Sigmoidoscopy should be performed shortly after removal of a foreign body to look for mucosal tears, but in patients where the rectum has been habitually overdistended rectal damage is not found.

¹ Bailey, H, and Love, J McN, in *A Short Textbook of Surgery*, ed A J H Rains and H D Ritchie, 16th edn, p 1013. London, Lewis, 1975.

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Resolution of cutaneous manifestations of systemic sclerosis after haemodialysis

Acute renal failure, usually in association with accelerated hypertension, is a rare but well-described feature of systemic sclerosis.^{1 2} It appears to develop relatively early in the disease, often without previous evidence of renal impairment, and carries a poor prognosis. Richardson³ has reported a patient in whom haemodialysis and renal transplantation resulted in survival for 18 months, and resolution of cutaneous manifestations, arthralgia and Raynaud's phenomena. We report here a patient in whom dialysis and hypotensive therapy have been instituted for 13 months.

Case report

A 42-year-old woman presented to Dr K Keczes in the dermatology department in July 1974. She had noticed tenderness, pain, and stiffness in the fingers of both hands for the previous 18 months; later she had developed Raynaud's phenomena and extensive thickening with tightness of the skin around her face, lips, and neck. She also had had considerable difficulty in opening her mouth. There was no family history of systemic sclerosis, although a sister had had vitiligo, both parents pernicious anaemia, and her mother rheumatoid arthritis.

She had waxy, yellowish, shiny thickening of the skin and subcutaneous tissues of her face, neck, hands, and around her mouth. She could not open her mouth more than 2 cm between the incisor teeth. There was considerable immobility of her fingers and she could not form a fist with either hand. The appearances were those of classical scleroderma. Her blood pressure was 150/100 mm Hg, but there were no other physical abnormalities. Serum urea was 3.5 mmol/l (21 mg/100 ml), electrolytes normal, and creatinine clearance 79 ml/min. Cold agglutinins were present in a titre of 1/4 at 4°C and urinary protein excretion less than 100 mg in 24 hours. A chest x-ray film showed interstitial fibrosis, and pulmonary function tests showed diminished diffusing capacity. The results of other extensive investigations were normal.

She was initially treated with azathioprine, 100 mg twice daily, and topical corticosteroids and emulsifying ointment. After two weeks she developed anorexia and vomiting. On her second admission to hospital her blood pressure was 240/120, while fundoscopy showed exudates and haemorrhages. She rapidly became oliguric and her creatinine clearance was found to be 4 ml/min. Renal biopsy showed appreciable endothelial proliferation of the arterioles with narrowing of the lumina.

She was established on intermittent haemodialysis twice weekly, and eventually her hypertension was successfully controlled with oxprenolol, 240 mg; diazoxide, 200 mg; and tolbutamide, 1 g daily. Azathioprine was discontinued because of leucopenia and prednisolone 5 mg daily maintained. Over 13 months she has developed deep pigmentation and hirsutism (attributed to diazoxide therapy), but she has been generally well, and increasingly active. Her scleroderma has become much less prominent, while repeated examination and investigations have shown no obvious evolving disease of the respiratory or gastrointestinal systems.

Discussion

The cause of acute renal failure in systemic sclerosis remains unexplained, and there is no evidence that any treatment will halt the progression of renal damage once it has begun. The survival of this patient would seem to be attributable to the early and adequate

combination of powerful antihypertensive therapy and intermittent haemodialysis. The unexpected remission of the subcutaneous manifestations is inexplicable. Possibly a spontaneous remission⁴ has taken place. Treatment with azathioprine or the low dosage of prednisolone was probably not responsible. Conceivably chronic uraemia suppresses fibroblastic function and thereby reduces excessive collagen formation.

Evidence suggests that haemodialysis or renal transplantation in systemic sclerosis associated with acute renal failure may partially reverse the progress of the disease and allow a more optimistic approach to management.

¹ Moore, H C, and Sheehan, H L, *Lancet*, 1952, 1, 68.

² Rodnan, G P, Schreiner, G E, and Black, R L, *American Journal of Medicine*, 1957, 23, 455.

³ Richardson, J A, *Arthritis and Rheumatism*, 1973, 16, 265.

⁴ Tuffanelli, D L, and Winkelmann, R K, *Archives of Dermatology*, 1961, 84, 359.

