Virchows Arch [Pathol Anat] (1983) 402:73–82

Virchows Archiv A

Case report

Spontaneous dissecting aneurysm of the renal arteries

A case and a review of the literature

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Summary. A case of dissecting aneurysm of the renal arteries is presented. The patient suffered from an intractable subarachnoid bleeding and the kidneys had been selected for transplantation. One kidney was never transplanted, the other was transplanted and rejected after few days. Dissecting aneurysms were present in the main artery and its major ramifications in both kidneys. Many investigators have claimed that dissecting aneurysm and fibromuscular dysplasia of the renal artery are different stages of but one disease. A review of the accumulated literature on dissecting aneurysm of the renal artery reveals, however, that this disorder shows a preponderance of middle-aged men, whereas fibromuscular dysplasia of the renal artery affects adolescent girls.

It is concluded that the two disorders of the renal artery most likely represent different vascular diseases.

Key words: Dissecting aneurysm – Fibromuscular dysplasia – Renal artery – Kidney transplantation

Case report

The patient was a 52-year-old woman. She had had two normal deliveries (1953 and 1955). In 1958 she had been admitted to hospital because of left peripheral paresis of the facial nerve and again in 1973 due to pains in the right side of the face. These symptoms were thought to be of myogenic origin and responded to physiotherapy. Blood pressure at that time was 160/95 mm Hg and serum creatinine 0.9 mg/100 ml.

One night in November 1980 she experienced sudden and severe pain in the neck and became unconscious after one hour, only reacting to painful stimuli. The blood pressure was 205/100 mg Hg and the serum creatinine 0.9 mg/100 ml on admission.

Acute CT-scanning showed subarachnoid haemorrhage and an aneurysm of the left pericalosal artery. Similar findings were seen in a cerebral angiogram and a slightly uneven diameter of all cerebral arteries was also noted (Fig. 1).

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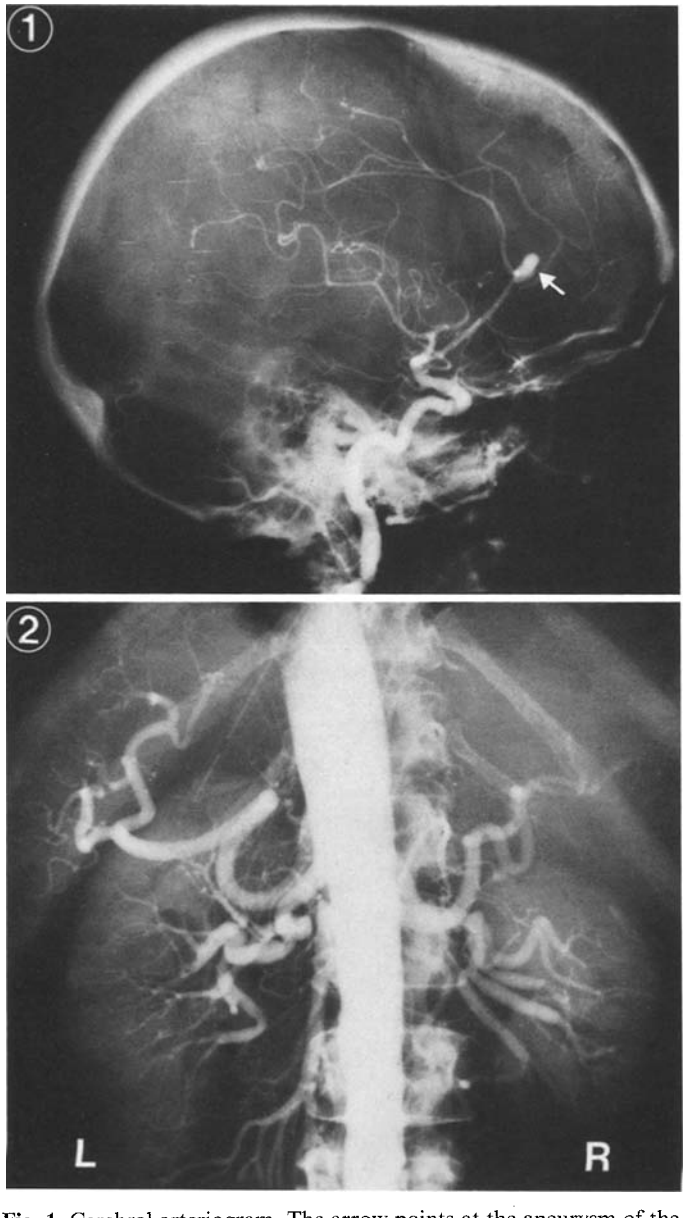

