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Case reports

Giant coronary aneurysm associated with aortic mucoid medionecrosis

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Summary. A case of sudden death due to rupture of a dissecting aneurysm of the ascending aorta in a 38-year-old man is presented. The patient had a clinical history of severe hypertension. The autopsy also revealed the presence of a voluminous aneurysm of the right coronary artery and a solitary multilocular cyst of the right kidney. It is thought that a prodromal influenza-like syndrome and the renal lesion could have played a role in causing the vascular pathology.

Key-words: Sudden death – Coronary aneurysm – Dissecting aortic aneurysm

Zusammenfassung. Ein 38 Jahre alter Mann starb plötzlich aufgrund der Ruptur eines dissezierenden Aneurysma der Aorta ascendens. Er hatte eine Anamnese mit einem schweren Hochdruck. Bei der Autopsie fand sich auch ein voluminöses Aneurysma der rechten Koronararterie und eine vereinzelte multilokuläre Zyste der rechten Niere. Die Autoren denken, daß die Läsion der Niere eine kausale Rolle bei der Ätiologie der Gefäßerkrankung spielen könnte.

Schlüsselwörter: Plötzlicher Tod – Koronaraneurysma – Dissezierendes Aortenaneurysma

Introduction

Aneurysms of the coronary arteries are lesions that are relatively rarely found [1–3] and may cause sudden death due to thrombosis or rupture [4]. Solitary multilocular cysts in the kidney are also rare lesions [5–8]. In the present report a case of sudden death due to the rupture of a dissecting aortic aneurysm in a 38-year-old hypertensive man is described. The autopsy also revealed the presence of a right coronary aneurysm and a solitary multilocular cyst of the kidney.

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Case report

In November 1987, a 38-year-old man was found dead sitting at the steering wheel of his car. The anamnesis revealed that an essential hypertension (220/150) had been diagnosed in 1981. In 1985, the patient was hospitalized with influenza complicated by bronchopneumonia. The echography, pyelography, angiophotoscintigraphy and laboratory investigations led to the general diagnosis of "chronic nephropathy with renal insufficiency". The patient was not diabetic nor syphilitic and responded well to the anti-hypertensive therapy.

Postmortem examination

The corpse was that of a robust man with no external signs of violence or trauma. The pericardium was filled with partially coagulated blood. The ascending aorta was eroded and showed a 19 mm long transverse breach of the adventitia which communicated with a 3 cm long vertical rupture of the intima beginning from the free edge of the right cusp of the aortic valve. A dissection of the aortic wall reaching the origin of the innominate artery was present. The heart weighed 390 g and showed marked hypertrophy of the left ventricle. The coronary arteries were pronounced, due to a diffuse enlargement of the calibre and whitish colour of the walls. After rounding the acute margin, the right coronary artery presented a 5×2 cm fusiform dilatation (Fig. 1). There was no arterio-venous fistula nor fistulous communication with the right venous chambers of the heart. The coronary arteries were removed together with the bulbus aorticus (Fig. 2) and an X-ray examination showed thin calcifications, mainly in the wall of the aneurysm (Fig. 4). The kidneys were reduced in size and their surface was finely granular. The upper pole of the right kidney showed a whitish mass 4 cm wide. The cut surface was multilocular with cysts which did not communicate with the renal pelvis. The abdominal aorta showed diffuse atherosclerosis. Histological examination showed marked intimal thickening and accumulation of mucoid material separating and disrupting the elastic fibres of the ascending aorta