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C3 activator and hypocomplementaemia in a "healthy" woman

Hypocomplementaemia and the presence of C3 activator (C3 nephritic factor) have been described in the following syndromes: mesangio-capillary nephritis^{1 2}; partial lipodystrophy and recurrent infections³; and partial lipodystrophy in combination with mesangiocapillary nephritis.⁴ Hypocomplementaemia induced by C3 activation has not been described in healthy persons, and we describe such a case here.

Case report

A 36-year-old woman was seen in February 1974, complaining of pain and stiffness in her thumbs, though these were neither red nor swollen and they moved easily. Her symptoms had started with tonsillitis in December 1972; One week later she had developed a swollen and red right Achilles tendon, followed by migrating pains and stiffness in different joints. She had improved after treatment with salicylates and oxyphenbutazone, but had since then occasionally suffered from arthralgia in her fingers. During the acute illness repeated examinations had shown neither haematuria nor proteinuria; the sedimentation rate (1 hour) was less than 10 mm, and no rise in antistreptolysin O titre had been detected.

Investigations showed ESR 4 mm; Hb 12.7 g/dl, white blood count $5.7 \times 10^9/l$ (5700/mm³); serum albumin 43 g/l, urea 1.7 mmol/l (10 mg/100 ml), creatinine clearance 91 ml per min. Urine deposit normal and no proteinuria. Tests for antinuclear factor were negative and antistreptolysin O titre normal. Several blood samples during 1974 showed the presence of very low C3 (3.5 mg/100 ml) and low total hemolytic complement (CH₅₀ less than 25 units per ml). C3 nephritic factor was present, as indicated by C3 breakdown in normal serum in vitro. The levels of Clq, Cls, C4, C3b inactivator (KAF), and properdin were all normal.

Discussion

Low C3 concentration induced by C3 activator was initially found in two-thirds of patients with mesangiocapillary nephritis. The role of hypocomplementaemia in this form of nephritis is obscure, since there is a poor correlation between the activity of glomerular disease and serum complement concentration.⁵ This type of hypocomplementaemia is, however, regarded as a predisposing factor in nephritis. Later C3 activation was described in a woman with partial lipodystrophy and increased susceptibility to infections but with normal renal function.³ Our patient with low concentration of C3 and C3 activator

is healthy and has had neither recurrent infections nor shown any signs of lipodystrophy, and her mild arthralgia has had no influence on her work as a clerk. The origin of the C3 activation in her case cannot be certain, but there are two possibilities. Firstly, a genetic deficiency, though her 14-year-old son has normal C3 and C4 (certainly this does not exclude a genetic origin, but it makes it less likely). Secondly, originally the complement activation might have been provoked by an infection, such as her tonsillitis and post-infectious arthritis.

At present this patient seems "healthy," apart from mild arthralgia in her fingers. Is the latter related to her hypocomplementaemia? Does her immune deficiency have any clinical importance? Finally, will her immunological deficiency eventually manifest itself as a nephritis?

Professor Anna-Britta Laurell, Lunds Lasarett, Sweden, analysed the complement factors and also gave good helpful advice.

¹ Spitzer, R E, *et al*, *Science*, 1969, **164**, 436.

² Peters, D K, *et al*, *Clinical and Experimental Immunology*, 1972, **11**, 311.

³ Alper, C A, Bloch, K J, and Rosen, F S, *New England Journal of Medicine*, 1973, **288**, 601.

⁴ Peters, D K, *et al*, *Lancet*, 1973, **2**, 534.

⁵ Vallota, E H, *et al*, *Journal of Pediatrics*, 1972, **80**, 947.

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Carcinoma in-situ of cervix in sisters

"There are no 'cancer families' in which the cervix has been the organ affected."¹ Yet if, as claimed,² early age at first coitus, having a first baby while a teenager, multiplicity of partners and pregnancies, and low socioeconomic class are relevant to the development of cervical cancer, it should not be surprising that example and environment might combine to lead to similar lesions occurring in sisters. In the family here described, the mother had four sons and four daughters and died at 50 from myocardial infarction. The pattern among the four daughters, all in socioeconomic class V, is as follows.

Case reports

Daughter No 1 was born in March 1936. First coitus was at age 18. She was pregnant when she married at 19 but aborted. She was still 19 when she had a baby. When 22 she divorced her husband and "had three chaps" before remarrying at 27, again when pregnant. That baby was premature and is backward. When 29 she had trichomonas vaginitis but her cervical smear was negative. She did not take the pill. She was 33 when at examination for secondary infertility her smear was reported positive. Cone biopsy confirmed carcinoma-in-situ of the cervix. Five and a half years later she is well, the cervix looks healthy, and the smear is negative.