Fig. 1. Cerebral arteriogram. The arrow points at the aneurysm of the left pericalosal artery

Fig. 2. Renal arteriogram. The dilatation of the extrarenal parts of the right and left renal artery is prominent. *L:* left, *R:* right

Although procedures to relieve the increased intracranial pressure were carried out, isoelectrical EEG developed and pan-arteriography showed cerebral swelling. With the intention of utilizing the kidneys for transplantation selective renal arteriography was performed. The kidneys were of normal size and form. On both sides slight arteriosclerotic changes and a conspicuously wide, even diameter of all the extrarenal parts of the arteries were seen. The dilatation stopped abruptly where the arteries entered the renal parenchyma (Fig. 2).

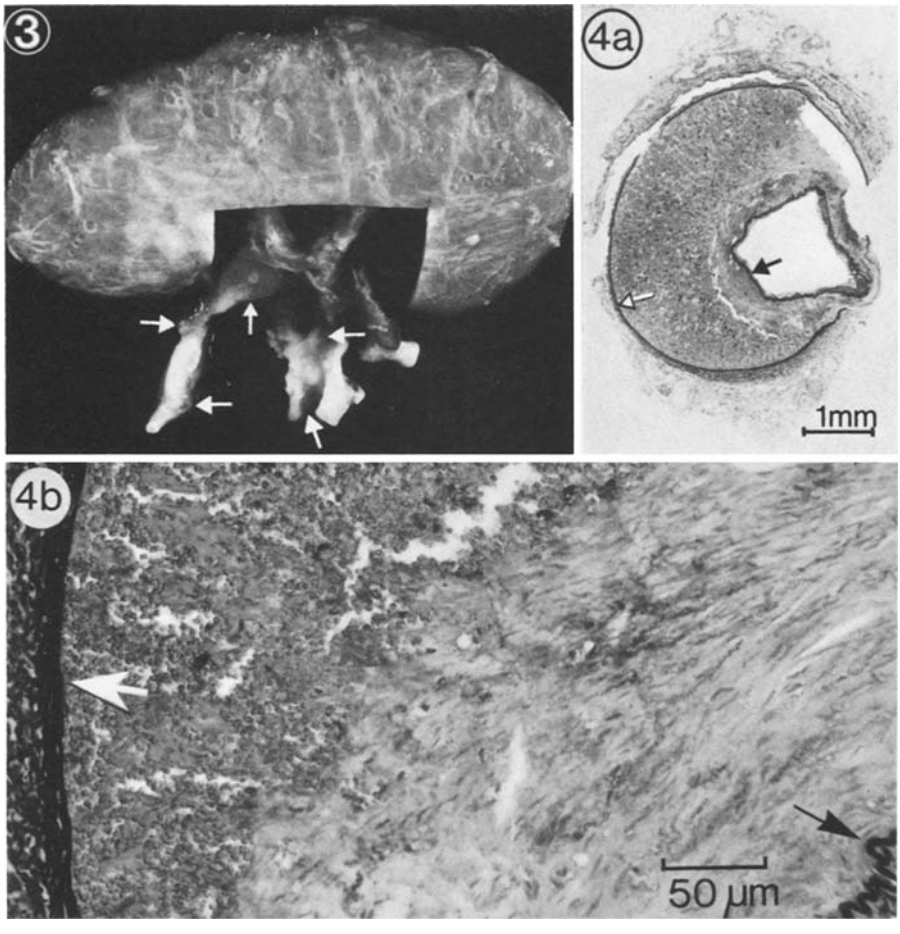

Fig. 3. Macrodissection of the left renal hilus. *Arrows* point at some of the aneurysms

Fig. 4a, b. Cross-section of renal artery showing dissecting bleeding in the external part of the media. *Arrows* indicate the internal (*black*) and external (*white*) elastic lamina. Elastic van Gieson stain

Death occurred 19 h after admission and immediately afterwards bilateral nephrectomy was performed.