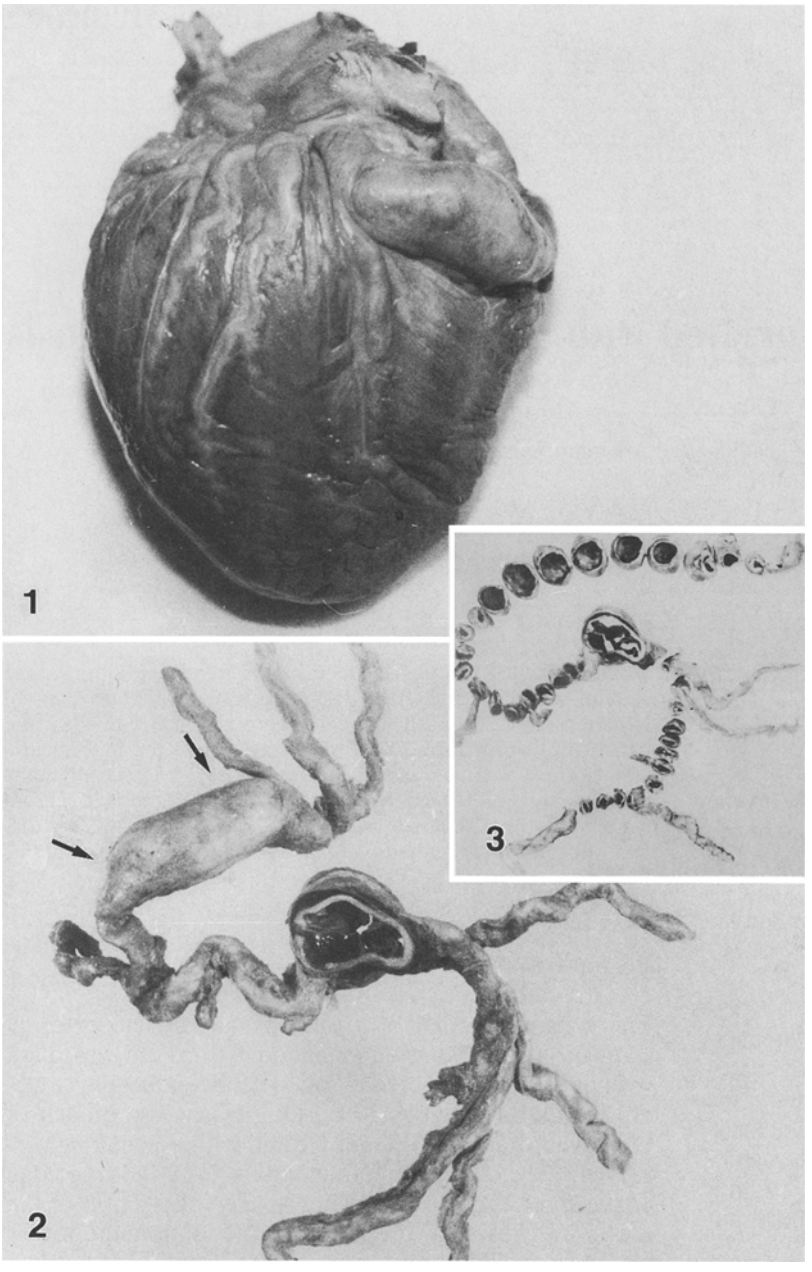

Fig. 1. Diaphragmatic aspect of the heart. Voluminous fusiform aneurysm of the right coronary artery

Fig. 2. The coronary arteries removed together with the bulbus aorticus. The *arrows* indicate the aneurysm

Fig. 3. The coronary arteries cross-sectioned at intervals of 5 mm

media (Fig. 7). The coronary arteries, cross-sectioned at intervals of 5 mm (Fig. 3), showed diffuse fibrosis and a slight lymphoplasmacytic infiltrate of the wall. The aneurysm (transverse diameters 14×20 mm) was almost completely thrombosed, with partial recanalisation and showed a 4 mm large residual lumen (Fig. 5). The wall of the aneurysm was 2 mm thick and there was an almost complete absence of smooth-muscle cells with fibrosis and calcium deposition in the media. The adventitia presented a lymphoplasmacytic inflammatory infiltrate containing germinal centres and intimal proliferation of the vasa vasorum. The kidneys showed diffuse glomerulosclerosis. The mass of the right kidney was composed by dilated tubules, many dilated to cystic proportions, lined by flattened or occasionally hobnailed cells (Fig. 8). Scattered throughout the stroma there were clusters of small tubules lined with clear cuboidal cells. The diagnosis of "hemopericardium due to the rupture of a dissecting aneurysm of the ascending aorta. Coronary arteritis with voluminous aneurysm of the right coronary artery. Solitary multilocular cyst of the right kidney.", was made. Marfan's syndrome was absent.

Discussion

The case studied presents an unusual association of a coronary aneurysm with a solitary multilocular cyst of the kidney in a young hypertensive man who died due to rupture of the aorta. Aortic ruptures are mainly posttraumatic or caused by dissecting aneurysms. Car accidents causing thoracic contusion are most often the cause

