Histology: Liver: early cirrhosis; it was not possible to tell whether this was alcoholic or cryptogenic. Spleen: weight 900 g; there was hyperplasia of the red pulp with some erythrophagocytosis and extramedullary haemopoiesis.

Discussion

The presenting feature of gout may have been related to the increased marrow proliferation. Gout is well documented in myeloproliferative disorders and in chronic haemolysis. In one series of patients with gout (Grahame & Scott 1971), 4.2% had an underlying haematological disorder. Since splenectomy, the serum uric acid level has fallen from 0.60 mmol/l to 0.50 mmol/l. Pancytopenia was due to sequestration of blood, and especially neutrophils, in an enlarged spleen. Evidence for splenic neutrophil sequestration is usually presumptive, being based on the results of a ^{99}Technetium-labelled red-cell scan, but in this case a direct estimation was made with an indium-labelled granulocyte scan. This technique makes use of a method for binding ^{111}In to cells, developed by Dr M Thakur (unpublished).

The aetiology of the cirrhosis, and its relationship to the splenomegaly and neutropenia, is uncertain. Alcoholic cirrhosis with portal hypertension is a possibility, but we think it is unlikely. No evidence of a collateral circulation to the portal system was seen at laparotomy, though this does not exclude portal hypertension (Turner *et al.* 1957). The spleen size would be unusual in portal hypertension due to cirrhosis alone. In two large series of cirrhotic patients the average spleen size was only 300 g, whereas in this case the spleen weighed 900 g (Kelynack 1897, Rolleston & Macnee 1929).

Splenomegaly secondary to cirrhosis usually produces mild pancytopenia, but occasionally a predominant neutropenia is found (Sherlock 1968). By contrast, in immune disorders with splenomegaly, such as Felty's syndrome, predominant neutropenia is typical.

ANA is rarely found in alcoholic cirrhosis, but is frequently present in cryptogenic cirrhosis. In one series of patients with cryptogenic cirrhosis, ANA was present in 38% of patients, but in only 3 out of 14 males. In only 2 patients was the titre as high as 400 units (Doniach *et al.* 1966).

We suggest that the high ANA titre may be a marker of an autoimmune process, of which the cirrhosis and neutropenia are separate manifestations. This supposition is supported by the subsequent appearance of circulating immune complexes, as demonstrated by the diffuse paraprotein band.

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Polyarteritis nodosa presenting with bilateral nerve deafness[1]

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In February 1976, one week after a 'chill', Mrs A G aged 63 suddenly developed tinnitus and profound bilateral deafness. In May she noticed painless redness of the eyes. She was examined at Moorfields Eye Hospital where old central keratitis and active marginal keratitis were

[1] Case presented to Clinical Section, 11 February 1977

diagnosed. In June she became aware of mild numbness in the soles of the feet, and of muscle pains mainly in the lower legs, neck and around the shoulders.

When she was 5 years of age tuberculous infection had occurred, involving both hip joints and resulting in severe arthritic damage. There was no evidence that active infection had recurred and she had remained well as an adult until the onset of the present illness. She had not received any medication in recent months.

At the time of admission to hospital on 23 June 1976, tachycardia and a low grade intermittent fever were present; blood pressure was 140/80; there was bilateral deafness. Audiograms (Figure 1) indicated bilateral sensorineural deafness; the results of electrocochleography pointed to end-organ impairment.

Investigations: Hb, 9.6 g/dl; WBC, 11.7×10^9/l; eosinophils, 8%; ESR, 130 mm/h (Westergren); serum B_{12} 150 ng/ml; blood cultures negative cerebrospinal fluid normal; electrolytes normal; urea 51 mg/100 ml (8.6 mmol/l); creatinine 0.8 mg/100 ml (70.5 μmol/l); urine albumin positive; chest X-ray, evidence of old tuberculosis; Mantoux test negative 1:10 000; Australia antigen negative; autoimmune complement fixation complete at 1:4; liver kidney microsomal antibody positive at 1:80; antinuclear factor negative; Rh factor negative. Gamma globulins: normal IgG, raised IgM and IgA. Complement: raised C3, normal C4.