Autopsy. The body was that of a 52-year-old caucasian woman weighing 46 kg and 154 cm tall. Incisions following boreholes in the scalp and bilateral nephrectomy were present.

The heart weighed 290 g. The thickness of the right ventricle was 3 mm, of the left ventricle 16 mm. The coronary arteries showed only slight arteriosclerosis. The aorta was moderately atheromatous with plaques and ulcerations in the abdominal part.

The brain weighed 1,400 g and there was heavy subarachnoid haemorrhage invading into the cerebral hemispheres. It was oedematous carrying the pressure marks of incarceration. On the left pericalosal artery 3 cm from the anterior communicating artery a 4 mm aneurysm was found with a 2 mm long tear. Otherwise the cerebral arteries were macroscopically normal.

The kidneys and spleen had been removed by surgery. The left kidney was intended for transplantation at our hospital, but this procedure was abandoned because of the findings at arteriography. The right kidney was sent for transplantation abroad (under the auspicies of the organisation of Scandia-Transplant), transplanted but removed a few days later due to the appearance of signs of severe acute rejection.

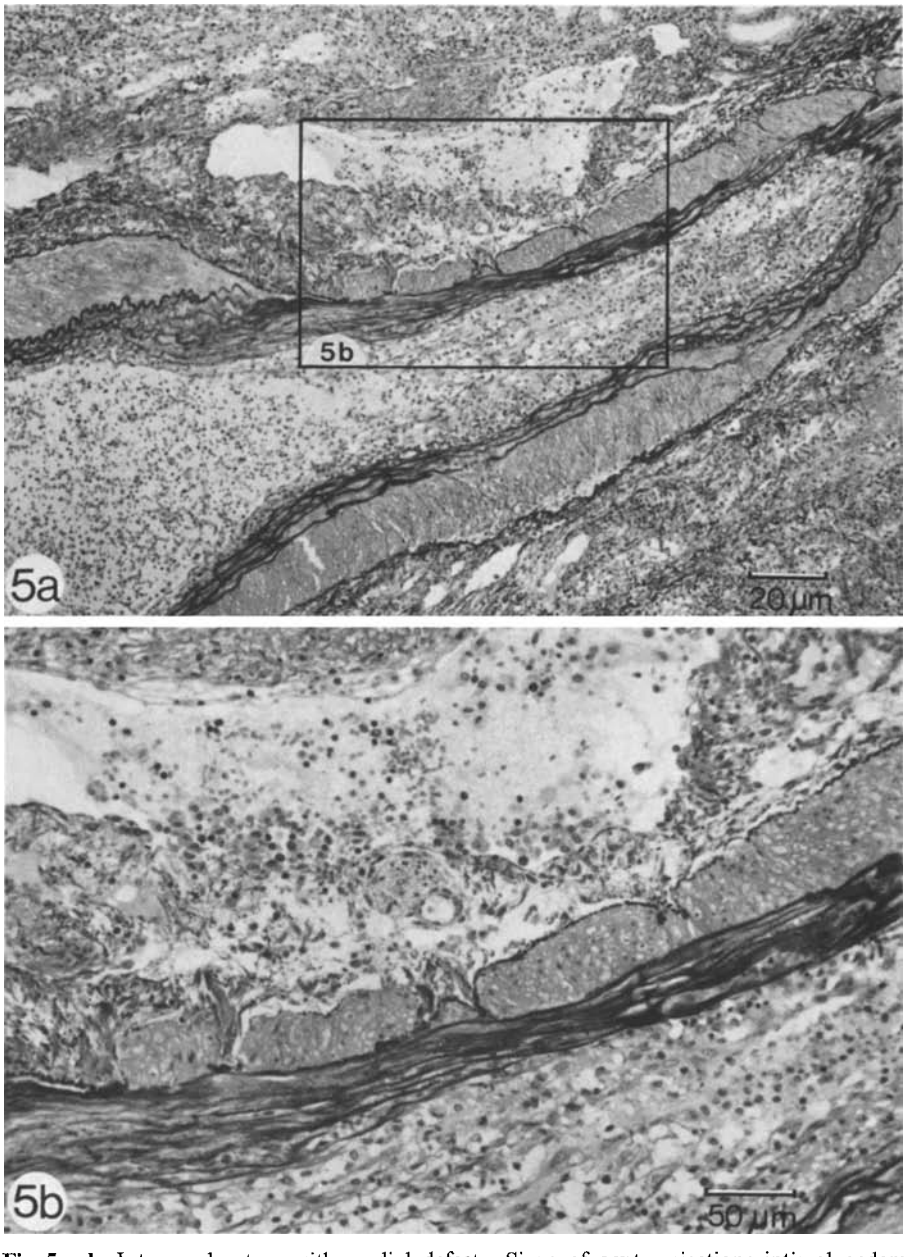

Fig. 5a, b. Intrarenal artery with medial defects. Signs of acute rejection: intimal oedema and lymphocytic infiltration. Elastic van Gieson stain. **b** Framed detail of **a**

The *left kidney* measured $12 \times 6 \times 3.5$ cm. The renal capsule was normal. The cut section of the renal parenchyma appeared normal. At the resection line the renal artery was normal but immediately distally multiple blue-black swellings were found (in distinct contrast to the wide, even structure seen radiographically) (Fig. 3). These changes affected the main artery as well as its primary branches. They subsided as the arteries entered the renal parenchyma.

Microscopy of this kidney showed a normal renal parenchyma. The swellings of the artery were due to lesions characteristic of dissecting aneurysms: there was haemorrhage in the external part of the media with a preserved external elastic lamina (Fig. 4). The elastic fibres were disorganised and ruptured in some areas and their number reduced. The grossly nodular appearance was due to an uneven thickness of the wall hematoma. Changes of fibromuscular dysplasia were not present in any section from the renal arteries, and the intrarenal arteries appeared normal.

