Case report

A 26-year-old man was admitted to hospital with a five-day history of a mild influenza-like illness and sudden onset of three grand mal convulsions. He was found to have a mild fever, absent reflexes, and extensor plantar responses. The cerebrospinal fluid was normal. His fever subsided spontaneously after three days but he remained areflexive. A diagnosis of influenzal encephalitis was made, which was supported by virological and electroencephalographic (EEG) findings.

At that time he was found to have raised serum levels of aspartate aminotransferase (SGOT), lactate dehydrogenase (LDH), aldolase, and creatine phosphokinase (CPK). The CPK level was initially 7000 IU/l (normal $\leq$ 90 IU/l), and it remained raised when the patient became asymptomatic. The peripheral blood film showed acanthocytes. Other routine investigations gave normal results. He was treated with phenytoin 100 mg three times a day and discharged.

Eight months later he was readmitted after he had had two grand mal fits. Clinical examination showed the same abnormalities as before. The fever again subsided spontaneously and he became asymptomatic. SGOT, LDH, and aldolase levels were raised. The CPK was 2130 IU/l and was identified electrophoretically as the muscle isoenzyme. Serum lipoprotein concentrations were repeatedly normal. The EEG was abnormal with generalised slow-wave activity. An EMI scan and the CSF were normal. Electromyography showed evidence of a mild axonal neuropathy.

Routine blood film examinations showed that half of all red cells were acanthocytes. Incubation studies showed that the red-cell changes were not reversed in normal serum. Normal red cells became acanthocytes in the patient's serum. Red-cell osmotic fragility was normal but there was increased autohaemolysis, which was corrected by glucose.

The patient's daughter, mother, and brother were asymptomatic but had acanthocytes, absent reflexes, raised CPK levels, and normal fasting lipoproteins. His sister had normal reflexes, CPK and lipoprotein levels, and red cells. The patient was the only member of the family to have had convulsions.

Comment

The line of inheritance found in this man with normolipoproteinaemic acanthocytosis associated with neurological dysfunction and his family supports previous reports of autosomal dominance in this condition.[1]

Incubation studies with compatible normal red cells in the affected sera showed the presence of an abnormal acanthocyte-inducing component. The patient's red cells remained abnormal in compatible normal sera. This may be interpreted as either an intrinsic defect of the membrane itself or an irreversible morphological change induced by the serum factor. Previous reports[2] support the latter hypothesis.

The abnormal electromyogram and serum enzyme levels can be explained if similar damage is caused to the membranes of other cells. Previous reports have described raised serum enzyme levels, but in only one case was a persistently raised CPK level documented.[2] In the family reported here all the affected members had persistently raised serum CPK levels.

In contrast to other families,[1-4] who showed fairly severe neurological abnormalities, this family showed an asymptomatic neurological disorder. The epileptic fits in our patient may have been causally related to a viral encephalitis or they might have been part of the neurological syndrome associated with the acanthocytosis.

A spectrum of disease seems to exist in patients with normolipoproteinaemic acanthocytosis. CPK estimation may help to determine the prognosis.

We thank Dr E C Hutchinson for permission to report this case, and Dr E Butterworth for his help.

[1] Levine, I M, *et al*, *Archives of Neurology*, 1968, **19**, 403.
[2] Aminoff, M J, *Brain*, 1972, **95**, 749.
[3] Critchley, E M R, *Postgraduate Medical Journal*, 1970, **46**, 698.
[4] Mars, H, *et al*, *American Journal of Medicine*, 1969, **46**, 886.
[5] *British Medical Journal*, 1971, **1**, 683.

(*Accepted 18 March 1977*)

Department of Haematology, Central Pathology Laboratory, Stoke-on-Trent

DAVID LOWE, MB, BS, senior house officer in haematology
R M IBBOTSON, MRCP, MRCPATH, consultant haematologist

Department of Neurology, North Staffordshire Royal Infirmary, Stoke-on-Trent

L A LOIZOU, MRCP, PHD, registrar in neurology

Transjugular liver biopsy

The risks of percutaneous needle biopsy of the liver are increased when there is impaired coagulation or thrombocytopenia, both of which are common in patients with severe liver disease. This risk of bleeding may be eliminated by taking the biopsy specimen through a hepatic vein, entering from the right internal jugular vein, an approach described by Rösch *et al.*[1 2] We report our experience of this unorthodox approach.

Method and results

Patients—All 26 patients had clinical and biochemical evidence of liver disease. Cirrhosis was suspected in 14, of whom nine gave a history of prolonged heavy alcohol intake. The indications for a transjugular biopsy were: prothrombin time ratio (PTR) > 1·3 (12); platelet count < 80×10^9/l (5); prlonged PTR and thrombocytopenia (4); pressure measurements or venography also required (2); failed percutaneous biopsy (2); extrahepatic biliary obstruction (1).

