Protean manifestations of Wilson's disease: a review of seven Saudi patients.

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Abstract

The clinical picture and laboratory data of five symptomatic and two asymptomatic cases of Wilson's disease in four unrelated Saudi families are reported. More than one member was affected in two families. The oldest and youngest patients were twenty and eight years respectively. The two oldest patients were siblings and presented with neurological disturbance only while the youngest two were unrelated and had predominantly hepatic involvement. A mixture of hepatic cirrhosis and extrapyramidal syndrome was encountered in one patient only and Kayser-Fleischer rings in three. The initial presentation of one patient who subsequently developed cirrhosis closely simulated subacute glomerular nephritis. Although only three patients showed clinical or biochemical evidence of liver disease either initially or in later stages, liver biopsy demonstrated a spectrum of morphological changes in all the seven cases. These findings indicate that a positive family history, Kayser-Fleischer rings and simultaneous hepatic and brain involvement need not be present and that subclinical hepatic involvement may be commoner than is generally realised in Wilson's disease. That seven cases were seen at one centre, which takes a largely unselected patient population, in a four-year period only, suggests that this preventable and treatable disease may be frequent in Saudi Arabia.