The parenchyma of the *right kidney* showed the features of severe acute rejection with interstitial mononuclear cell infiltration, arterial intimal swelling and multiple small infarcts. This kidney also showed dissecting aneurysms in the extrarenal parts of the arteries. In one location an intrarenal artery showed a peculiar reduction of the thickness of the media, but without rupture or bleeding (Fig. 5).

Microscopical investigation of the cerebral arteries showed a ruptured aneurysm of the left pericalosal artery located in the position indicated above. Many sections of other cerebral arteries showed occasional mucinous degeneration of the media in the layer adjacent to the internal elastic lamina. Dissection or haemorrhage was, however, not present.

Varying degrees of atheromatosis of the *aorta* were present. There were no lesions compatible with cystic medial necrosis. Dissection or bleeding were not observed.

Discussion and review of the literature

The incidence at autopsy of *renal artery aneurysms of all types* is 0.01% (Altebarmakian et al. 1979; Ippolito and LeVeen 1960). Tcherdakoff et al. (1971) in order to evaluate hypertension, performed arteriography in 2,036 patients. In 27 (1.32%) of these patients a renal artery aneurysm was revealed by this method. Kincaid et al. (1968), however, in a selected series of 78 hypertensive patients with renal artery stenosis found 12 aneurysms, corresponding to an incidence of 15%.

Dissecting aortic aneurysms are relatively frequent. Hirst et al. (1958) in their review of the English literature of the period 1933–1953 found an incidence of 0.28% in several series representing 175,045 autopsies. Extension of an aortic dissection into the major peripheral branches is the rule rather than the exception. Thus this occurred in 437 out of 505 patients. The renal arteries were involved in 63 cases.

Spontaneously occurring dissecting aneurysms of peripheral arteries without aortic involvement is very rare. Nothing can be said about the incidence in general or about the incidence of dissection of the renal arteries in particular. From the English, German and French literature 48 cases can be traced, to which is added the present case.

Table 1 displays the pertinent data of these cases.

