Letters

Focal nodular hyperplasia of the liver

Str.,—I wish to add to the reported association of focal nodular hyperplasia of the liver and intracranial pathology as described by Goldin and Rose (Gut 1990; 31: 554–5). The original pathological study by Albrecht et al.1 in Tübingen where it was found that in a postmortem series of 13 patients with multiple focal nodular hyperplasia all had associated lesions, often vascular, in the circle of Willis: including meningioma, astrocytoma, cerebral telangiectasia, and berry aneurysm.3

In 1984, a 30 year old woman who neither smoked nor drank alcohol presented with iron deficiency due to menorrhagia. It was noted that her liver function was normal save for gamma glutamyltransferase (GGT) 237 U/l (N<35). She next presented in 1988 with limbic epilepsy. Investigations with computed tomography of the head showed a right frontal astrocytoma (later confirmed at open brain biopsy in 1989). The GGT was 403 U/l but other liver function tests normal. She was treated with many different anticonvulsants which did not adequately control her epilepsy. In the summer of 1989 she underwent radiotherapy with subsequent improvement of the epilepsy. In November 1989 she complained of right upper abdominal pain and was noted to have 4 cm hepatomegaly. Ultrasound scanning of the liver showed multiple mixed density lesions mainly in the right lobe of the liver approximately 10 cm in diameter. Isotope scanning showed hepatomegaly but normal distribution of colloid uptake. Computed tomography showed several slightly hypodense lesions (largest being 12 cm) which enhanced after contrast injection (Figure). This combination of radiological findings is typical of focal nodular hyperplasia that diagnosis was confirmed by ultrasound guided biopsy of the liver. Biopsy specimens were also taken of adjacent but sonographically normal liver and this was found to be normal histologically.

No treatment has been given for the liver lesion and the clinical, biochemical, and radiological features have not changed over the ensuing six months. Her only medication during this time has been anticonvulsants (clobazam, phenytoin, carbamazepine). At no time was she taking an oral contraceptive drug. It is possible that between 1988 and 1990 the enzyme inducing effect of her various anticonvulsant drugs has influenced the size of her lesions. This case supports the suggestion of a new syndrome linking focal nodular hyperplasia with cerebral tumours and vascular malformations.

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A case of nodular regenerative hyperplasia of the liver, CREST syndrome, and primary biliary cirrhosis

Str.,—I took a special interest in the recent article by McMahon and colleagues. Moreover, I read with great interest a letter to the editor by Cadranel et al on the article and the reply.1,2 Cadranel et al claim that the patient reported was merely a new case (the fifth case in the published literature) of the association between CREST syndrome and nodular regenerative hyperplasia of the liver, because anti-mitochondrial antibody can be found in scleroderma, increased levels of serum IgM or alkaline phosphatase may not be in various disorders such as nodular regenerative hyperplasia of the liver, and because no histological features consistent with primary biliary cirrhosis were present. I do not intend to refute all of their ideas. However, I would like to comment on this reply of McMahon et al that the patient has also primary biliary cirrhosis for following reasons.

In the original paper by Ahrens et al on primary biliary cirrhosis, microscopic study of the liver had not been carried out in five of the 25 cases of primary biliary cirrhosis in the historical review nor in two of the 17 patients with primary biliary cirrhosis in their series. Furthermore, in Japan in the national survey, despite the lack of histological evidence suspected of having primary biliary cirrhosis due to positive antimitochondrial antibody, clinical features, and course were diagnosed as having primary biliary cirrhosis.2 Though the patient has not exhibited paresthesia, I would like to note that an increasing number of cases of primary biliary cirrhosis, including asymptomatic ones, have recently been detected, having "biochemical" and "immunological" findings characteristic for this disease. I expect that statistics of various laboratory data on nodular regenerative hyperplasia of the liver, such as antimitochondrial antibody IgM, alkaline phosphatase, globular proteins, serum levels of IgM, and alkaline phosphatase, will be more accurate, meaningful, and comparable if such cases of asymptomatic or biopsyneudirmed primary biliary cirrhosis are classified separately.

From the rheumatological point of view, I should like to make some comments. Firstly, it is important, from now on, to find out and to accumulate cases of nodular regenerative hyperplasia of the liver complicated with diffuse "sclerodera" as well as CREST syndrome—that is, limited cutaneous scleroderma. Secondly, to complete the report of this extremely rare case, I would like to have information on whether or not the patient has any evidence (clinical, immunological, or histological) of Sjögren’s syndrome, because the sicca syndrome often associated with primary sclerosis.