysms. Benson pointed out that preponderance of females (17 out of 24 cases) was worthy of note as was the fact that 7 of the females suffered this fatal disease in the postpartum period.

Reviewing the literature, a total of approximately 100 cases of coronary aneurysms have been reported, when not counting the dissecting aneurysms. Of these only 16 have been classified as congenital. In the present paper 3 cases of coronary aneurysms in young persons — 2 resulting in sudden death — all classified as congenital aneurysms are reported.

Case Reports

Case 1 (P. O. 18/52). This 33 year old woman suddenly fell unconsciuos in a streetcar while picking up a package from the floor. Dead on arrival at the hospital.

5 years earlier she had collapsed while running after a train. Shortly before death she had been complaining of shortness of breath when exercising and also had tendency to headache and dizziness.

Autopsy. Height: 168 cm. Weight: 57 kg.

There were a few superficial bruises in the face with underlying small haematomas (fall-lesions). The heart was of normal size (240 g). The coronaries were slightly arteriosclerotic only. 5 mm from the origin of the anterior descending branch from the left coronary artery a saccular aneurysm with a diameter of 10 mm was located. The coronary artery flew into the aneurysm on one side and continued from the exactly opposite side of the aneurysm. The thin wall of the aneurysm was calcified and the lumen was occupied by abundant thrombotic masses. On cut-section the myocardium appeared normal.

Except for the very slight arteriosclerotic changes in the aorta the autopsy-finding was negative. No microscopical examination.

Cause of Death. Myocardial ischaemia due to thrombotic occlusion of a congenital coronary aneurysm.

Case 2 (Sect. 457/66). 14 year old boy who 4 years previously had suffered from febrile attack in connection with throatsymptoms and aches in the fingerjoints. Shortly after he developed severe abdominal pains and laparotomy revealed a bleeding aneurysm on the hepatic artery. The aneurysm was removed and the artery ligated. Microscopy of the wall of the aneurysm showed slight calcification but no indication of inflammation.

Half a year later cardial symptoms occurred with dyspnea and chestpains. X-ray showed an enlarged heart. Congestion of the liver was obvious and he was treated with digitalis and diuretic drugs. Cardiac catheterization revealed insufficiency of the right as well as the left ventricle but no abnormal anastomosis. The condition was unaltered until he was hospitalized in a terminal stage which resulted in death 8 weeks after admission.

Autopsy. Height: 159 cm. Weight: 47 kg.

The heart tremendously enlarged (630 g). The mitral valves were covered with small vegetations. The chordae tendineae were normal but in the anterior wall of the left ventricle a big fibrous scar was present. 5 mm from the origin of the left coronary artery was a thinwalled aneurysm located with a diameter of approximately 13 mm. The wall was calcified but there was no thrombus in the lumen. Just peripheral to this aneurysm a similar aneurysm was located but contrary this was completely occupied by old thrombus masses. The wall of this aneurysm was calcified. From the aneurysms first mentioned a wide and normal looking

Fig. 1 (case 2). The heart was opened and the view is from the inside of the aortic ostia. The aneurysm is located between the arrows. The origin of the left coronary artery (*a*) and the circumflex branch (*b*)

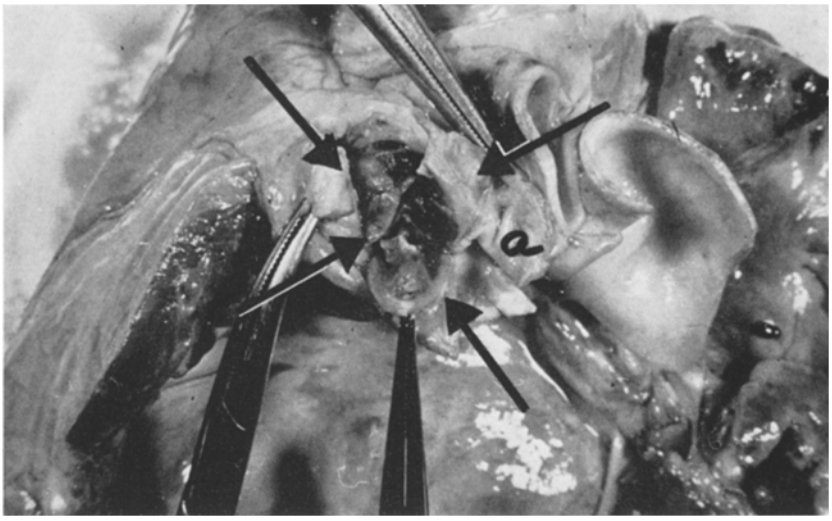

Fig. 2. Same case as Fig. 1. Viewed from outside. The second aneurysm occluded by thrombus masses surrounded by arrows. The first mentioned aneurysm can barely be seen (*a*)