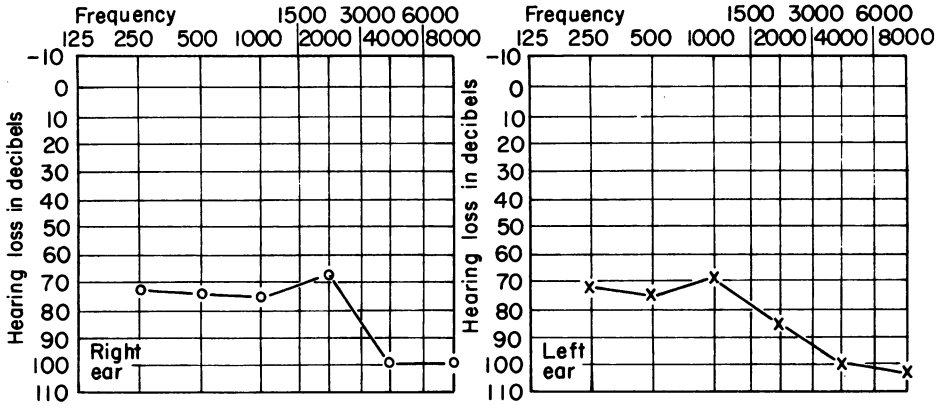

Figure 1. Audiograms on admission

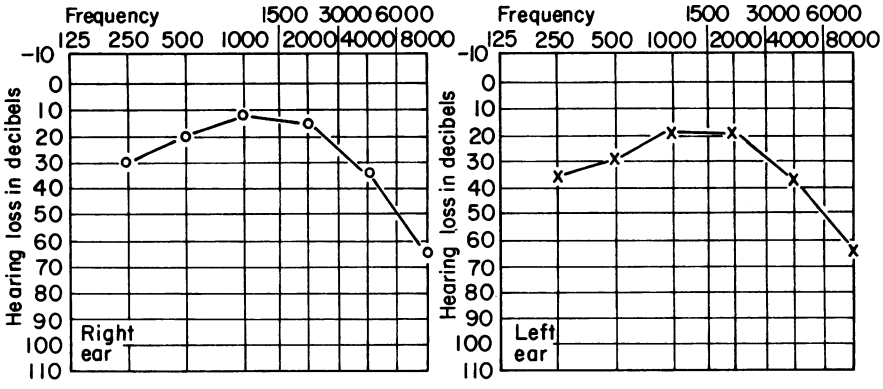

Figure 2. Audiograms following spontaneous return of hearing

Raised alkaline phosphatase (233 iu/l) and aspartate aminotransferase (130 iu/l). Liver biopsy normal.

Course and progress

Four days after admission, sensory impairment in an ulnar nerve distribution was noted on the left and, a day later, similar changes occurred in the right hand. During the next two weeks asymmetrical weakness and sensory disturbance occurred in both upper and lower limbs. Right wrist and left foot drop occurred suddenly, within four days of each other. Apart from auditory nerve involvement, there was no impairment of cranial nerves.

Muscle tenderness was present. Biopsy from the left biceps showed inflammatory myopathy and arteriolar inflammation consistent with polyarteritis nodosa. Electromyography and nerve conduction studies showed changes consistent with proximal myopathy and mononeuritis multiplex.

On 4 July, before treatment with prednisolone had been started, the patient noticed spontaneous improvement in her hearing, to an extent that she was able to hear normal conversation. Subsequent audiograms (Figure 2) showed that her hearing, by then, had returned virtually to normal.

Treatment with oral prednisolone, initially 100 mg daily for three days and then in reduced dosage, was started on 9 July; azathioprine 2.5 mg/kg was added later. The patient developed mild hypertension which was controlled by propranolol and bendrofluazide. The urine became free from protein. The ocular symptoms did not recur. Some neurological impairment persisted with distal asymmetrical weakness and wasting in the upper limbs and patchy sensory impairment in both arms and legs. There has been no recurrence of deafness.

