Virchows Arch [Pathol Anat] (1981) 391:241–247

Virchows Archiv A

Dissecting Aneurysm of the Pulmonary Artery

Helmut Lüchtrath
Pathologisches Institut, Koblenzerstraße 115, D-5400 Koblenz, Federal Republic of Germany

Summary. Dissecting aneurysm of the aorta is often seen; similar changes in the pulmonary artery are rare. In the German literature they are unknown. 11 previously described cases have been compiled with their clinical and pathological records, and a new added. The patient, a 45 year old woman, suffered from pulmonary hypertension which resulted in medionecrosis and a large aneurysm of the trunk of the pulmonary artery. She died of haemopericardium after rupture of the artery in two stages, with a tear of 8 cm in the trunk which reached to the bifurcation of the vessel.

Key words: Pulmonary dissecting aneurysm – Pulmonary hypertension – Haemopericardium

Introduction

Dissecting aneurysm of the aorta is a well defined clinical and pathological entity. It is astonishing that dissecting aneurysm of the pulmonary artery has been observed only few times although pulmonary hypertension is not a rare disease (Horat 1970) and the architecture of the two large vessels is similar. There are a few reports of aneurysm of the pulmonary artery (Rokitansky 1852; Heeger and Kubicek 1968; Hartwell and Tilden 1943; Wilkinson 1940; Deterling and Clagett 1947) with or without persistent ductus arteriosus Botalli or with malformations of the great vessels (Crumpton 1950). In literature there are only 11 descriptions of the dissecting aneurysm of the pulmonary artery (Watson 1956; Foord and Lewis 1959; Epstein and Naji 1960; Best 1967; D'Arbela et al. 1970; Tikoff and Bloom 1970; Ferenc 1977; Tomov et al. 1979) excluding those which are caused by tuberculosis or bronchogenic carcinoma within the lung. The remaining 11 cases have many features in common (Table 1). The patients were predominantly young, the oldest being 50 years of age, the peak reaching from 20–25 years. The anatomical changes always

Offprint requests to: H. Lüchtrath

0340-1227/81/0391/0241/$01.40

Table 1.

Author	Age	Sex	Clinical findings	Localisation of the rupture. Autopsy	Malformations basic diseases
1. Walshe (1862) (cit. a. Watson)	21	m	Idiopth. pulmonal. hypertension	Trunk of the pul. art. haemopericardium	–
2. Duffield (1882) (cit. a. Watson)	50	f	Pulmonal. hypertension emphysema sudden death	Right branch of the pulm. artery	–
3. Durno (1908) (cit. a. Watson)	33	m	Pulmonal. hypertension sudden death	Trunk of the pul. art. haemopericardium	Duct. art. Bot. persistens
4. Crumpton (1950)	19	m	"Blue baby" since birth cyanosis, heart failure, pulmonal. hypertension. Death 10 h after attack of pain	Trunk, 2 cm after origin. rupture into mediastinum	Truncus com. of the heart
5. Odinokova (1956)	43	f	Since childh. rheumat. heart failure a. cyanosis, death 2 days after hospitalization	Trunk 2,5 cm behind pulmon. valves haemopericardium	Rheumatic stenosis of mitral valves
6. Foord (1959)	26	f	Connat. heart failure, sudden substernal pain shock, death 30 min after begin	Trunk a. right branch, tear of ∅ 6 cm. rupture into mediastium	Duct. art. Bot. persistens/ sclerosis of pulmon. art.
7. Epstein and Naji (1960)	19	m	At birth cyanotic, at 6 years Blalock operation, at 17 years episodes of haemoptysis. Sudden dull precordial pain, cyanose, later death	Trunk of the pulm. art. tear of 3 cm distal to the atretic pulm. valves haemopericardium	Tetralogy of Fallot
8. D'Arbela (1970)	18	f	Dyspnoea, pains, pulmonal. hypertension, death at defaecation, 4 weeks after admission	Trunk of the pulm. art. haemopericardium	Duct. art. Bot. persistens, medionecrosis, hypertens. disease of the pulm. arteries
9. Tikoff (1970)	23	m	Connat. heart failure, clubbings, suspicion of trunk, com. Sudden substernal pain. Death 5 h later	Trunk of the pulm. art. 4 cm behind the valves, haemopericardium	Ventricular septal defect, duct. art. persist. Malformation of the thoracic aorta
10. Ferenc (1970)	newborn		Hungarian original only abstract available	–	–
11. Tomov (1979)	adult	m	Prim. pulmonal. hypertension, bulgarian original, only abstract available	–	–
12. Own case	45	f	Since 5 years dyspnoea pulmonal. hypertension. Sudden pain, shock, died 18 h later	Trunk a. bifurcation of the pulm. art. 8 cm tear, haemopericardium	Hypertens. disease of the pulm. arteries. Prim. pulmon. hypertonie.