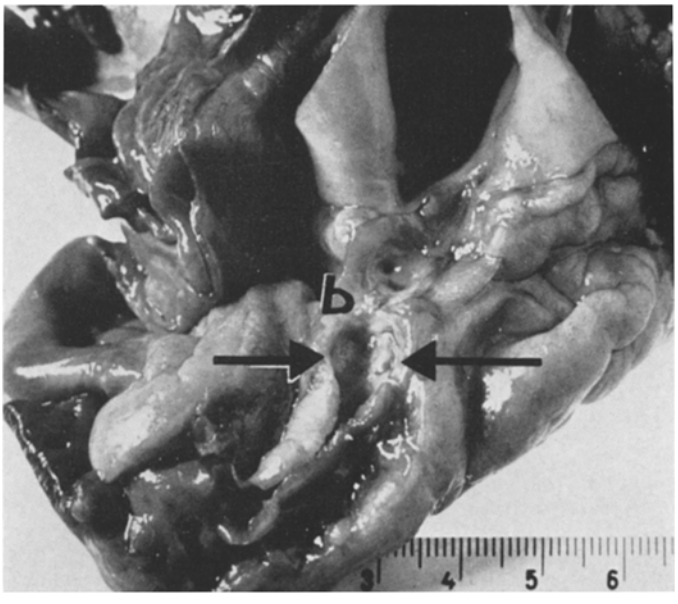

Fig. 3 (case 3). Aneurysm in the right coronary artery (arrows). The right coronary ostia (*a*). The artery is moderately arteriosclerotic and stenotic (*b*)

circumflex branch originated. From the occluded aneurysm it is believed that the descending branch has radiated but it was impossible to demonstrate this artery. The big scar corresponded exactly to the area which received blood-supply from this artery. The right coronary artery was normal.

Additional findings were only sequaelae after ligation of the hepatic artery and removal of the gallbladder surgically.

Microscopy: The aneurysmatic walls showed severe atheromatosis with calcification. The scar in the myocardium was verified. The valvular vegetations did not reveal a characteristic histological picture and no evidence of rheumatic lesions was found. Numerous sections from the vascular system showed normal pictures.

Discussion: The aneurysms were regarded as congenital. The occlusion of the peripheral aneurysm probably occurred 3 years previously at the time when the symptoms of incompensation became evident — due to the big infarction in the anterior wall of the left ventricle.

Cause of Death. Cardial incompensation due to myocardial fibrosis as a result of thrombotic occlusion of a congenital coronary aneurysm.

Case 3 (D 306/70). 21 year old draftee collapsed during exercise. Resuscitation was immediately started, he was dead on arrival at the hospital.

There was no medical history and he had done rather well in the military service. He had been satisfactory in physical exercises.

Autopsy. Height: 182 cm. Weight: 66 kg.

There were two small excortiations in the left side of the face (fall-lesions).

The heart was of normal size (350 g). The coronary arteries were moderately atheromatous. 3 cm from the origin of the right coronary was a marked stenosis immediately followed by an aneurysm with a diameter of 7 mm. In the left coronary artery, 6 mm from the origin were two aneurysms located. The first had a diameter of 15 mm, the second of 12 mm. From the last mentioned aneurysm a very delicate artery branch originated, probably the hypoplastic circumflex branch. The anterior descending branch was not demonstrable. The aneurysmatic walls were all calcified. In the myocardium, especially in the septal part, slight interstitial fibrosis was observed.

In addition congenital malformations were demonstrated in the genito-urinary tract. The right kidney was absent and there was hypospadia glandis.

Serumcholesterollevel in post mortem blood was 145 mg%.

Microscopy: The aneurysmatic walls showed evidence of atheromatosis with calcium deposits. The myocardium showed slight interstitial fibrosis while several sections from the vascular system revealed normal structure.

Cause of Death. Coronary insufficiency due to multiple congenital aneurysms in a person with additional congenital malformations.

