Adult Dominant Polycystic Kidney Disease and Atrial Myxoma

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Sir,

We wish to report a case of adult dominant polycystic kidney disease (ADPKD), in association with atrial myxoma and suggest that there may be a pathological link between the two conditions.

A 79-year-old lady presented with a 3-week history of palpitations, dizziness and transient weakness of the left side of the face. She had a family history of ADPKD. The main findings on examination was a completely irregular pulse of 130/min, blood pressure of 140/110, no evidence of heart failure or pulmonary oedema, bilateral palpable kidneys and a left homonymous hemianopia. Following digitisation and slowing of the heart rate auscultation revealed a mid-diastolic murmur, and a subsequent echo-cardiogram revealed the presence of a left atrial mass. The latter was removed uneventfully and proven to be a myxoma. The patient has since remained well and in sinus rhythm.

This patient presented with well-known features of atrial myxoma as documented in a report of a 50-year review of the condition [1]. The not-so-well-recognised association between ADPKD and cardiovascular lesions has also been recently highlighted [2, 3]. This association has led to the postulate that ADPKD is due to a primary genetic defect leading to a connective tissue abnormality. The mechanisms currently favored are production of an abnormal extracellular matrix or a circulating factor inducing cyst formation [4], with resultant increased compliance of tubular basement membrane.

The pathogenesis and nature of atrial myxoma are similarly uncertain, being thought of as either due to thrombus formation or a primary tumor. Numerous studies have not clearly defined the cell type involved, although evidence suggests they are fibroblasts in a myxoid stroma [5]. The arguments against thrombus has been made on the basis of site of the tumor, its occurrence in otherwise normal hearts and features of recurrence and embolism. All these points have been countered in the extensive study by Salyer et al. [6].

Patients with cardiac lesions and ADPKD, usually present with heart disease and mitral valve abnormalities are commonest, the available histology showing myxoid degeneration with loss and disruption of collagen [7]. The latter features are very similar to those found in Ehlers-Danlos and Marfan's syndrome. The occurrence of ADPKD and atrial myxoma has not been recorded before, but similarities in the histology of these lesions and the link with cardiovascular abnormalities suggests a common aetiology. Further definition of the various gene loci associated with ADPKD may clarify the basis of possibly this and other extrarenal manifestations.

References

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