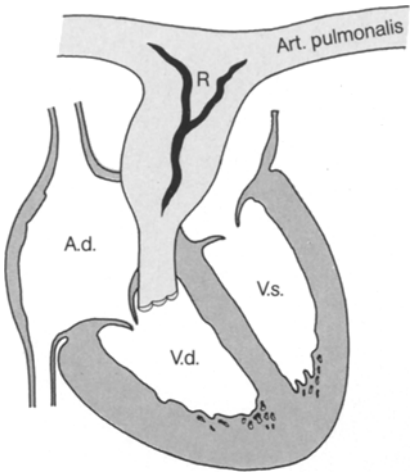

Fig. 1. Rupture (*R*) of the trunk of the pulmonary artery and branches. Aneurysm of the artery. *A.d.* Atrium dextr.; *V.d.* Ventricul. dext.; *V.s.* Ventricul. sinistr.

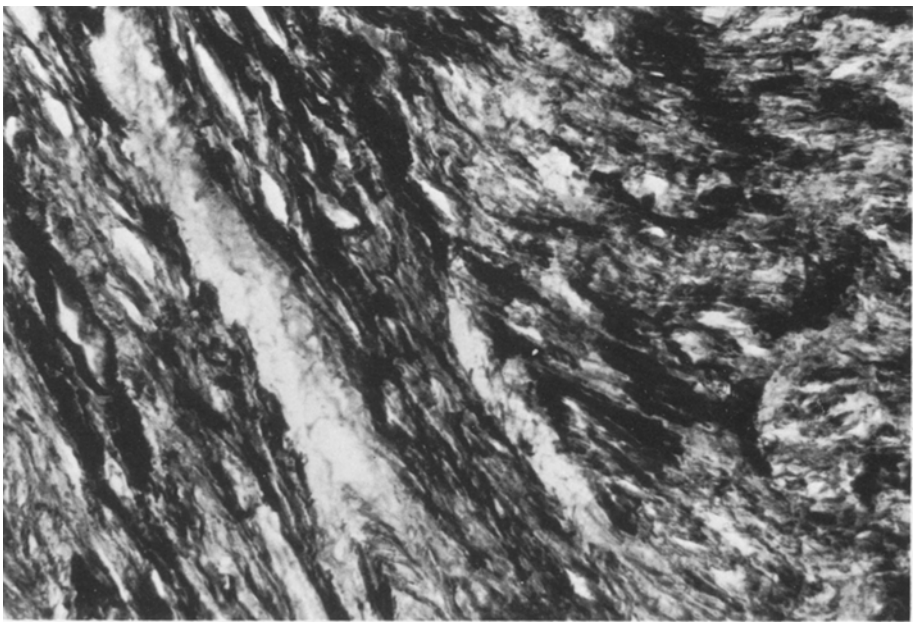

Fig. 2. Medionecrosis with small fissures. Elast.-van Gieson, 520:1

showed that there had been a secondary pulmonary hypertension, which was the cause of the aneurysm. Only Walshe (1862) reported a case with primary pulmonary hypertension but his clinical record is fragmentary.

