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CASE REPORT

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Fibromuscular dysplasia of coronary arteries as a rare cause of death

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Abstract A case of fibromuscular dysplasia of the coronary arteries in a 15-year-old boy is reported. After a quarrel involving no violence the boy suddenly suffered from ventricular fibrillation, collapsed and was initially successfully defibrillated. After 37 days of deep unconsciousness the boy died of bronchopneumonia. The cause of the ventricular fibrillation was clarified only after histological investigations. Fibromuscular dysplasia of the coronary arteries with narrowing was found, which has very occasionally been described in the literature. However, its localization in the A-V node artery, as described here, only seems to have been observed once.

Key words Fibromuscular dysplasia · Coronary artery · A-V node · Cardiac death

Introduction

In 1938 Leadbetter and Burkland [8] described a disease of the renal arteries, which in 1958 was named fibromuscular hyperplasia by McCormack et al. [13] and nowadays is known as fibromuscular dysplasia of arteries [4]. Fibromuscular dysplasia is most often found in the renal and cervical arteries. The aorta and other arteries are rarely affected [9], but in this case fibromuscular dysplasia of coronary arteries is reported.

Case report

After having a quarrel with friends a 15-year-old boy suddenly slipped off an armchair and collapsed. No violence was involved. After 8 min of unconsciousness the ambulance arrived and the electrocardiograph revealed ventricular fibrillation. Defibrillation was successful, and while the boy remained deeply unconscious, intubation and artificial respiration were carried out before he was admitted to hospital. The computer tomogram showed no signs of any intracranial disease. Poisoning was excluded by a toxicological analysis. During the boy's stay in hospital, electrocardiographic records revealed no pathologic changes. The main symptom was the deep irreversible unconsciousness, which was considered to be a consequence of cerebral ischaemia caused by ventricular fibrillation of unclear genesis. The boy died of bronchopneumonia 37 days after the sudden event.

Autopsy findings

The autopsy findings included purulent tracheobronchitis and bronchopneumonia and signs of general hypoxaemic damage to organs. The heart weight was 180 g with no congenital malformations and no acquired valvular heart defect. Coronary arteries were macroscopically inconspicuous. The histological investigations of the organs revealed an abnormal, extreme narrowing of the A-V node artery with distinct disturbances of the framework of the arterial wall in the heart. In these areas of the vessel, a proliferation of smooth muscle cells with fibrosis of the media and partly of the intima were found. The internal elastic lamina was partly fragmented and partly doubled in thickness (Figs. 1–3). The immunohistochemical investigations showed evidence of actin in the media and intima, whereas the reaction to desmin was only focal. Similar changes, but less intense, were found in further small branches of the coronary arteries. Arteriosclerosis, amyloidosis or myocarditis with the corresponding affection of vessels did not exist. The A-V node and the bundle of His showed no signs of necrosis or increased fibrosis. The histological investigations of further organs proved that there were no significant changes in the arterial walls. Hypoxaemic damage of ganglion cells was found in the brain. The diagnosis was fibromuscular dysplasia of the A-V node artery and other small branches of coronary arteries.