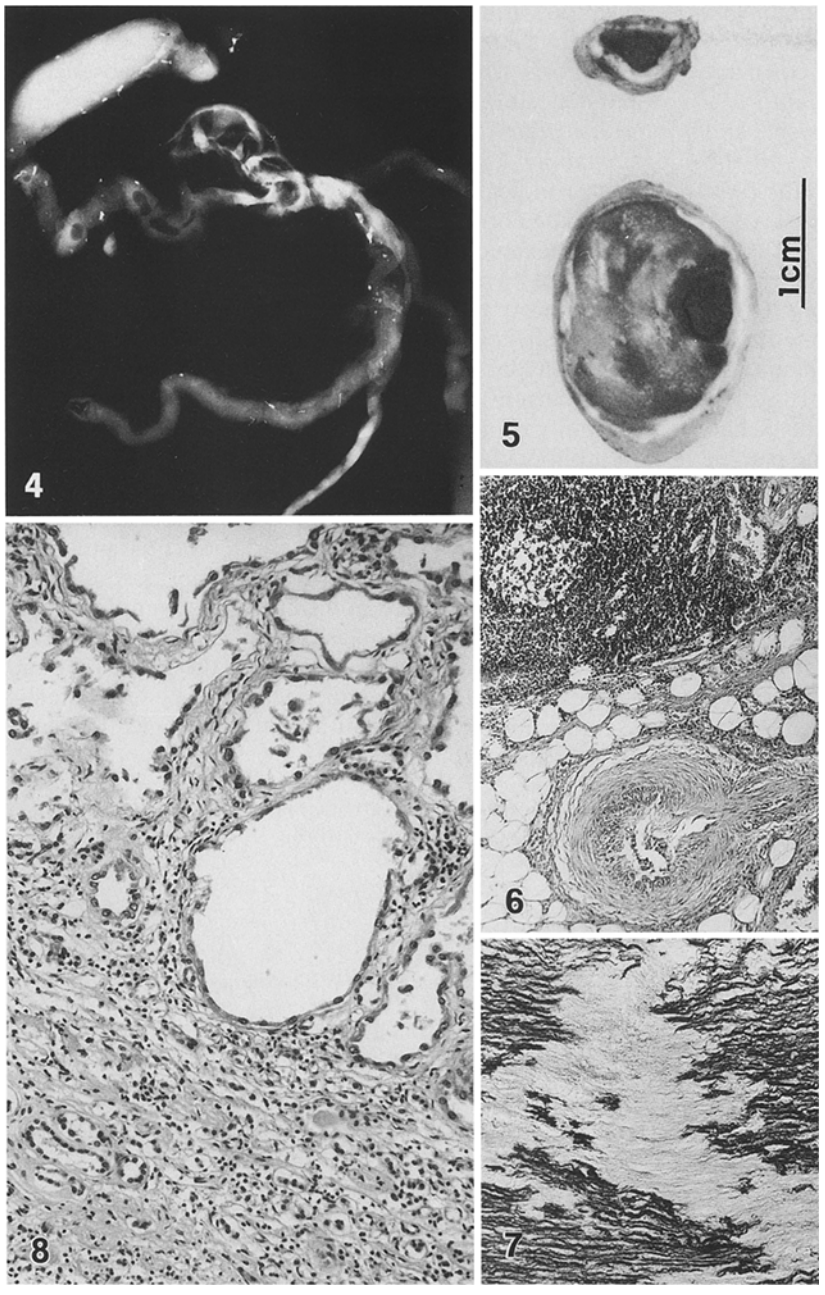

Fig. 4. The X-ray examination shows thin calcifications, mainly in the aneurysm wall

Fig. 5. Confrontation between the transverse section of the aneurysm and that of the left coronary artery origin. The aneurysm is nearly completely thrombosed

Fig. 6. Lymphoplasmacytic inflammatory infiltrate containing germinal centers and intimal proliferation in the vasa vasorum in the adventitia of the aneurysm wall. H-E $\times 6.3$

Fig. 7. Section of the ascending aorta media showing accumulation of mucoid material separating and disrupting the elastic fibres. Elastic stain $\times 10$

Fig. 8. The solitary multilocular cyst of the kidney shows dilated tubules lined by flattened or occasionally hobnailed cells. H-E $\times 25$

of traumatic aortic rupture which may occur immediately or be delayed (two-staged) [9, 10]. The main factor in the pathogenesis of dissecting aneurysms is the association with a degenerative lesion involving the media (i.e. mucoid medionecrosis). This condition was first recognized in 1928 by Gsell [11], while Erdheim [12] in 1930 described it in detail and coined the name "medionecrosis aortae idiopathica cystica". The lesion occurs frequently in Marfan's syndrome in which there is an inherited defect of mesodermal tissues. In this case the histological examination of the ascending aorta wall, showing lesions in the tunica media such as an accumulation of mucoid material separating and disrupting the elastic fibres, leads to the diagnosis of dissecting aortic aneurysm due to mucoid medionecrosis. More interesting is the interpretation of the etiology of the giant coronary aneurysm. Mycotic-embolic aneurysms are dominant in younger persons and arteriosclerotic aneurysms in older persons [13, 14]. More recently it has been reported that in young people inflammatory [3] and congenital [4] aneurysms are the more frequent types, even if there is no age peak for the latter. The most frequent inflammatory aneurysms in children and young adults are secondary to the mucocutaneous lymph node syndrome [15, 16], or Kawasaki's disease which was first described in 1967 in Japan [17] and then in many other countries [18, 19]. Congenital aneurysms are often associated with other visceral malformations [20] and they may have secondary arteriosclerotic lesions such as calcifications in the media and inflammatory infiltrate in the adventitia. Simonsen [4] proposed the following diagnostic criteria for congenital aneurysms: low age in connection with histo-

