LETTERS TO THE EDITOR

Focal nodular hyperplasia of the liver

Sr.,—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 1989; 30: 554–5). The original paper by Albrecht et al. (1) in 1989 described the association where it was found that in a postmortem series of 13 patients with multiple focal nodular hyperplasia all had associated lesions, often vascular, in their cerebral brain including meningioma, astrocytoma, cerebral telangiectasis, and bery 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 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 but 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 (clofazin, 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 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, Freij JV. Multiple focal nodular hyperplasia of the liver associated with vascular malformations and neoplasia of the brain and meningi: a new syndrome. Lab Invest 1989; 60: 2A.


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

Sr.,—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 original literature) of both a CREST syndrome and nodular regenerative hyperplasia of the liver, because anti mitochondrial antibody can be found in scleroderma, increased levels of serum IgM or alpha-2-antiplasmin 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 like to add, in reply, 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 or in two of the 17 patients with primary biliary cirrhosis in their series. Furthermore, in Japan in the national survey, described a lack of histological evidence of having primary biliary cirrhosis due to positive anti mitochondrial antibody, clinical features, and course were diagnosed as having primary biliary cirrhosis. Though the patient has not exhibited positive antinuclear antibodies, 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 anti mitochondrial antibody, IgM and alpha-2-antiplasmin, may be more accurate, meaningful, and comparable if such cases of asymptomatic or biopsyunproved 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 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 Sjogren's syndrome, because the sicca syndrome often associated with primary


biliary cirrhosis is usually preceded by the diagnosis of primary biliary cirrhosis (89%) and is not correlated with the duration or severity of liver disease.\(^{14}\)

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**BOOK REVIEWS**


There is certainly no shortage of books about gastro-oesophageal reflux and it is reasonable to ask if we really need another one just now. I suppose it is better to have a lot of books about a common condition than about a rare one, but do we really want to read them all?

Gastro-oesophageal reflux is the commonest upper gastrointestinal disorder in most developed countries and there has been a pronounced improvement in our ability to make an accurate diagnosis and to treat what we find. The arrival of this latest offering from a very respectable author is timely in the light of new pharmaceutical developments and many gastroenterologists will already have received a free copy. As one would expect from the authors the book is well written, balanced in outlook, and extremely well referenced. Controversy has not been glossed over and the opinions of the contributors come over clearly. I certainly do not agree with every statement that has been made, but the book has the continuing interest of the subject and the justification for another book.

At £39.50 this is a good buy. If you got it for nothing I suspect you didn’t read it. Get it off the shelf and have a look. It wasn’t the usual useless freebie.


This is a good compact book in which Dundee and London surgeons combine to edit a multiauthor review of the state of the slow progress in the very difficult fields of bile ducts and pancreatic cancer. Fifteen of the 17 chapters concentrate on title topics while two chapters are included in the medical and surgical approaches to endocrine tumours of the pancreas.

There are currently few aspects on the research or therapeutic approaches to these notorious tumours which can be a cause for greater hope for the future of those unfortunate enough to suffer from these diseases. A theme throughout the book is to refer these patients to regional or national experts to minimise suffering, maximise effective remaining life, and occasionally achieve cure. Limited encouragement is presented in the initial improved palliation of cholangiocarcinoma using the iridium wire techniques. More hard data on this important subject would be valuable. Alfred Cuschieri emphasises the potential role of laparoscopy in making accurate diagnosis and minimalising the need for open laparotomy. Endoscopic and percutaneous stent placement in the management of obstructive jaundice, and the role of the specialist surgeon in resection are subjects well covered in a coherent manner.

Resection surgery alone as a treatment for cancer of the pancreas is usually inadequate and there is emphasis on the potential value of adding either chemotherapy or radiotherapy or both to the management of these difficult problems.

Prospects for improved treatment through greater knowledge of steroid receptors of the common cancer of pancreas have not yet materialised but the reviews by Swedish workers provide a window into an intriguing new area.

The challenge to produce monoclonal antibody therapy is also fascinating, but the abiding problem of the late presentation of advanced cancer so dominates this field that the current advances represent only a glimmer of improvement from 40 to 60 watts in a cavern of darkness the size of a football stadium.

This reviewer was impressed by the helpful detail in the last chapter on pain control, and this crucial area calls for dedicated professionalism in techniques of nerve block and ablation. The failure to match hospice and hospital pain control at home in the terminal stages of the disease remains an important challenge.


Patients who suffer from ‘functional’ disorders may find it difficult or even impossible to get an intelligible explanation and constructive advice from their physicians. This is less than surprising, given that many if not most physicians are not temperamentally equipped to deal with patients who have symptoms suggestive of pathology that appears to be absent.

In this situation, explanation and advice in book form is a potential remedy, potentially because of the dearth of suitable books. Now we have two books which aim to fill the gap.

The scope of these books differs in two ways. Grant Thompson covers a broad spectrum of disorders from globs to pteralgia fugax. Some might quarrel with the mixture of ill understood problems -- such as burblence and irritable bowel -- with syndromes of known pathophysiology such as gastro-oesophageal reflux. Geoff Watts has tried to insure himself against such quibbles with an all-embracing title, but if non-ulcer dyspepsia is in, why is duodenal ulcer disease out? I suppose you have to draw a line somewhere. In contrast, Geoff Watts has not confined his book to a single but common enough syndrome.

The second and major difference is in the training, orientation, and purpose of the authors. Grant Thompson is a physician who has made important contributions to our understanding of the field. His book is intended ‘... for patients, their relatives and those nurses, physicians, and allied health workers who may want to understand digestive symptoms better.’ Geoff Watts is a scientist by training and communicator by profession; he will be known to many readers as the presenter of ‘Medicine Now’ on BBC Radio. His book is aimed at helping patients to deal with the grips which divides disgruntled IBS sufferers from resentful physicians, Thompson is one of ‘us,’ while Watts, as he reveals at the conclusion of his book, is one of ‘them.’

Grant Thompson guides the reader with considerable skill through the maze of conflicting data, speculation, and mythology; his advice on diagnosis and management is wise, restrained, and sensitive. There is no better manual available for doctors, nurses, and dietitians. But for patients? Well, one has to allow for a difference in the health (or illness) cultures of North America and Britain, but...
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