PULMONARY HEART DISEASE

PATHOLOGICAL CLASSIFICATION OF PULMONARY VASCULAR DISEASE

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Haemodynamic alterations in the pulmonary circulation, whether due to acquired or congenital heart disease or to lung disease, as a rule are accurately reflected by changes in the pulmonary vasculature.

Conversely, the pathologist, by studying the histological lesions of the lung and its vessels in autopsy material or in lung biopsies, may contribute to a better understanding of the cardiac or pulmonary abnormalities involved. These lesions generally form a pattern which may fit into a useful classification.

Group 1. Pulmonary hypertension due to prae-tricuspid or post-tricuspid shunts, such as atrial or ventricular septal defects or patent ductus arteriosus.

Medical hypertrophy of muscular pulmonary arteries is the first and most wide-spread alteration. In cases of ventricular septal defect, when pulmonary hypertension will be present from birth, it may be observed almost immediately after birth. This has given rise to the term "carry-over of the fetal type of vessel", suggesting that the thick-walled fetal arteries fail to become thinner in the post-natal period. It is unlikely that this is completely true. There is evidence that in any infant with a ventricular septal defect there is an initial thinning of the media due to the normal post-natal dilatation of the arteries, although this is followed within a few weeks by marked medial hypertrophy which may become severe within a few months (Wagenvoort et al., 1961).

This medical hypertrophy is likely to result from an increased tone and vasoconstriction thus induced, contributes to an increased pulmonary vascular resistance and pressure (Wagenvoort et al., 1967). The medial thickness is roughly proportional to the pulmonary hypertension. As far as we know, the lesion is reversible should the defect be repaired (Dammann et al., 1961).

Intimal fibrosis is a later phenomenon, rarely observed in young infants. In principle it is a non-specific change as it may be the end-result of any reparative process or of organization of thrombi or emboli, while it is also a normal age change. Even so, the type of intimal proliferation and fibrosis observed in congenital cardiac shunts differs from that of other forms by being concentric and laminar with an onion-skin arrangement (Fig. 1). Various phases in its development can often be recognized. It may lead to marked narrowing or to complete obstruction of the vessel. As medial hypertrophy, it is also roughly proportional to the pressure in the pulmonary artery. It is not or only to a limited degree reversible.

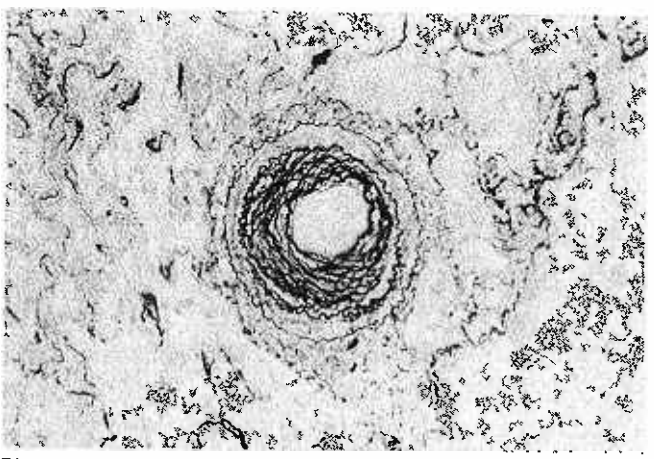

Fig. 1. Muscular pulmonary artery with medial hypertrophy and concentric laminar intimal fibrosis. There is an older phase with dense elastic tissue and a more recent phase with intimal proliferation (E.v.G., × 150).

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Fibrinoid necrosis with or without arteritis is less often encountered (Fig. 2). It almost certainly results from intense vasospasm and is limited to the more severe forms of pulmonary hypertension. In turn it may give rise to the so-called **plexiform lesions** (Fig. 3), complicated structures in which organization of the fibrin clot by intimal cells leads to a plexus in a small segment of the artery, usually with destruction of its wall and with dilatation of the peripheral part of the artery (Wagenvoort et al., 1964). Isolated dilatation of convoluted arteries, the so-called **dilatation lesions** may also be present.

When medial hypertrophy is accompanied by concentric intimal fibrosis, and particularly by fibrinoid necrosis, plexiform lesions and dilatation lesions, a prae- or post-tricuspid shunt must be the cause of the pulmonary vascular disease. To this rule there are only two exceptions. Vaso-constrictive primary pulmonary hypertension may give exactly the same pattern, usually in its more severe forms (Wagenvoort et al., 1970). Also in pulmonary schistosomiasis the various lesions just described may be present, although in these cases

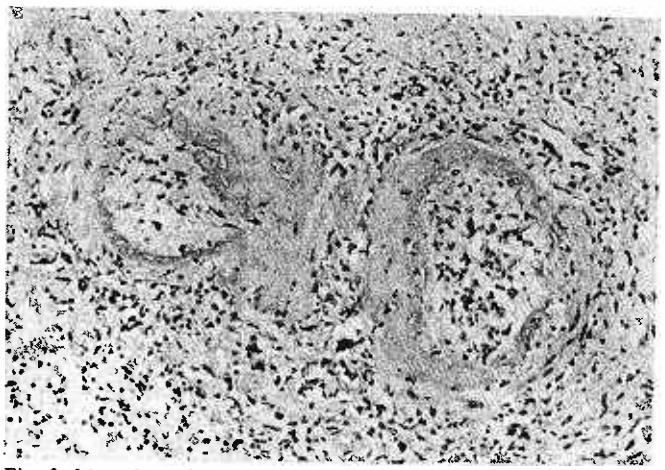

Fig. 2. Muscular pulmonary artery with fibrinoid necrosis and arteritis (H. and E., × 100).

