Filling defect in lower pole of caecum.

laparotomy mandatory. A caecal diaphragm must therefore become part of the differential diagnosis of a space-occupying lesion of the lower pole of the caecum found by radiology. The chance of diagnosing correctly a caecal diaphragm by radiology alone must be remote, but if at laparotomy for suspected pathology of the pole of the caecum faeces are palpated in the caecal pole and attempts to move them into the ascending colon fail then the presence of a caecal diaphragm must be seriously considered.

(Accepted 18 March 1977)

Bradford Royal Infirmary, West Yorkshire

A D GAYNER, FRCS, registrar

J J PRICE, FRCS, consultant surgeon

O J FOLLOWS, MRCP, FRCP, consultant radiologist

Giant-cell arteritis and hypothyroidism

The aetiology of giant-cell arteritis is unknown, though there is increasing evidence to suggest an immunological basis in which the arterial elastic tissue acts as an antigen.¹ * An association of giant-cell arteritis with currently well-recognised autoimmune disorders would therefore be of interest and might provide indirect supporting evidence of an underlying abnormal immune host response. The simultaneous presentation of giant-cell arteritis and autoimmune thyroiditis with hypothyroidism has not previously been reported.

Case report

A 73-year-old woman was admitted to hospital with a 12-month history of lethargy and depression for which antidepressants had been prescribed. During the previous six weeks she had also complained of intolerance to cold, acroparaesthesiae, and bitemporal headaches. Relatives had noticed her to be slow and reported the recent appearance of puffiness around the eyes and hoarseness. She denied any visual disturbance, muscle pain, or joint stiffness and there was no history of endocrine disease.

On examination she looked myxoedematous with dry skin and periorbital puffiness. The thyroid was moderately enlarged and the relaxation phase of ankle jerks was slow. Both superficial temporal arteries were clinically normal, pulsatile, and non-tender. There were no abnormal vascular bruits, and all

peripheral pulses were present. The locomotor system showed no abnormality. Results of investigations were: haemoglobin 9.9 g/dl, erythrocyte sedimentation rate (ESR) 115 mm in first hour, protein-bound iodine (PBI) 250 nmol/l (3.2 µg/100 ml) (normal 315–630 nmol/l (4.0–8.0 µg/100 ml)), and four-hour thyroidal uptake of ¹²⁵I 5.4% (normal 10–35%). Anticytoplasmic thyroid antibody titre was strongly positive and antithyroglobulin antibody titre positive at 1/2.5 × 10.⁶ Antigastric antibody titre was negative, and serum vitamin B₁₂ and folate concentrations were within the normal ranges. Biopsy of a right superficial temporal artery showed the characteristic changes of giant-cell arteritis.

The patient was treated with oral prednisolone 40 mg daily and at the same time began thyroxine replacement therapy 50 µg daily, later gradually increased to 200 µg daily. Within one month she was entirely asymptomatic; the haemoglobin had risen to 12.1 g/dl and the ESR fallen to 33 mm in the first hour. We later reduced the dose of prednisolone with no untoward effects. At 18 months follow-up she remained well and euthyroid and was taking 2.5 mg prednisolone and 200 µg thyroxine daily.

Comment

This patient presented with the clinical features of hypothyroidism, and the diagnosis was confirmed by the findings of a goitre, strongly positive antithyroid antibody titres, low PBI, and subnormal thyroidal radioiodine uptake. The coexistence of giant-cell arteritis was suspected from the history of temporal headaches of recent onset, and, although the temporal arteries were clinically normal, the histological features on biopsy were typical of giant-cell arteritis. The protean manifestations of hypothyroidism and giant-cell arteritis are well known, and this case illustrates the importance of recognising their possible coexistence in elderly patients, as both conditions are eminently treatable.

The simultaneous presentation of giant-cell arteritis and hypothyroidism has not previously been reported. Polymyalgia rheumatica, however, which is closely related to giant-cell arteritis, was found to co-exist with thyroiditis and hypothyroidism in an elderly woman.³ In addition, Niarchos and Finn⁴ described an association between hypothyroidism and abdominal aneurysm and speculated that the arterial lesions were related to an underlying immunological mechanism. In a retrospective survey, Thomas and Croft⁵ reported a higher incidence of thyrotoxicosis but not of hypothyroidism in a group of women with giant-cell arteritis than in a control group.

The coexistence of giant-cell arteritis and autoimmune thyroiditis with hypothyroidism suggests a common cause, and further studies may contribute to our understanding of the pathogenesis of giant-cell arteritis.

¹ Mowat, A G, and Hazleman, B L, *Journal of Rheumatology*, 1974, 1, 190.

² Liang, G C, Simkin, P A, and Mannik, M, *Annals of Internal Medicine*, 1974, 81, 19.

³ Fauchald, P, Rygvold, O, and Øystese, B, *Annals of Internal Medicine*, 1972, 77, 845.

⁴ Niarchos, A P, and Finn, R, *British Medical Journal*, 1973, 4, 110.

⁵ Thomas, R D, and Croft, D N, *British Medical Journal*, 1974, 2, 408.

(Accepted 18 March 1977)

University Department of Therapeutics and Clinical Pharmacology, Aberdeen AB9 2ZD

J HOW, MB, MRCP, lecturer

P D BEWSHER, MD, FRCP, senior lecturer

W WALKER, MA, FRCP, regius professor

Benign normolipoproteinaemic acanthocytosis

Acanthocytes, or spiky red cells, in neurological disorders have been described in association with normal serum lipoproteins,¹⁻³ hypobetalipoproteinaemia,⁴ and abetalipoproteinaemia.⁵ These cells are morphologically indistinguishable from "burr" cells, or "spur" cells in American reports (*acanthos*, thorn).⁶ The third family in the United Kingdom with normolipoproteinaemic acanthocytosis is described here.

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 influenza 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 ≤ 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 normolipoprotein-aemic 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 reports² 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 normolipoprotein-aemic 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 Röscher *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 $< 80 \times 10^9/l$ (5); prolonged 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 vein³ 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 $\times$ 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 respiratory 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,