Comment

The occurrence of deafness during the course of polyarteritis nodosa is unusual and has rarely been reported. Bernstein (1935) described increasing bilateral nerve deafness in a 44-year-old man in the course of polyarteritis nodosa. A few other authors have described similar cases (Middleton & McCarter 1935, Spiegel 1936, McNeil *et al.* 1952). Among 300 patients with polyarteritis nodosa reviewed by Foster & Malamud (1941), only 2 developed nerve deafness.

Cogan (1945, 1948) described a syndrome of nonsyphilitic interstitial keratitis with vestibuloauditory symptoms occurring in young adults. Bellucci *et al.* (1974) reported two patients with Cogan's syndrome who also developed bilateral nerve deafness. In certain patients with the features of Cogan's syndrome there is evidence of a systemic disorder, and a few have subsequently developed manifestations of polyarteritis nodosa (Cody & Williams 1960, Oliner *et al.* 1953, Boyd 1957, Crawford 1957). However, in the majority of cases the aetiology is obscure.

We have been able to find only one other reported case of a patient presenting with sensorineural hearing loss as the initial feature of polyarteritis nodosa (Peitersen & Carlsen 1966). In that case the patient improved after starting steroid treatment; in our case, recovery took place spontaneously, even before steroid treatment was started. Peitersen & Carlsen (1966) also described a similar presentation in Wegener's granulomatosis.

Our patient provides the first reported example of severe deafness occurring as the initial sympton of polyarteritis nodosa, with spontaneous improvement in the sensorineural impairment before starting steroid treatment.

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Systemic lupus erythematosus in a patient with Hashimoto's thyroiditis and pernicious anaemia[1]

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Mrs M Z, now aged 68 years, presented in 1964 at the age of 55 with a history of intermittent pain and swelling in her hands, and with pain in her elbows and wrists since 1944. She had felt cold, tired and constipated and admitted to dry hair and skin and a deep voice since 1957. A clinical diagnosis of Hashimoto's thyroiditis was supported by the following investigations: protein-bound iodine 1.9 g/100 ml; I^{131} thyroid uptake in 24 hours of 13% (normal 22–50%) with no increase in iodine uptake following thyroid-stimulating hormone administration, and thyroid tanned-cell agglutination titre 1/3200; electrocardiogram showed bradycardia, T-wave inversion and low voltage. Although the haemoglobin was 13.4 g/100 ml, there was macrocytosis present and the serum B_{12} was 130 pg/ml (normal 140–900 pg/ml). Histamine test meal revealed complete achlorhydria. A Schilling test was performed, 8.5% of oral dose was excreted in 24 hours. Immunofluorescent antibodies to gastric parietal cells were strongly positive and the complement fixation titre was 1/128. Antibodies to intrinsic factor were positive on one occasion and negative on another. A diagnosis of pernicious anaemia was made. In 1964, she had no active rheumatoid disease; erythrocyte sedimentation rate (ESR) was 12 mm/h (Westergren) and the latex agglutination titre was negative. She was started on oral thyroxine and vitamin B_{12} injections.

In 1969 the patient developed pain and swelling of the metacarpophalangeal joints of both hands. On examination these joints were hot, tender and swollen, and there was swelling of the proximal interphalangeal joints with Heberden's nodes. The radiographic appearances were those of osteoarthritis superimposed on a primary rheumatoid process. Latex agglutination titre and sheep cell agglutination titre were both negative, and the ESR was 18 mm/h. In January 1975, at the age of 66, the patient presented with a history of diarrhoea for eighteen weeks, and swelling of her tongue and face after starting neomycin.

On examination: The patient was acidotic, pyrexial and dehydrated, with a tachycardia (120/min) and a blood pressure 70/50 mmHg. She had a dry skin, large tongue, angioneurotic oedema and an urticarial rash on her face. During the next few weeks she developed pleurisy and diffuse abdominal pain.

Investigations: On admission she was anaemic (Hb, 10 g/100 ml) and the ESR was 117 mm/h. Chest X-rays showed fluctuating areas of atelectasis and a pleural effusion which was tapped on one occasion. A diagnosis of senile systemic lupus erythematosus was made and confirmed by the presence of lupus erythematous cells and antinuclear antibody (greater than 1600 units

[1] Case presented to Clinical Section, 11 February 1977
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