logically normal vessels, multiplicity of the aneurysms and presence of additional congenital malformations. Virmani et al. [3] reported 52 cases of acquired coronary arterial aneurysms. The patients were divided into two groups: 38 with atherosclerotic coronary aneurysms and 14 with aneurysms secondary to inflammation. In the atherosclerotic group, aneurysms were found in the right coronary artery (18 cases), in the left coronary artery (13 cases) and in the right and left coronary arteries (7 cases). Aortic aneurysms were present in 8 of these patients and in 6 cases they had ruptured. A history of systemic hypertension was found in 52% of the patients. None of the patients had ruptured coronary arterial aneurysms. In the inflammatory group, isolated aneurysms of the left coronary artery were seen in 6 patients, while 8 had multiple right and left coronary aneurysms. One patient had an aortic aneurysm. Two of the coronary arterial aneurysms had ruptured. In the case described here it was necessary to consider the possible relation between the coronary aneurysm and the aortic pathology. Extension of the aortic dissection to the coronary artery or the contemporary compromission of the coronary wall by the mucoid medionecrosis were excluded after histological examination. The young age and the "malformative" renal lesion could have induced a congenital origin of the aneurysm. On the other hand the chronic hypertension, the diffusion of the arteriosclerotic lesions to the whole arterial system and the fusiform shape [14] of the aneurysm would tend towards an acquired origin. Therefore in the light of two anamnestic data, the hypertension dating from 1981 and the influenza and of an autoptic element i.e. the solitary multilocular cyst of the kidney, the etiology of the coronary aneurysm must be considered. The hypertension itself, with the related systemic arteriosclerotic lesions, could have been sufficient to cause the aneurysm. The influenza in turn could have caused a coronary arteritis leading to a reduction in resistance of the vessel wall. The solitary multilocular cyst of the kidney is a benign renal tumor that can sometimes cause hypertension [7]. Virmani et al. [3] classified acquired coronary aneurysms on the basis of the following criteria. Atherosclerotic aneurysms were characterized by lipid deposition; foam cells; pultaceous debris containing cholesterol clefts, with or without hemorrhage; fibrous caps; hyalinization; focal calcification, usually in the intima, with or without extension in the media; extensive destruction of musculo-elastic elements of the media and often lymphocytic aggregates in the adventitia, usually around the vasa vasorum and/or mild lymphocytic infiltrates within atherosclerotic plaques. Inflammatory aneurysms on the other hand, were characterized either by the presence of inflammatory infiltrates in the media and adventitia or by minimal inflammation, with or without medial fibrinoid necrosis. In some quiescent inflammatory aneurysms the only change was focal destruction of the media, with replacement by fibrous tissue. Thrombi were sometimes present in the lumen; the thrombi showed varying degrees of organization, with granular tissue containing small multiple endothelium-lined capillaries. In our case the histological examination revealed alterations in the coronary wall (slight intimal thickening, fibrosis and calcifications in the media, lymphoplasmacytic infiltrate in the adventitia, intimal proliferation of the vasa vasorum, thrombosis with partial recanalisation and absence of lipid deposition and of pultaceous debris containing cholesterol clefts) which in combination with the history of the patient (young age, prodromal influenza-like syndrome) indicate a previous general inflammatory process of the coronary arteries and the inflammatory etiology of the aneurysm. The chronic pre-existent hypertension might have strengthened the damaging action of the inflammatory process of the coronary wall. The absence of family history and the protracted positive response to the anti-hypertensive therapy would lead to doubt that the hypertension really was primitive. Therefore the possibility of nephrogenic hypertension due to the solitary multilocular cyst of the kidney, as it has been sometimes reported in the literature [7], must be considered.

A final consideration regards the association of inflammatory aneurysms of the aorta, aortitis and coronary arteritis occasionally reported in literature [21, 22]. In our case the absence of diffuse peri-adventitial fibrosis with intense lymphoplasmacytic infiltrate, the hallmark of inflammatory aneurysms of the aorta [23] and the presence of mucoid medionecrosis exclude a systemic vasculitis and indicate a different pathological etiology of aortic and coronary arteries.

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