Case Report

A 45 year old woman complained of dyspnoea, cough and tachicardia a 5 year period. One year before death a pulmonary hypertension had been found with a mean pressure of 65 mm

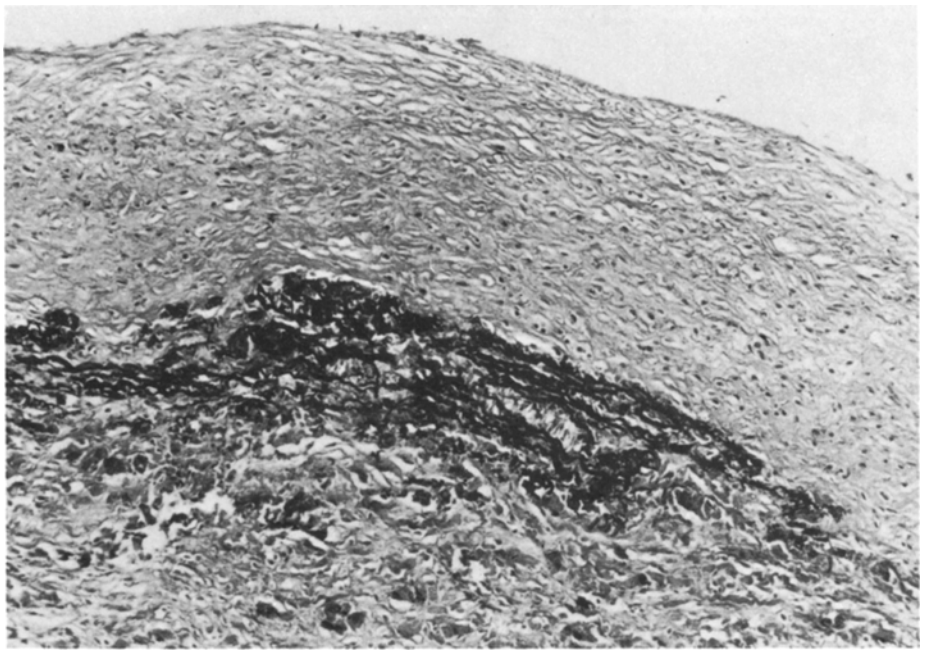

Fig. 3. Destroyed arterial wall with collagen tissue and few remaining elastic fibers. Elast.-van Gieson, 210:1

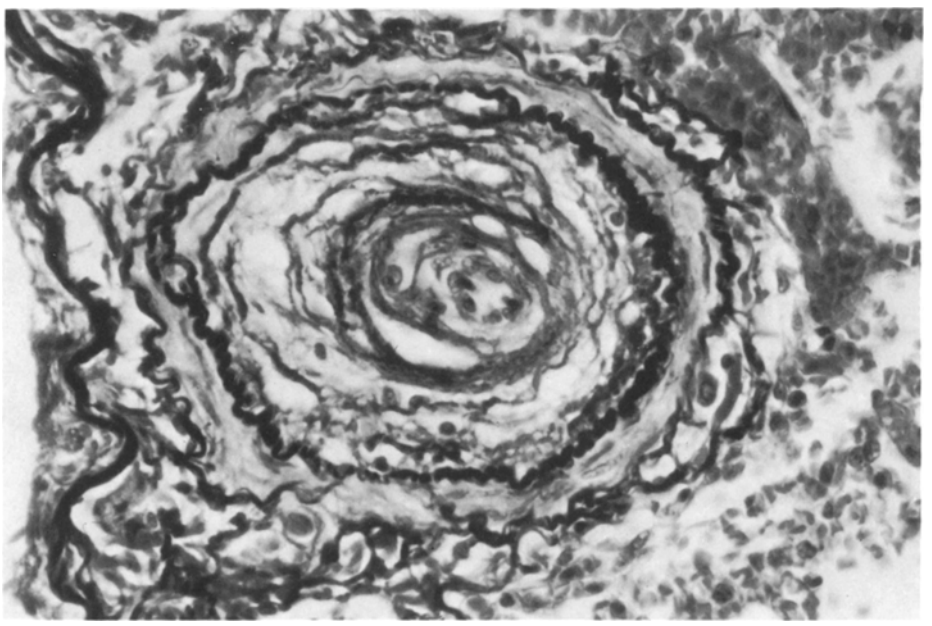

Fig. 4. Hypertensive lesion in the pulmonary arteries: Concentric fibroelastosis of the intima. Elastin, 400:1