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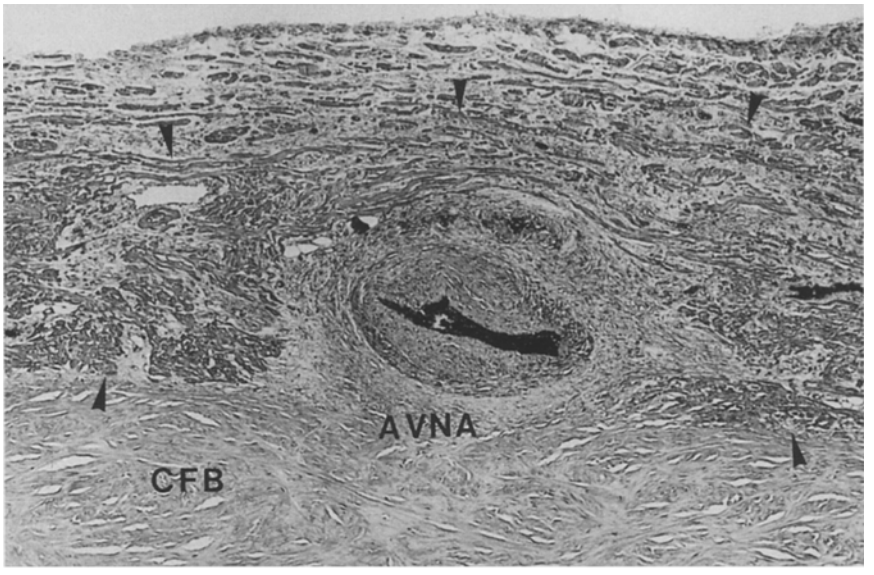

Fig. 1 The A-V node artery (*AVNA*) adjacent to the central fibrous body (*CFB*) of the heart. Strong thickening of the arterial wall and narrowing of the lumen. The A-V node is marked by *arrows*. Masson-Goldner, × 64

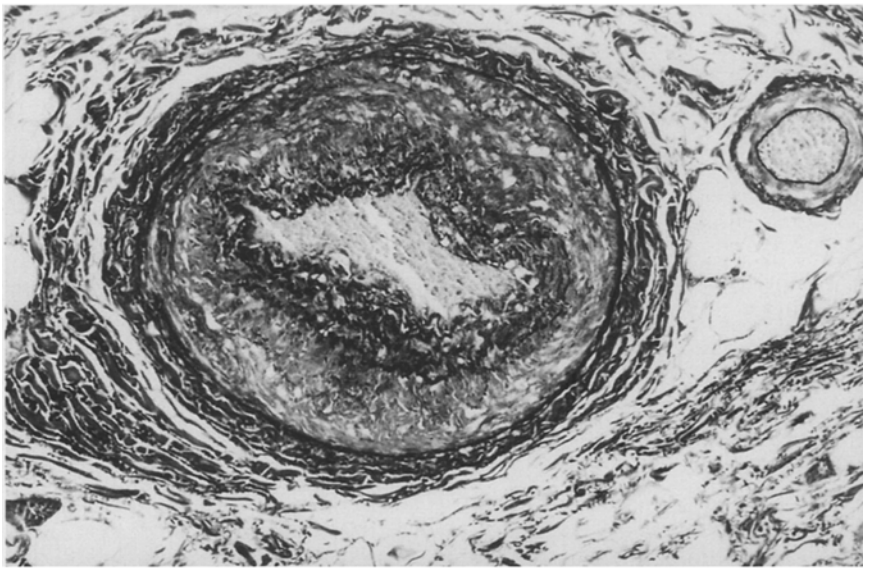

Fig. 2 The same artery shortly before it enters the A-V node. Considerably narrowed lumen. Destruction of the internal elastica lamina in all parts. Additionally a small branch of the A-V node artery without any pathological changes is seen. Elastica-Domagk, × 128