There is no consensus concerning relation to other vascular diseases or pathogenesis. Harrison et al. (1967) include dissecting aneurysm along with other stenosing and disruptive lesions of the renal artery (arteries) under the heading of fibromuscular dysplasia. These authors state that of 66 patients with renovascular hypertension due to idiopathic, non-atherosclerotic disruptive and hyperplastic lesions the stenoses were due to medial thickening in 44 cases, perimural fibrosis in 16 and dissecting aneurysm in 6 cases. McCormack et al. (1966) regard dissection as a complication of intimal and medial fibroplasia or fibromuscular hyperplasia. Excluding

Table 1. Published cases of spontaneously occurring dissecting aneurysms of the renal arteries without aortic involvement

Authors	Ref. no.	Sex	Age	Past/present inform. of hypertension	Right/left renal artery	Renal infarct.	Involvement of other arteries (except the aorta)	Operation	Death	Dysplasia
Gilfillan et al. (1956)	15	♂	47	+	L	+	−	+	−	−
Liebow et al. (1956)	25	♂	57	+	R/L	+/−	−	−	+	−
Watson (1956)	35	♂	52	0	R	−	−	−	+	−
		♀	49	+	R/L	+/+	−	−	+	−
Foord and Lewis (1959)	12	♂	27	+	R (L absent)	+	−	+	+	−
		♂	58	+	R	+	+[a]	−	+	−
		♂	79	−	R/L	+/+	+[b]	−	+	−
		♀	72	−	R	+	−	−	+	−
		♀	76	−	R	−	−	−	+	−
		♂	52	+	R	−	−	−	+	−
		♂	72	+	R/L	+/−	−	−	+	−
Ralston and Wasdahl (1960)	30	♂	51	−	L	−	+[c]	−	+	−
Henry and Burke (1963)	20	♀	66	0	R	+	−	−	+	−
		♂	72	−	R	+	?[d]	−	+	−
		♀	80	0	R	−	−	−	+	−
Neuman and Sahin (1965)	28	♂	43	+	L	+	−	−	+	−
Tuqan (1965)	34	♂	70	+	R	+	−	−	+	−
Englund (1966)	11	♂	49	+	R	+	?[e]	−	+	−
Rosenblum (1966)	31	♂	55	−	R	+	−	−	+	−
Boquist and Berg (1970)	6	♀	52	+	R	+	+[f]	−	+	−
Hare and Kincaid-Smith (1970)	17	♂	47	+	R	+	−	+	−	−
		♂	43	+	R	+	−	+	−	−
		♂	44	+	L	+	+[g]	−	−	−
		♂	39	+	L	0	−	−	−	−
		♀	42	+	R	+	+[h]	−	−	−
Tcherdakoff et al. (1971)	33	1 case	0	0	0	0	0	0	0	−
Perry (1971)	29	♂	37	+	R	0	−	+	−	+
Arnold and Evans (1972)	4	♂	59	+	R/L	+/−	−	0	+	−
		♀	41	+	R/L	−	−[i]	0	+	−
		♂	57	+	R	+	−	0	+	−

Cormier et al. (1972)	8	♂	45	+	L	+	+ [j]	+	−	−
Guthrie and McLean (1972)	16	♀	70	+	R/L	−	+ [k]	−	+	−
		♂	60	0	L	−		−	+	−
		♂	35	+	L	−	−	+	−	−
		♂	59	+	L	−	−	+	−	−
Cummings et al. (1973)	9	♂	56	+	R	−	−	+	−	−
		♀	44	+	L	+	−	+	−	+
Kaufman et al. (1973)	22	♂	57	+	R	−	−	+	−	+
Khanna et al. (1973)	23	♂	35	+	R	+	−	+	−	−
Meyers et al. (1974)	27	♂	31	+	L	+	−	+	−	+
Galenaud et al. (1977)	14	♂	38	+	R	+	−	+	−	−
Bakir et al. (1978)	5	♂	44	+	R/L	+/+	−	+	−	+
		♂	50	+	R/L	+/+	−	+	−	−
Foulon et al. (1978)	13	♂	43	+	L	−	−	−	−	−
Abet et al. (1980)	1	♀	44	+	R	−	+ [l]	+	−	−
		♂	33	+	L	−	−	+	−	+
		♂	51	−	L	+	−	+	−	−
		♂	29	+	R	−	−	+	−	−
Present case		♀	52	+	R/L	−/−	?[m]	−	+	−