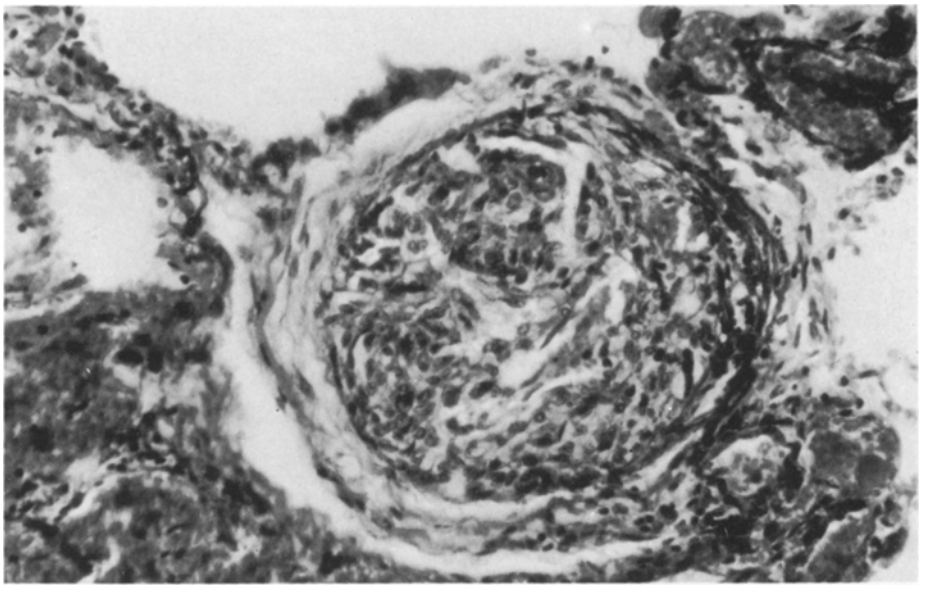

Fig. 5. Hypertensive changes in the pulmonary arteries: Plexiform lesion. Elastin, 250:1

Hg in the pulmonary artery. The resistance in the lesser circulation was increased 8–10 times. An electrocardiogram showed hypertrophy of the right ventricle. There was a systolic parasternal cardiac murmur loudest in the second left intercostal space. A year later she was admitted to hospital after a sudden onset of abdominal pain with the symptoms of shock and cyanosis. The blood pressure was not measurable. The next day she died after having had another attack of pain in the chest.

Autopsy showed a dissecting aneurysm in the trunk and bifurcation of the pulmonary artery. There was a tear of 8 cm within the intima and media of the vessel (Fig. 1) and a haemorrhage into the adventitia with tamponade of the heart and 600 ml blood in the pericardium. The pulmonary artery showed severe dilatation with advanced arteriosclerosis, especially in the middle and peripheral branches. There was a marked hypertrophy (1.1 cm) of the right ventricle with dilatation and relative insufficiency of the pulmonary valve. The wall of the aneurysm, the pulmonary artery and the lungs were examined histologically after embedding in paraffin. The slides were stained with haematoxylin, picrin-acid-fuchsin, resorcin-fuchsin, alcian blue, PAS and silver impregnation (Gomori).

In the wall of the pulmonary artery there was spotty oedema, some basophil foci with mucoid degeneration and defects of the elastic fibers. There were also small fissures (Fig. 2). No signs of inflammation were noted but some minor haemorrhages had occured. The findings were those of medionecrosis idiopathica of the aorta. The elastic fibers within the aneurysm wall were destroyed, leaving only a few bundles (Fig. 3). Scar tissue was apparent. The wall was extremely thin and extended. Near the rupture the blood had lacerated the intima and media and separated an outer layer of the media. In the intima severe arteriosclerotic deposits were seen.

The lung revealed severe sclerosis of the large and middle sized arteries. The small vessels showed marked hyperplasia of the media with proliferation of the intima (Figs. 4 and 5).

Discussion

This woman suffered from pulmonary hypertension of long duration. Lung changes corresponded with the histological findings of hypertensive changes grade III–IV (Heath and Edwards 1958) as recently described by Leu et al.

(1979). The hypertension was followed by severe sclerotic changes in the trunk and bigger branches of the pulmonary artery with degeneration in the media similar to that seen in the aorta in medionecrosis idiopathica (Erdheim 1930) with mucoid spots, destroyed elastic fibers replaced by scar tissue, with an important structural failure which resulted in weakness and aneurysmal expansion of the wall. Rupture obviously occured in the inner layers and was accompanied by shock which led to hospitalization. The 2nd stage was haemorrhage in the pericardium, the cause of death.