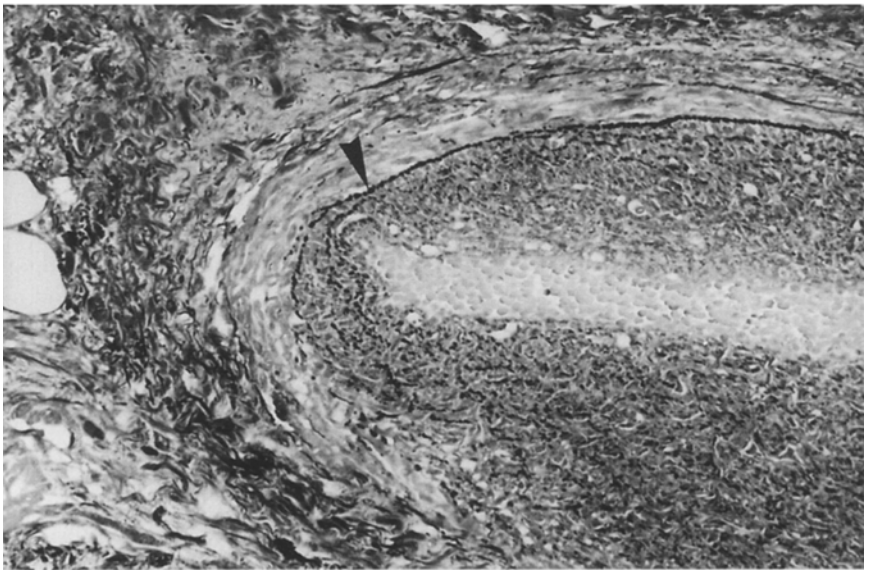

Fig. 3 Destruction of the internal elastic lamina. The arrow marks the change-over from normal to altered internal elastic lamina. Elastica-Domagk, × 256

Table 1 Cases of fibromuscular dysplasia of coronary arteries recorded in the literature

No.	Reference	Year	No. of cases	Age (years)	Sex	Localization
1	Brill et al. [1]	1971	1	18	f	Coronary artery
2	James and Marshall [5]	1976	2	7 64	m m	Sinus node artery Sinus node artery
3	Lie [10]	1983	1	34	f	Left ant. desc. coronary artery
4	Lie and Berg [11]	1987	1	24	m	Right coronary artery
5	Schneider et al. [16]	1987	1	56	m	Right coronary artery
6	Nichols et al. [14]	1989	1	28	f	Sinus node artery
7	Ogbuihi [15]	1989	1	0.8	m	Sinus node artery
8	James and Riddick [6]	1990	1	29	m	Branch of AV node artery supplying the His bundle
9	Lüders [12]	1990	1	25	m	Small branches of coronary arteries
10	Kanzaki et al. [7]	1992	1	16	m	Coronary arteries
11	Yamada et al. [18]	1993	1	37	m	Coronary arteries
12	Tanaka et al. [17]	1993	1	17	m	Left and right coronary artery

Discussion

Fibromuscular dysplasia is a non-atheromatous, non-inflammatory disease of the arteries with segmental stenosis, but its aetiology has not yet been clarified. The disease is characterized by a fibrous or fibromuscular thickening of the vessel wall with varying degrees of affection of the intima, media and/or adventitia and also by destruction of elastic elements of the vessel wall. Although fibromuscular dysplasia mainly affects the renal and cervical arteries, it is occasionally also found in other vessels. With an incidence of 1% in renal arteries from unselected autopsies and approximately 0.5% in cervical angiographies, fibromuscular dysplasia is a rare disease. Approximately 5 times as many women as men and all age groups, including the paediatric age group, are affected [9].

Fibromuscular dysplasia of coronary arteries has only very rarely been described. In the literature we found a total of 13 cases, which are summarized in Table 1. In contrast to fibromuscular dysplasia in other localizations, men outnumber women by 3 to 1 as far as localization in the coronary arteries is concerned. Owing to the small number of cases, this distinction between the sexes in the literature should not be accorded too much importance. A uniform histological classification of fibromuscular dysplasia of arteries does not exist. In 1971 Harrison and McCormack introduced a classification based on findings in renal arteries, which was later applied to arteries in other locations [2]. This classification distinguishes between intimal, medial and adventitial types. The present report concerns a medial type with the described focal, multifocal or tubular stenoses [2, 3]. Such disturbances in the framework of the arterial wall (Figs. 1–3) were found at all levels in the series of sections ($n = 85$). In some places they were limited to opposite sides of the vessel, but in others they were found around the whole circumference. The proliferation of smooth muscle cells with accumulations of collagenous connective tissue were found to be concentric in the tubular form. In the focal and multifocal forms, the stenoses were predominantly excentric and not circular [2, 9]. The varying degrees to which these findings were present are shown in the Figs. 1–3.

