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Plexiform lesions with giant cells

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The plexiform lesion is one of the most striking histological lesions associated with pulmonary hypertension. It is the diagnostic feature of plexogenic pulmonary arteriopathy, the designation given by a working party of the World Health Organisation[1] to that form of hypertensive pulmonary vascular disease which may complicate pre- and post-tricuspid congenital cardiac shunts, primary pulmonary hypertension, and rare cases of cirrhosis of the liver and portal vein thrombosis.[2] This lesion has been studied intensively and both its histological and ultrastructural features are familiar to morbid anatomists. Recently we saw a histopathological variant with giant cells which was so unusual as to merit report.

Pulmonary vascular pathology

The lung tissue studied was obtained at necropsy from a woman of 26 years, with an aortopulmonary septal defect complicated by pulmonary hypertension, who had undergone cardiac catheterisation. The "muscular pulmonary arteries" (100 to 1000 μm in external diameter) showed medial hypertrophy and concentric-laminar intimal proliferation of the "onion-skin" variety. There were many plexiform lesions composed of dilated, sac-like branches of pulmonary arteries in which had developed a plexus of thin-walled vessels separated by septa containing plump cells with rounded nuclei. In some of the plexiform lesions, presumed to be older, the septa were thicker and fibrous. The parent vessels showed patchy "fibrinoid necrosis" which is known to be the basis for the development of plexiform lesions.[3] Some of the elastic pulmonary arteries showed calcification and disruption of their elastic laminae with small mineralised deposits in the intima and adventitia. The histology of the plexiform lesions was unusual for, scattered among the anticipated cells forming the proliferation, there were large foreign body giant cells with multiple nuclei (fig 1). Many of them had engulfed small fragments which were pale blue in sections stained with haematoxylin and eosin. These fragments proved to be brightly refractile in polarised light (fig 2). Fortuitous sections revealed that the intracellular foreign bodies were derived from long fibres, up to 66 μm in length, with a central linear split.

Discussion

The plexiform lesion is one form of "dilatation lesion" which develops in the late stage of hypertensive pulmonary vascular disease complicating a congenital cardiac shunt. Its appearance is associated with a fall in pulmonary blood flow below normal levels and a pronounced increase in pulmonary vascular resistance[4] which becomes irreversible.[5] It is formed by striking localised dilatation of a branch of a muscular pulmonary artery,[6] to form a sac. In this sac develops a plexus of thin-walled vessels which are separated by septa formed by an associated proliferation of cells streaming in from the parent vessel. This proliferation of small vessels and cells appears to represent the organisation in the sac of fibrin which is washed into it from areas of fibrinoid necrosis in the parent artery. Electron microscopy reveals that the proliferated cells are myofibroblasts and "fibrillary cells" which are closely related to the so-called vasoformative reserve cells.[3] Giant cells are not a usual feature of the histopathology of the plexiform lesion.

The plexus of vessels and cells in these sacs probably

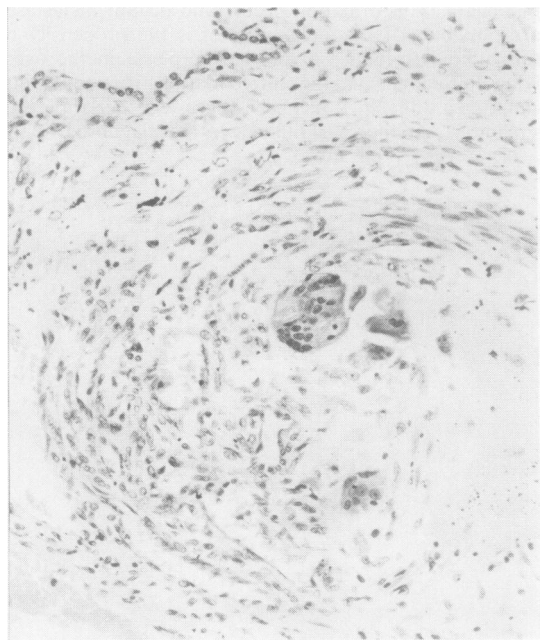

Fig 1 *Plexiform lesion composed of thin-walled vessels separated by septa of plump cells with rounded nuclei. Also present are multinucleated giant cells. Careful examination of these reveals small intracellular foreign bodies. Haematoxylin and eosin, × 190.*

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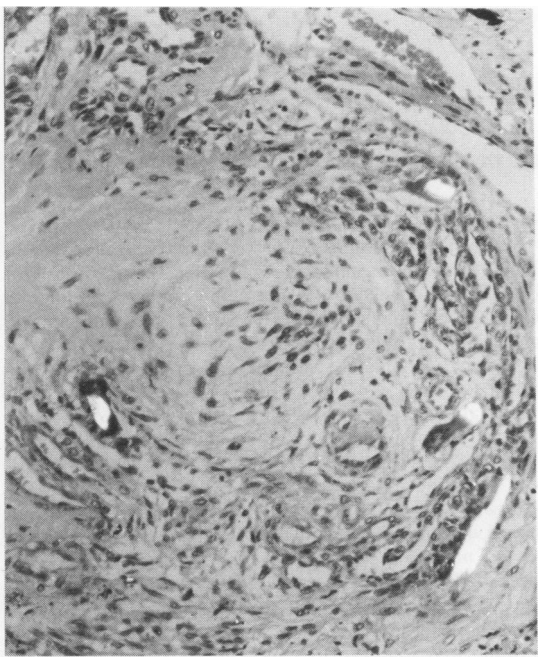

Fig 2 *Plexiform lesion photographed in polarised light. Four brightly doubly refractile foreign bodies are seen. The three upper bodies are partly engulfed by giant cells. The lower is linear in form and is consistent in appearance with being a fragment of cotton fibre. Haematoxylin and eosin, in polarised light, × 243.*

constitutes an effective sieve, filtering off particulate matter flowing into it from the pulmonary arteries. In this case large, doubly-refractile fibres containing a central slit circulating in the blood appear to have been trapped by the plexiform lesions and then provoked a vigorous foreign-body giant cell reaction. From our previous experience we know that these foreign bodies are cotton-fibre emboli. It is well known that such emboli may be introduced inadvertently by clinicians into systemic veins and then impact in pulmonary arteries leading to a giant-celled granulomatous reaction. We have reported such a case in a woman with a patent ductus arteriosus who was subjected to cardiac catheterisation and in whom the catheter was contaminated by wisps of cotton wool.[7] More recently we have carried out experimental studies in rats to study the details of the formation of such granulomas around cotton fibres and their passage through the pulmonary arterial wall to the adjacent alveolar spaces.[8] In the present case the fibres have been trapped in plexiform lesions rather than become impacted in the walls of pulmonary arteries and have produced the unusual histological appearances described.

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