Technique—The procedure is performed in an intensive therapy unit where an *x*-ray image intensifier and ECG monitor may be used. When possible the patient is sedated with intravenous diazepam. A 9 F catheter is introduced through the right internal jugular vein[3] into the right atrium and directed under fluoroscopic control into a hepatic vein. Its position is confirmed by injection of contrast medium. The 16 G 55-cm pre-curved transjugular needle, a modified Ross trans-septal needle (William Cook, Europe A/S, Hitchin, Herts), connected to a syringe of saline, is advanced within the catheter into the hepatic vein. The catheter is flushed with saline from the syringe, advanced to a wedged position, and its position confirmed with the image intensifier. The needle is then rapidly advanced 2 cm beyond the catheter tip and the biopsy specimen retained within the needle by gentle suction on the syringe. The needle remains in the liver parenchyma for about one second as when using the Menghini percutaneous biopsy needle. The free and wedged hepatic vein pressures, relative to the sternal angle, are also routinely measured in several branches of the hepatic vein using a 7 F Cournand catheter.

Histological findings—Biopsy specimens were obtained in all 26 patients and measured 4–12 mm × 1 mm. The specimen was adequate for full histological assessment in 16 of these (see table).

Histological findings

Diagnostic biopsies	
HBsAg-positive active cirrhosis	2
Cirrhosis and alcoholic hepatitis	3
Micronodular cirrhosis	2
Active chronic hepatitis	2
Alcoholic hepatitis	1
Postanaesthetic hepatitis	1
Granulomatous hepatitis	1
Leukaemic infiltration	1
Venous congestion	1
Non-specific minor inflammatory changes	2
Non-diagnostic biopsies	
Suggestive of cirrhosis	5
Suggestive of alcoholic hepatitis with or without cirrhosis	3
No diagnostic features	2

Complications—No patient showed any clinical signs of blood loss after the procedure. One patient with heart disease developed transient atrial fibrillation during the procedure. None complained of undue discomfort during or after the investigation, and there were no local complications in the neck.

Comment

Although 21 patients had an increased bleeding tendency, there were no serious complications in this, or other, series. Additional information is readily obtained during the procedure: if thrombus is suspected, venography of the hepatic veins and inferior vena cava may be performed; and portal hypertension may be detected by measuring pressures in the hepatic vein in the free and wedged positions. It may be the easier biopsy route in the presence of ascites, and may be safer in semiconscious or anxious patients as repiratory co-operation is unnecessary. In one patient the procedure was performed uneventfully through the left internal jugular vein, as the right side could not be cannulated.

The major drawback of this technique is the size of the biopsy specimens obtained: in 10 patients these were inadequate for full histological assessment. All but two of these, however, had cirrhosis,

Br Med J: first published as 10.1136/bmj.2.6079.100 on 9 July 1977. Downloaded from http://www.bmj.com/ on 17 April 2024 by guest. Protected by copyright.

a condition in which the results of Menghini percutaneous biopsies are also often unsatisfactory. Specimens may be improved by incorporating a stilette within the needle to prevent possible aspiration and fragmentation of the specimen in the syringe.

In summary, while this technique should be attempted only by those experienced in catheterisation and cannulation of the internal jugular vein, transjugular liver biopsy is valuable when percutaneous biopsy is hazardous, or when pressure measurements or venography are also required.

We are grateful to Miss V Arnold for her technical help. ITG is in receipt of a Medical Research Council Training Fellowship.

[1] Rösch, J, *et al*, *New England Journal of Medicine*, 1973, **289**, 227.
[2] Rösch, J, Antonovic, R, and Dotter, C T, *American Journal of Roentgenology, Radium Therapy, and Nuclear Medicine*, 1975, **125**, 602.
[3] Branthwaite, M A, and Bradley, R D, *Journal of Applied Physiology*, 1968, **24**, 434.
[4] Menghini, G, *Gastroenterology*, 1958, **35**, 190.