In several cases fibromuscular dysplasia of coronary arteries has been described as a cause of sudden cardiac death [1, 5, 6, 10–12]. In the case presented here, we consider the narrowing of the A-V node artery consequent on fibromuscular dysplasia to be the cause of the sudden ventricular fibrillation of the heart, which led to irreversible ischaemic cerebral damage.

References

1. Brill IC, Brodeur MTH, Oyama A (1971) Myocardial infarction in two sisters less than 20 years old. JAMA 217: 1345–1348
2. Harrison EG, McCormack LJ (1971) Pathologic classification of renal arterial disease in renal vascular hypertension. Mayo Clin Proc 46: 161–167
3. Harrison EG, Hunt JC, Bernatz PE (1967) Morphology of fibromuscular dysplasia of the renal artery in renovascular hypertension. Am J Med 43: 97–112
4. Hunt JC, Harrison EG, Sheps SG, Bernatz PE, Davis GD (1965) Hypertension caused by fibromuscular dysplasia of renal arteries. Postgrad Med 38: 53–58
5. James TN, Marshall TK (1976) De subitaneis mortibus. XVII. Multifocal stenoses due to fibromuscular dysplasia of the sinus node artery. Circulation 53: 736–742

6. James TN, Riddick L (1990) Sudden death due to isolated acute infarction of the His bundle. J Am Coll Cardiol 15: 1183–1187
7. Kanzaki T, Kobayashi T, Shimzu H, Miura Y, Takayasu M, Abe T (1992) Aneurysm in the skin: arterial fibromuscular dysplasia. J Am Acad Dermatol 27: 883–885
8. Leadbetter WF, Burkland CE (1938) Hypertension in unilateral renal disease. J Urol 39: 611–615
9. Letsch R, Kantartzis M, Sommer T, Garcia M (1980) Arterial fibromuscular dysplasia. Report of a case with involvement of the aorta and review of the literature. Thorac Cardiovasc Surg 28: 206–210
10. Lie JT (1983) The cardiovascular system in sudden death. In: Silver MD (ed) Cardiovascular pathology, vol 2. Churchill Livingstone, New York London Melbourne, pp 1191–1192
11. Lie JT, Berg KK (1987) Isolated fibromuscular dysplasia of the coronary arteries with spontaneous dissection and myocardial infarction. Hum Pathol 18: 654–656
12. Lüders H (1990) Plötzlicher Herztod bei fibromuskulärer Dysplasie der Koronararterien. Zentralbl Allg Pathol Pathol Anat 136: 711–714
13. McCormack LJ, Hazard JB, Poutasse EF (1958) Obstructive lesions of renal artery associated with remediable hypertension. Am J Pathol 34: 582–586
14. Nichols GR, Davies GJ, Lefkowitz JB (1989) Sudden death due to fibromuscular dysplasia of the sinoatrial nodal artery. J Ky Med Assoc 87: 504–505
15. Ogbuihi S (1989) Zum forensischen Stellenwert von Läsionen des Sinusknotens bei unklaren plötzlichen Todesfällen. Z Rechtsmed 102: 315–322
16. Schneider J, Coester C, Meyer W, Oechslin E (1987) Ungewöhnliche Koronartodesfälle. Schweiz Med Wochenschr 117: 1577–1582
17. Tanaka M, Watanabe T, Tamaki S, Ichihara T, Yasushi T, Abe T, Masakazu T, Nakashima N (1993) Revascularization in fibromuscular dysplasia of the coronary arteries. Am Heart J 125: 1167–1170
18. Yamada A, Nakamoto S, Sakamoto M, Matsumoto T (1993) An autopsy case developing both marked stenosis of the coronary artery and dilated phase of hypertrophic cardiomyopathy. Respir Circ 41: 689–692