Total number ♀/♂ = 12/36; mean age ♀ = 57.3 years (range 41–70 years); mean age ♂ = 49.3 years (range 27–79 years)

+ : present; − : not present; o : information not available
Dysplasia: + indicates a possible relationship between dysplasia and dissection; − that such a relationship has been denied or not commented upon

[a] Superior mesenteric artery
[b] Left common iliac artery
[c] Coronary artery
[d] Cerebral aneurysm
[e] Cerebral haemorrhage
[f] Coronary, coeliac, superior mesenteric and hepatic artery. Cerebral haemorrhage
[g] Left common iliac artery
[h] Hepatic, splenic and left internal iliac artery
[i] Subarachnoid and intracerebral haemorrhage
[j] Hepatic, splenic and jejunal artery
[k] Left costocervical, left gastroepiploic, left gastric, splenic and superior mesenteric artery
[l] Common hepatic artery
[m] Subarachnoid aneurysm and intracerebral bleeding

35 patients with intimal atherosclerosis from their series of 97 patients with renal arterial disease there were 45 females and 22 males with dysplastic changes, of these 33 had dissecting aneurysms. Hare and Kincaid-Smith (1970) emphasize that minor degrees of dissection in the renal artery lead to the formation of stricture and to the pathological changes which are now grouped together as fibromuscular dysplasia, rather than vice versa.

The female/male ratio of dysplasia shows a distinct female preponderance (Harrison et al. (1967): 3/1; McCormack et al. (1966): 2/1) which is opposed to a heavy male preponderance of dissection: in the series collected by the present authors there were 13 females and 36 males.

The average age of dissecting aneurysms is about 50 years, which is also in contrast to the mean age of the patients with dysplasia which usually presents in adolescence. These differences indicate that the two diseases are distinct entities.

Trauma following selective arteriography (Delin et al. 1979; Hare and Kincaid-Smith 1970; Talner et al. 1975) or blunt external trauma (Boyd and Watson 1956) has been reported as a cause of dissection of the renal arteries, as has a subadventitial angioma (Acconia and Manganelli 1978).

Unfortunately it is not possible by studying the literature to state how often or for how long hypertension has been present in patients with dissection of the renal artery. Thus, a distinction is not always made between hypertension present before symptoms of dissection and a recognized development of hypertension in the course of dissection. In our patient hypertension was not present before she fell ill.

Although the reason for the anomaly remains obscure we think it is fair to state that our patient suffered from a generalized disease of the arteries as she also had certain changes in the cerebral arteries. However, we do not feel inclined to group the changes under a specific diagnostic heading, and they differ from what is named fibromuscular hyperplasia in general, and also from the concept of the medionecrosis of Erdheim (there are no pools of mucinous material in the renal arteries and the focal mucinous degeneration observed in the media of the cerebral arteries is positioned just beneath the internal elastic lamina). Our case does not fulfill the angiographical signs of Moyamoya disease (abnormal net-like mesh of arterial vessels at the base of the brain), an entity which has mainly been reported from Japan and of which the pathology apparently still has to be defined (Oka et al. 1981; Rose 1983).

The treatment of the ailment is surgical (nephrectomy or arterial reconstruction (e.g. Abet et al. 1980)) – if intervention is possible at all – to prevent hypertension and renal infarction.

The circumstances of the reported case are exceptional and it is not reasonable to draw any conclusions as to the general practice of selecting and investigating kidneys for transplantation. It is evident, however, that such kidneys, if found, should not be used for the purpose of transplantation.

Acknowledgements. We would like to thank Dr. E. Reske-Nielsen, Department of Neuropathology, and Dr. H. Halaburt, Department of Neuroradiology, Aarhus Kommunehospital, for advice and practical help with this study.

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Accepted August 2, 1983