(*Accepted 23 March 1977*)

St Thomas's Hospital, London SE1 7EH

I T GILMORE, MRCP, research fellow, gastrointestinal laboratory, Rayne Institute, and honorary senior registrar in medicine
R D BRADLEY, BSC, MB, consultant physician, department of clinical physiology
R P H THOMPSON, DM, MRCP, consultant physician

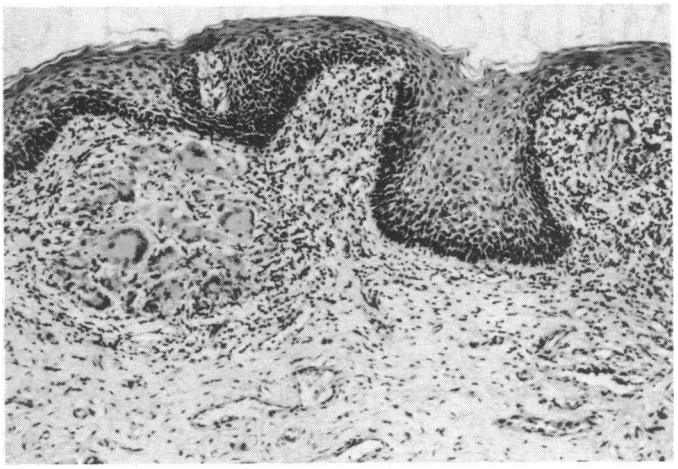
Crohn's disease: an unusual cause of dyspareunia

The commonest dermatological manifestation of Crohn's disease is direct extension of the disease from the affected bowel, such as perianal lesions or extension on to the anterior abdominal wall after surgical treatment. A much rarer and only recently recognised complication is metastatic Crohn's disease, in which skin lesions occur in a site distant from the gastrointestinal disease. Disease of the groins, male genitalia, submammary region, and postauricular area have been reported.[1-3] The present case describes vulval lesions and resulted in gynaecological referral.

Case report

A 45-year-old woman was referred from another hospital for further investigation of diarrhoea. Her major complaints, however, were of persistent vulval soreness and dyspareunia, for which she had attended gynaecological outpatients for three years, having had dilatation and curettage twice and treatment for one proved monilial infection without any improvement. Her gastrointestinal complaints had lasted four years, being diarrhoea with the passage of 3-4 loose explosive stools daily. She had a 15-year history of recurrent iritis and a 2-year one of recurrent oral ulceration and arthropathy of the knees, hips, and elbows. She was a fit woman in whom the only abnormal physical finding was an area of small reddened nodules with surrounding erythema on the right labia majoras with no other perianal or perineal lesions.

Although the result of a barium enema was normal, a diagnosis of Crohn's disease with total disease of the large bowel was established by sigmoidoscopy and colonoscopy with histological evidence from rectal biopsy specimens. A barium follow-through showed ileal narrowing consistent with Crohn's disease and *x*-ray films of the sacroiliac joints confirmed the presence of sacroileitis. A biopsy specimen of the vulva showed very similar changes to the rectal biopsy specimen with numerous granulomata (figure).



Giant-cell granulomas and mononuclear inflammation beneath vulval epidermis. (Haematoxylin and eosin. ×100.)

Her gastrointestinal symptoms failed to improve with simple symptomatic measures but responded rapidly to oral steroid treatment. Nevertheless, there was no change in her vulval symptoms or lesion after treatment with oral, local, or intradermal steroids.

Discussion

Parks, Morson, and Pegum[4] first drew attention to metastatic Crohn's disease and described it as cutaneous disease occurring remote from the gastrointestinal tract and separated from it by normal skin. Only six subsequent cases have been reported and, although there is no explanation for this cutaneous lesion, Mountain[2] noted that it occurred where skin surfaces were in close apposition and tended to be moist. Vulval disease, which has not been described, would also fit this association.

Although her gastrointestinal symptoms resolved completely with oral steroids, she still complained bitterly of dyspareunia. As cutaneous sarcoid lesions occasionally respond to intradermal steroids, we tried these in this patient, but there has been no symptomatic or objective improvement.

[1] McCallum, D I, and Kinmont, P D C, *British Journal of Dermatology*, 1968, **80**, 1.
[2] Mountain, J C, *Gut*, 1970, **11**, 18.
[3] McCallum, D I, and Gray, W M, *British Journal of Dermatology*, 1976, **95**, 551.
[4] Parks, A G, Morson, B C, and Pegum, J S, in *Proceedings of the Royal Society of Medicine*, 1965, **58**, 241.

(*Accepted 23 March 1977*)

University Departments of Medicine and Histopathology, Bristol Royal Infirmary, Bristol BS2 8HW

P A HAMILTON, BSC, MB, house physician
P BROWN, MB, MRCP, lecturer in medicine
J D DAVIES, MD, MRCPATH, consultant and senior lecturer in pathology
P R SALMON, MB, MRCP, consultant senior lecturer in medicine

Princess Margaret Hospital, Swindon
K D CROW, FRCP, consultant dermatologist

Br Med J: first published as 10.1136/bmj.2.6079.100 on 9 July 1977. Downloaded from http://www.bmj.com/ on 17 April 2024 by guest. Protected by copyright.