All such cases described in literature showed severe changes in the heart and large vessels (Table 1), partially in form of congenital malformations (Truncus comm., Tetralogy of Fallot). Valvular defects are important (rheumatic stenosis of the mitral valves) with secondary pulmonary hypertension leading to dissecting aneurysm of the pulmonary artery. In nearly all cases the rupture of the aneurysm took place in the truncus and caused a haemo-pericardium. This is the explanation for the sudden attacks of pain in the chest and the death of most of the patients within minutes or hours.

In our case there were neither malformations nor a failure of the heart. The ductus arteriosus was closed. No drugs were used which could have caused a pulmonary hypertension. We assume therefore that a primary pulmonary hypertension existed. Inflammatory or sclerotic depots were not seen.

Acknowledgements. I wish to thank Dr. Stein, Krankenhaus Marienhof, Koblenz for the clinical informations and Prof. Dr. Rüttner, Zürich for his kindly support. I am grateful to Mrs. G. Clos for technical assistance.

References

Best J (1967) Dissecting aneurysm of the pulmonary artery with multiple cardiovascular abnormalities and pulmonary hypertension. Med J Aust 2:1129–32

Crumpton M (1950) Congenital heart disease and dissecting aneurysm of pulmonary artery. Br Med J 1303–1308

D'Arbela PG, Mugerwa JW, Patel AK, Somers K (1970) Aneurysm of pulmonary artery with persistent ductus arteriosus and pulmonary infundibular stenosis. Br Heart J 32:124–126

Deterling RA, Clagett OT (1947) Aneurysm of the pulmonary artery: Review of the literature and report of a case. Am Heart J 34:471–499

Epstein S, Naji AF (1960) Pulmonary artery aneurysm with dissection after Blalock Operation for tetralogy of Fallot. Am J Cardiol 5:560–564

Erdheim J (1930) Medionecrosis Aortae idiopathica cystica. Virchows Arch [Pathol Anat] 276:187–229

Ferenc B (1977) Dissecting aneurysm in the pulmonary artery of a newborn infant. Morphol Igazsagugyi Orv SZ 16:293–295 (in ungarischer Sprache)

Foord AG, Lewis RD (1959) Primary dissecting aneurysm of peripheral and pulmonary arteries. Arch Pathol 68:553–577

Hartwell AS, Tilden JL (1943) Aneurysm of the pulmonary artery. Am Heart J 26:692–698

Heath D, Edwards JE (1958) The pathology of hypertensive pulmonary vascular disease. Circulation 18:533–547

Heeger H, Kubicek F (1968) Idiopathisches Aneurysma der Arteria pulmonalis. Zur Ätiologie und Diagnose. Z Kreislaufforsch 57:306–312

Horat W (1970) Primäre vasculäre pulmonale Hypertonie. Z Kreislaufforsch 59:91–93

Leu HJ, Rüttner JR, Hürlimann P (1979) Hypertensive Pulmonalarterienveränderungen bei chronisch-entzündlichen Lungenerkrankungen. Virchows Arch [Pathol Anat] 383:283–292

Odinokova VA (1956) Dissecting aneurysm of the pulmonary artery. Arkh Patol 18:87–89
Rokitansky K (1852) Über einige der wichtigsten Krankheiten der Arterien. Kaiserl.-Königl. Hof- und Staatsdruckerei, Wien
Tikoff G, Blomm S (1970) Complete interruption of the aortic arch in an adult associated with a dissecting aneurysm of the pulmonary artery. Am J Med 48:782–786
Tomov J, Nikolov G, Elenkova A, Goranova J (1979) Case of primary pulmonary hypertension leading to a dissecting aneurysm of the pulmonary artery. Vutr Boles 18:88–93 (in bulgarischer Sprache)
Watson AJ (1956) Dissecting aneurysm of arteries other than the aorta. J Pathol Bacteriol 72:439–449
Wilkinson KD (1940) Aneurysmal dilatation of pulmonary artery. Br Heart J 2:255–258

Accepted November 27, 1980