



Pathology Page

Pulmonary veno-occlusive disease



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A 57-year-old woman with end-stage renal disease received a kidney transplant using a kidney from a cadaver. The post-operative creatinine level gradually recovered from 9.6 mg/dL to 2.8 mg/dL. A kidney biopsy was performed 9 months post-transplantation because of worsening renal function and pathology revealed tacrolimus nephrotoxicity. Her immunosuppressant

was switched to sirolimus. Ten months after the biopsy, she was admitted to hospital because of pulmonary hypertension with a systolic pressure of 90 mmHg and right ventricle overload. She died of sepsis 4 days later. After autopsy, histopathology of the lungs confirmed changes in the pulmonary venous system with the lumina narrowed by extensive intimal fibrosis diagnostic of pulmonary veno-occlusive disease (PVOD; Fig. 1).

PVOD is currently classified as a subgroup of pulmonary artery hypertension (PAH) and accounts for 5–10% of cases initially considered to be idiopathic PAH. The cause of PVOD is not known. PVOD is associated with respiratory infections (especially virus infections), exposure to toxins, (chemotherapy), bone marrow transplantation, and renal transplantation. Only one case of PVOD following renal transplantation has been reported. Pathologists suggest the obstructive changes in the pulmonary veins are thrombotic in origin, such as in our case. PVOD is characterized by a poor prognosis and the possibility of developing severe pulmonary edema with specific PAH therapy. Lung transplantation is the treatment of choice.

Further reading

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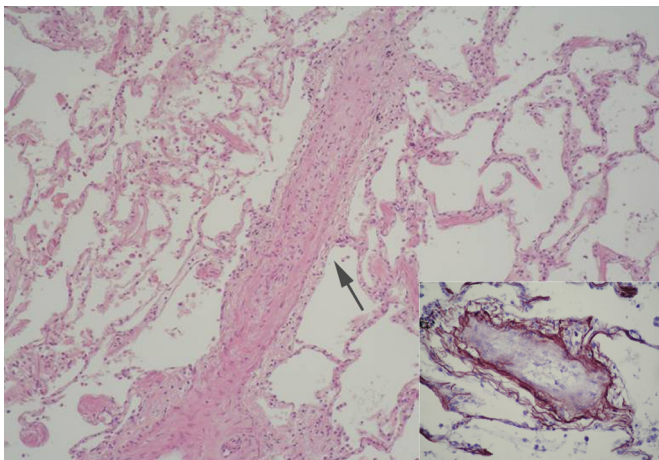


Fig. 1. Histopathology shows fibrotic tissue occluding the pulmonary venules in the septum (hematoxylin and eosin stain, magnification $\times 100$). Venous wall occlusion is demonstrated by an orcein stain (inset, orcein stain magnification $\times 200$).

Conflict of interest: none.

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