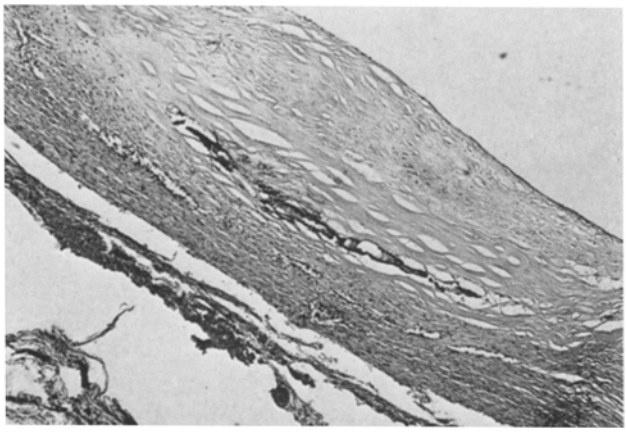

Fig. 4. Same case as Fig. 3. Wall of the aneurysm with deposits of calcium, haematoxilin-eosin × 56

Discussion

It appears that coronary aneurysms are observed more often now than in the past. Whether the reason for this is due to an increase in the literature on the topic or due to an alteration in the criterias of evaluation is difficult to determine. Probably both factors are contributing to this.

Daoud *et al.* (1963) pointed out that cases with coronary aneurysms in their material were as frequent as 1.4% of the total number of autopsies in adult persons. In 694 hearts from persons more than 16 years of age they were able to find 10 cases of coronary aneurysms including 3 with multiple aneurysms. The examinations were very extensive and the criteria of selection were very liberal. The coronaries were cross-sectioned at intervals of 5 mm and even very small changes in the diameter were recorded. All the aneurysms were arteriosclerotic in origin and found in elderly persons (51—86 years); a completely different type than those in the present material. It is questionable if other investigators would adopt criteria as liberal as those mentioned. Characteristic for arteriosclerotic aneurysms is the more fusiform shape. The authors asserted that aneurysms of the abdominal aorta is a frequent combination.

Coronary aneurysms are not frequent (Packard and Wechsler, 1929), and this statement is more true when congenital coronary aneurysms are concerned (Scott, 1948; Sayegh *et al.*, 1968). Among approximately 100 cases mentioned in the literature only 16 are regarded as congenital.

The 3 present cases which are regarded as congenital in origin are found among more than 20000 autopsies, which means that the frequency is less than 0.015% in routine, but very carefully performed autopsies.

From the literature it is evident that congenital and arteriosclerotic aneurysms are still more dominating among the reported cases, as it is also evident that the cause of death due to occlusion of the abnormal vessel is now the most common finding as compared to the previous time, when rupture resulting in haemopericardium was the dominating cause of death. It is interresting that Packard and Wechsler in their extensive survey in 1929 did not mention the possibility of a congenital etiology for these cases. The above mentioned alteration in type and symptoms is not surprising when taking in to consideration that mycotic-embolic complications as well as syphilis have become more and more seldom.

The problem at the present time, for this reason is more and more limited to the discussion whether a demonstrated aneurysm is of arteriosclerotic or congenital origin. In both cases the wall of the aneurysm is often calcified. The shape may be of some help because the arteriosclerotic aneurysms are more often fusiform while the congenital aneurysms commonly are saccular. Young age and additional congenital malformations, especially of the vascular system (Crocker *et al.*, 1957) support very strongly the possibility of congenital origin. In addition, if the vascular system including the coronaries shows normal appearance or very slight atheromatous changes this is strong support for a congenital etiology.

In the 3 present cases there were almost no pathological findings in the coronaries except for the aneurysms, which all were saccular in shape and in 2 cases multiple, all the persons were young and finally 2 of the cases showed concomitant

congenital malformations. For these reasons the author finds it justified to classify all 3 cases as congenital aneurysms of the coronary arteries.

As mentioned above, the etiology seems to change during the time, but other alterations are worthy of mention. It becomes still more common to diagnose the abnormality during life by coronary angiography and lately it looks like a new type is coming up, the dissecting coronary aneurysm which is far more common among women than men, and it is also worthy to note that especially women in the post partum period seem to be predisposed to this fatal disease.

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Prof. Dr. med. Jørn Simonsen
Retsmedicinsk institut
J. B. Winsløwsvej 15
DK-5000 Odense
Denmark