Daughter No 2 was born in February 1941. First coitus was at 18 and first baby (premature) at 19. She married when 22 and had three babies and three miscarriages. The last pregnancy, a successful one, was at 28. She was 31 when at request for sterilisation she was found to have a positive smear. She had been taking the pill for the previous 18 months. The cone biopsy taken at tubal ligation showed carcinoma-in-situ. Three and a half years later she is well, the cervix looks normal, and the smear is negative.

Daughter No 3 was born in May 1942. First coitus was at 15 and she married at 16. She had one baby at 18 before she was divorced. She remarried and had three babies. She was on the pill from age 24 to 30. She was 32 when at request for sterilisation she had a positive cervical smear. Biopsy showed varying degrees of dysplasia and one area of carcinoma-in-situ. As she also had prolapse, she was treated by vaginal hysterectomy with repair. One year later she is well with a negative vault smear.

Daughter No 4 was born in October 1947. First coitus was at 17. She had her first baby when 19 and the second, by the same consort, when 24. Since then she has had two other partners, one for a few months and the other for nearly 3 years, and is still single. When 26 she had vulval warts, vaginal

thrush, and trichomonas vaginitis at different times, but her cervical smear was negative. She took the pill for 18 months up to this time. She was 28 when two smears were reported as positive. Cone biopsy in August 1975 showed areas of dysplasia but no carcinoma-in-situ.

Comment

Note that none of the sisters was aged more than 33 years at the time of the first positive smear. The moral seems to be that if a woman of low socioeconomic level has an intra-epithelial lesion of the cervix, she should be asked if she has any sisters.

¹ *Corscaden's Gynecologic Cancer*, ed S B Gusberg, and H C Frick, 4th edn, p 176. Baltimore, Williams and Wilkins, 1970.

² Jeffcoate, T N A, *Principles of Gynaecology*, 4th ed, p 394. London, Butterworth, 1975.

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Aplastic anaemia and hair dye

Patients with aplastic anaemia for which no obvious cause is found continue to be seen. Recent reports on the carcinogenicity and mutagenicity of hair colourants^{1 2} have re-emphasised the potential dangers of these products, which are in widespread use. We should like to report a case of fatal aplastic anaemia in a patient using a new hair dye. Although anecdotal and in no way establishing a cause and effect relationship, this case emphasises once more the need for careful questioning and continuing enquiry of such patients.

Case report

A 52-year-old housewife was admitted to hospital for investigation of spontaneous bruising and purpura of one month's duration. Four months previously she had received a short course of oxytetracycline and then penicillin from her general practitioner for a mild upper respiratory tract infection. One month before the onset of her haemorrhagic tendency she had started using a new hair dye containing paratolylenediamine. So far as she or her husband could remember there had been no other exposure to new cosmetic, household, or other toxic agents. She looked well but was anaemic and had numerous petechiae and ecchymoses on the skin and mucous membranes. There was no lymphadenopathy or hepatosplenomegaly and she was not jaundiced. Investigations showed a haemoglobin of 9.9 g/dl, with slight macrocytosis; the WBC was $8.5 \times 10^9/l$ ($8500/mm^3$) and platelet count $8 \times 10^9/l$ ($8000/mm^3$). Coombs test was negative. The LE phenomenon was not detected and tests for ANF were negative. Leucocyte and platelet antibodies were not found, and there was no evidence of haemolysis. Bone marrow examination showed complete absence of megakaryocytes and erythroid hypoplasia.

She was treated initially with prednisolone in high doses and infusions of platelet-rich plasma. There was no appreciable response to steroids and oxymethalone, 300 mg per day, was added, again without benefit. Over the next five months she received fresh blood and platelet transfusions as necessary but remained thrombocytopenic, anaemic, and, during the later stages, leucopenic. Repeat bone marrow examinations during this time showed severe hypoplasia of all three haemopoietic components and she died six months after presentation from an intracranial haemorrhage. Necropsy confirmed bone marrow aplasia.

Discussion

It is unlikely that either tetracycline or penicillin caused this patient's aplastic anaemia. The hair dye used contained paratolylenediamine (2-methyl 1.4 phenylenediamine) in a concentration of 0.03%. Phenylenediamine has been used for many years in the dyeing industry and its effects on the bone marrow are well known.³ Two groups have recently reported studies on the mutagenic and carcinogenic properties of the various paraphenylenediamine dyes.^{1 2} The mutagenic properties are retained even after mixing with hydrogen peroxide, the oxidant used in the lotion developer, and appreciable