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Commentary

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These four case reports have a common theme – the pulmonary circulation – but the subject matter of each is actually quite different.

Gosney and Rešl, in a post mortem study, report histopathological findings in a case of primary pulmonary hypertension, associated with liver cirrhosis of unknown aetiology, in a 44 year old woman. In particular, they failed to find the usual proliferation of pulmonary endocrine cells which they have described as a feature of pulmonary hypertension with plexiform lesions. The cirrhosis was "long standing" but the pulmonary hypertension was never suspected in life, so we do not know the date of onset of either condition. Two questions arise: (1) is there a causal association between plexogenic pulmonary arteriopathy and cirrhosis, and (2) is lack of proliferation of pulmonary endocrine cells (whose products are linked with gastrin, calcitonin, and serotinin) a significant finding in this one case or does it represent just one end of a spectrum? The answer to the second question must await the analysis of more cases.

The most frequent pulmonary vascular pathology in cirrhosis is a diffuse structural *dilatation* of the microvasculature leading to pulmonary *hypotension* and severe hypoxaemia due to "intravascular shunting". This is now called the "hepatopulmonary syndrome" and up to 30% of patients in liver failure may be affected, but the true incidence – particularly in subclinical forms – has never been assessed. This structural vasodilatation is consistent with other features of liver failure such as clubbing, spider naevi, varices, a high cardiac output, and a hyperdynamic circulation. The molecular pathogenesis remains unknown.

Primary pulmonary hypertension in liver cirrhosis is a much rarer phenomenon. It has clinical importance as a contraindication to liver transplantation. On the other hand, intrapulmonary shunting generally improves after liver transplantation and is a positive indication for it. The incidence of plexogenic pulmonary hypertension in portal hypertension is about 2%; the vascular changes are so different from the more common "hepatopulmonary syndrome" that I feel pulmonary hypertension is a "personal" response to liver disease rather than part of the syndrome of hepatocellular failure.

Case reports frequently involve rare or unique cases, sometimes called "nature's experiments". Their value is to stimulate "lateral thinking". Koga *et al* report the fourth ever case of pulmonary involvement in neoplastic angioendotheliosis (a B cell lymphoma invading arterial walls and leading to vascular occlusions).

How does one diagnose an angiopathic neoplasm of the lung? In pulmonary function testing a lone diffusion defect (in this case, FEV_1 and VC >80% predicted, TLCO <30%) points towards selective pathological involvement of pulmonary blood vessels, and this was supported by an abnormal perfusion scan (not typical of pulmonary embolic disease). Pulmonary vasculitis was (quite correctly) considered at this stage. The patient was too ill for open lung biopsy, and empirical treatment with corticosteroids and anticoagulants was instituted. Tumour cells were eventually found in the bone marrow and in the peripheral blood; pulmonary involvement was confirmed ultimately after a post mortem examination. This case illustrates the specificity of a lone low TLCO value as a marker of pulmonary vascular involvement, even though it is quite non-specific for the particular type of vascular pathology.

Another form of pulmonary vasculopathy is featured in the case reported by Jolliet *et al.* The focus was not on the rarity of the pathology, which was mixed connective tissue disease, but on the use of a nitric oxide inhalation challenge to assess the reversible component of the patient's pulmonary hypertension. Nitric oxide at 24 ppm for 10 minutes reduced pulmonary vascular resistance (PVR) by 14% and mean pulmonary artery pressure (PAP) by 21% (from 37 to 29 mm Hg) without any change in cardiac output or systemic blood pressure. In contrast, prostacyclin (maximum dose 6 ng/kg/min) or oral nifedipine (50 mg over two hours) lowered PAP by 16% and 10% respectively, and PVR by 50% and 20%, but at the cost of a 60% increase in cardiac output and a 9–17% fall in systemic blood pressure. Interestingly, Pao_2 fell (by 2·1–2·8 kPa) with nitric oxide and nifedipine but not with PGI_2. Nitric oxide is a useful agent for investigating the pulmonary circulation, especially as its effects wear off immediately.

In the last case report in this series Khan and Klapper remind us that Kaposi's sarcoma accompanies the immunosuppressed state from any cause, not just HIV disease. The incidence following renal transplantation is 400–500 times higher than in a control population; particular risk factors include treatment with cyclosporin and a non-white racial origin. The patient (a 32 year old black man) presented with persistent haemoptysis seven months after receiving a renal transplant for systemic hypertension. At bronchoscopy, haemorrhagic fluid was aspirated easily from several bronchial orifices. Bronchoalveolar lavage specimens were negative for opportunistic organisms, but the transbronchial biopsy showed spindle-shaped cells, which were von Willebrand antigen positive, compatible with Kaposi's sarcoma.

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