patients in both relapse and remission were upper respiratory tract infections (URTI). Other factors associated with relapse were stress (2), *Escherichia coli* urinary tract infection (1), and gastroenteritis (1).

For patients with CD, 18 relapses were observed during the 61 clinic attendances. Of these 18 relapses, 13 were associated with a precipitating factor (table). Of 43 attendances of patients with quiescent disease, only 10 were associated with a precipitating factor ($P < 0.0005$). Most of the precipitating factors were again URTIs. One patient who was in relapse had begun divorce proceedings in the previous month.

Even when upper respiratory tract infections were considered alone, patients with a relapse of either UC or CD were significantly more likely to have had an infection in the previous month than those in remission (UC relapse *v* remission $P < 0.01$, CD relapse *v* remission $P < 0.005$). No patient with an URTI had received an antibiotic during the previous month.

Comment

The present study has shown that there is a highly significant association between URTIs and a relapse of either UC or CD. Thus 60% of patients with UC with a relapse had had an URTI during the previous month compared with 22.7% of patients who presented in remission. A similar relationship was present in patients with CD. Of the other precipitating factors investigated, none appeared with sufficient frequency to enable meaningful results to be obtained.

The reasons why URTIs should be associated with a relapse of either UC or CD are uncertain. One hypothesis is that circulating immune complexes resulting from the infection may be deposited in the diseased intestine. In rabbits deposition of immune complexes in the colon may cause colitis.^{3,4} Since animals infected with upper respiratory tract viruses may have circulating complexes,⁵ similar complexes occurring in human URTI may localise to the diseased bowel and lead to a relapse.

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Presumed ataxia-telangiectasia in a man

Ataxia-telangiectasia is a complex, multisystem genetic disorder.^{1,2} It is usually fatal from uncontrolled infection in adolescence, though it shows some variability in severity, age of onset, and rate of progression. We report an unusual case of the condition, resembling Friedreich's ataxia, in a man.

Case report

The patient is a 37-year-old Italian lawyer with two children, who has been diabetic from the age of 35. His parents were first cousins, and there had been consanguineous marriages in previous generations. From infancy he had had recurrent infective diarrhoea and subsequently recurrent respiratory and urinary infections. He had had two operations for hypospadias in infancy, but his early development had otherwise been normal. Neurological symptoms began in his late teens, with weakness and wasting beginning distally in legs, later spreading proximally. Progressive ataxia developed later, though on 30 October 1977 he still walked unaided. He complained of "hypersensitivity" of the skin of the feet and legs.

His paternal grandfather and great-grandfather had had progressive walking difficulty and required wheelchairs in old age. In 1976 his father, though asymptomatic, had impaired proprioceptive sensation in the legs and an absent left ankle jerk.

The patient had telangiectases in both popliteal fossae, gonadal atrophy, bilateral pes cavus, and normal intellect. He had fine, phasic horizontal nystagmus on lateral gaze bilaterally; the eye movements were otherwise normal, as were the remaining cranial nerves. There were no bulbar conjunctival lesions. The arms showed symmetrical mild intention tremor and

diminished tendon reflexes; the legs showed symmetrical weakness and wasting, mainly distally. Posterior column sensation was absent and spinothalamic sensation impaired in both legs below the knees; knee jerks were present, ankle jerks absent, plantar responses flexor. His gait was ataxic and Romberg's sign positive. There was no peripheral nerve hypertrophy.

Serum IgA was undetectable on repeated examination; IgG 12.45 g/l (normal 11.4 ± 1.7 g/l) and IgM 2.1 g/l (normal 1.0 ± 0.4 g/l). Serum α -fetoprotein 36 MRC units/ml (raised). Hb 13.4 g/dl, normal red cell indices and morphology (no acanthocytes); WBC $15.8 \times 10^9/l$, 86% polymorphs; platelets normal; ESR (Westergren) 108 mm in first hour. Blood concentrations of urea, creatinine and electrolytes, bilirubin, aminotransferases, alkaline phosphatase, and fasting calcium and phosphate were normal; fasting blood sugar 6.4 mmol/l; VDRL negative; serum folate $1.8 \mu g/l$, $B_{12} > 1000$ ng/l. Fasting blood cholesterol and serum triglyceride concentrations were normal, as was lipoprotein electrophoresis. Urine contained 1 pus cell high-power field, no growth on culture. Chest x-ray film showed lingular consolidation; skull x-ray film was normal; electrocardiogram was normal.

Results of clinical neurophysiological studies are given in the table. Electromyography (concentric needle electrode) showed no spontaneous activity and no motor units under volitional control in the right extensor digitorum brevis. In the right tibialis anterior there were fibrillation potentials, positive sharp waves, and a reduced interference pattern on volition; motor units were of normal configuration but increased duration. The findings indicated a mixed motor/sensory axonal neuropathy. A gastrocnemius muscle biopsy done in 1976 in Italy showed non-specific changes.

Results of clinical neurophysiological investigations

		Terminal latency (ms)	Evoked potential duration (ms)	Evoked potential amplitude (μV)	Conduction velocity (m/s)
Motor	L median	4.3	15.4	7000	64.1
	R ulnar	2.7	15.9	6500	50
	R lat popliteal	9.8	4.4	250	35.8
	R digit II	3.2	1.6	0.8	46.9
Sensory	R digit V	1.9	1.8	2.0	65.8
	R sural	..	..	..	..

Sensory nerve action potential not obtainable

Comment

In our view the telangiectasia, deficiency of IgA, and raised alpha-fetoprotein concentration confirm the diagnosis of ataxia-telangiectasia. The onset of neurological symptoms was late, the course relatively benign, and the predominance of the peripheral neuropathy unusual. Cases have been reported with survival into the fifth decade,³ presumably owing to the increasingly effective treatment of infection. Evidence of cerebellar dysfunction in our patient was slight, but in one report⁴ cerebellar dysfunction was minimal at the age of 34. Usually cerebellar ataxia is the predominant neurological manifestation, beginning in childhood. Nevertheless, peripheral neuropathy⁵ certainly occurs in this condition and in our patient was the initial and predominant neurological manifestation.

The nature of the disorder present in previous generations is unknown but the pattern suggests pseudo-dominant inheritance due to multiple consanguinity since ataxia-telangiectasia is usually due to an autosomal recessive gene. Though our patient and his relatives possibly suffered from a previously unreported hereditary axonal neuropathy associated with telangiectasia, immune deficiency, and a raised serum α -fetoprotein concentration, on balance we believe that it was probably an atypical presentation of ataxia-telangiectasia.

This patient was referred to JNW by Professor Mario Manfredi, professor of neurology at the University of Rome. We thank him for permission to publish this report, and also for providing details of the histological findings in the patient's muscle biopsy specimen.

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