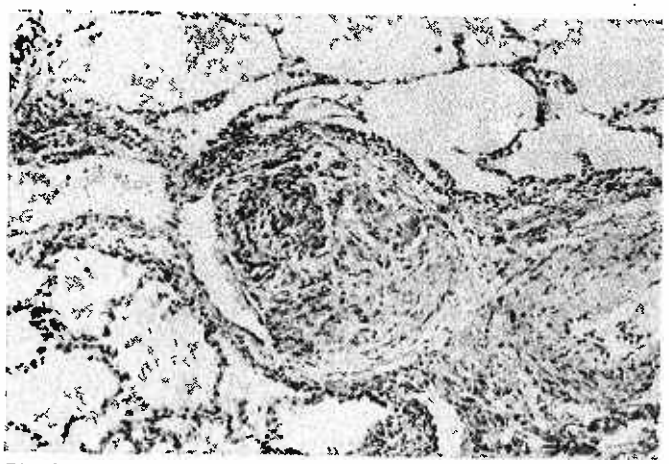

Fig. 3. Muscular pulmonary artery with plexiform lesion. In part the channels contain fibrin thrombi. The wall is partly destroyed with some inflammatory reaction (H. and E., × 100).

granulomas, often with ova of the parasite, are added to the picture.

It must be added that severe pulmonary vascular disease is particularly found in cases of ventricular septal defect, more than in patent ductus arteriosus, while it is rare in atrial septal defect. A large pulmonary flow, in the absence of a markedly elevated pressure, as often is characteristic for an atrial septal defect, apparently has but little influence on the lung vessels.

Group 2. Pulmonary hypertension due to chronic pulmonary embolism. In this condition there is an entirely different pattern. The muscular pulmonary arteries may be narrowed or even obliterated by intimal fibrosis but this is of an excentric patchy type without laminar arrangement. Usually the fibrosis, which is due to organization of emboli, extends over a relatively short distance. Recanalization is common and may lead to the formation of intraluminal fibrous septa (Wagenvoort et al., 1970).

Medial hypertrophy is generally slight to moderate but may even be absent. This probably indicates that mechanical obstruction in these cases is more important than vasoconstriction in the production of the elevated pressure. Arteritis is very rare, plexiform lesions and dilatation lesions are never observed in these instances.

Group 3. Pulmonary hypertension due to obstructed pulmonary venous outflow, such as mitral valve disease.

In these conditions both the pulmonary arteries and veins are affected. In the arteries medial hypertrophy is always present and often severe. In contrast to cases of congenital cardiac shunts, some fibrosis of the media is regularly observed, while there is no good correlation of the medial thickness with pulmonary arterial pressures, at least not with those recorded at rest.

Intimal fibrosis is a regular feature and may be severe, although it does not often lead to obliteration. It may be either excentric or concentric, but not laminar. Arteritis is uncommon; there are no plexiform lesions or dilatation lesions.

The walls of the pulmonary veins are usually increased in thickness. Both the media and the intima are usually thicker than normal, but the most characteristic feature is arterialization of the venous wall which acquires the elastic configuration of a pulmonary artery with internal and external elastic laminae.

Wide-spread intimal fibrosis of pulmonary veins and venules with obliteration of the lumina is only observed in the rare cases of pulmonary veno-occlusive disease. In these instances there are no cardiac malformations but the obstruction is thrombotic in origin.

Group 4. Pulmonary hypertension due to chronic respiratory disease and hypoxia, such as chronic bronchitis and emphysema. Even in cases in which elevation of pulmonary arterial pressure and right ventricular hypertrophy are marked, medial hypertrophy as a rule is absent or only slight to moderate. The degree of intimal fibrosis varies greatly and is often dependent on the presence of inflammation or fibrosis in the lung tissue.

The only lesion in these cases which is almost pathognomonic for chronic hypoxia, is the presence of longitudinal smooth muscle bundles in the intima of small and medium-sized muscular pulmonary arteries (Hicken et al., 1965). This feature is also commonly observed in individuals living at high altitudes and exposed to the hypoxia prevailing there.

Group 5. Decreased pulmonary vascular flow as in pulmonary stenosis and tetralogy of Fallot.

In these cases of decreased pulmonary flow, the pulmonary arterial pressure is only slightly lower than normal. Even so this may affect the media in that it becomes thinner, sometimes to a pronounced degree of medial atrophy with loss of all medial muscle tissue. The vessels are relatively wide. Medial atrophy may be present at the time of birth as in infants with pulmonic atresia.

Intimal fibrosis is common, particularly in older children and adults and is always of the patchy, excentric type, indicating organization of thrombi. A remarkable feature is excessive recanalization leading to the formation of delicate septa within the lumen (Rich, 1948).

Although with these five patterns the diagnostic possibilities in studying the pulmonary vasculature, are not yet exhausted, they include the most important and most frequent forms of vascular disease in the lung.

Sometimes features of various patterns may be present. In a case of primary pulmonary hypertension for instance the condition may be complicated by alterations in the pulmonary veins, as might happen, although uncommonly, when chronic cardiac insufficiency is added to the picture. Also chronic pulmonary embolism may complicate mitral valve disease. When this is realized, an accurate diagnosis can be made.

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