LETTERS TO THE EDITOR

Focal nodular hyperplasia of the liver

SIR,—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 proposal came from Albrecht *et al*[1 2] in Toronto 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 liver and brain including meningioma, astrocytoma, cerebral telangiectasis, and berry aneurysm.

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 glutamyltranspeptidase (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 hypertrophy[3] and 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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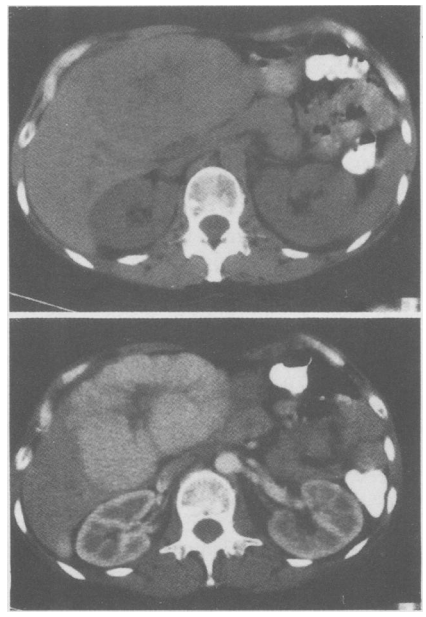
1 Albrecht S, Wanless IR, Bilbao J, Frei JV. Multiple focal nodular hyperplasia of the liver associated with vascular malformations and neoplasia of the brain and meninges: a new syndrome. *Lab Invest* 1989; **60**: 2A.
2 Albrecht S, Wanless IR, Bilbao J, *et al.* Multiple focal nodular hyperplasia of the liver associated with vascular malformations of various organs and neoplasia of the brain: a new syndrome. *Mod Pathol* 1989; **2**: 456-62.
3 Welch TJ, Sheedy PF, Johnson CM, *et al.* Focal nodular hyperplasia and hepatic adenoma: comparison of angiography, CT, US and scintigraphy. *Radiology* 1985; **15**: 593-5.



Top: Computed tomography of the liver showing hypodense lesions. Bottom: After injection of contrast.

Coeliac ganglionectomy

SIR,—We read with interest the paper by Johnson *et al* on 'Pancreatic exocrine responses to secretin, 2-deoxyglucose, a meal, and ethanol after coeliac ganglionectomy in the conscious dog' (*Gut* 1989; **30**: 1765–70). In the introduction to the paper the authors claim that 'There are no published data on the effect of stimulation or secretion of the splanchnic nerves in the conscious dog. . . .' This statement is wrong.

In 1986 we reported on the action of coeliac and superior mesenteric ganglionectomy alone or in combination with truncal vagotomy on the pancreatic secretory response to exogenous secretin in conscious dogs.[1] Johnson *et al* do not cite this investigation nor our other study in which we investigated the action of coeliac and superior mesenteric ganglionectomy on gastric and pancreatic responses to 2-deoxy-D-glucose in conscious dogs.

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1 Singer MV, Niebel W, Kniesburges S, Hoffmeister D, Goebell G. Action of atropine on the pancreatic secretory response to secretin before and after cutting the extrinsic nerves of the pancreas in dogs. *Gastroenterology* 1986; **90**: 355–61.
2 Becker S, Niebel W, Singer MV. Nervous control of gastric and pancreatic secretory response to 2-deoxy-D-glucose in the dog. *Digestion* 1988; **39**: 187–96.

Reply

SIR,—Thank you for the opportunity to reply to the letter of Professor Singer and colleagues. They draw attention to two reports from their group on the effects of coeliac ganglionectomy and superior mesenteric ganglionectomy on pancreatic secretory responses. The effects observed in those experiments were different from the effects seen in our experiments on the responses to secretin and 2-deoxyglucose. It is difficult to know how to interpret these differences because the extent of the denervation in the two experiments were different. We carried out coeliac ganglionectomy alone. There is, as far as I am aware, no report which compares the effect of coeliac ganglionectomy with coeliac ganglionectomy and superior mesenteric ganglionectomy. The differences between our work are interesting, but difficult to explain.

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

SIR,—I took a special interest in the recent article by McMahon and colleagues.[1] Moreover, I read with great interest a letter to the editor by Cadranel *et al* on the article and the reply.[2 3] Cadranel *et al* claim that the patient reported was merely a new case (the fifth case in the world literature) of the association between CREST syndrome and nodular regenerative hyperplasia of the liver, because antimitochondrial antibody can be found in scleroderma, increased levels of serum IgM or alkaline phosphatase may be noted 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 agree with the 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.[4] Furthermore, in Japan in the national survey, despite a lack of histological features, patients suspected of having primary biliary cirrhosis due to positive antimitochondrial antibody, clinical features, and course were diagnosed as having primary biliary cirrhosis.[5] Though the patient has not exhibited pruritus or jaundice, 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, and alkaline phosphatase, will be more accurate, meaningful, and comparable if such cases of asymptomatic or biopsy-unproved 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 scleroderma[6] 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