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References


Myelofibrosis as a cause of pancytopenia in systemic lupus erythematosus

We read with interest the article by Dr Daly and Dr Scott describing the association between systemic lupus erythematosus and myelofibrosis. The association between systemic lupus erythematosus and myelofibrosis is rare and we report an additional patient showing this association.

CASE REPORT

A 28 year old white man had a four year history of polyarthritis, weight loss, and fatigue. Investigations showed that he had positive antinuclear factor and lupus erythematosus cells and systemic lupus erythematosus was diagnosed. Subsequent admissions were for several attacks of bilateral pleurisy and joint swelling (elbows, knees, ankles). Urinalysis also showed the presence of proteinuria (1.1 g/24 h). He received steroids in decreasing doses.

Bone marrow examination was performed two months before his final admission for investigation of progressive anaemia and thrombocytopenia. The peripheral blood picture at this time showed anaemia (haemoglobin 9.4 g/dl), leucopenia (white cell count 2.4 × 10^9/l), and thrombocytopenia (platelets 82 × 10^9/l). There was no history of exposure to irradiation or benzene. He had not received any immunosuppressive agents.

Bone marrow from the sternum and iliac crest was similar showing variable cellularity—hypocellular in some areas and hypercellular in others. Megakaryocytes were decreased in number; myeloid and erythroid activity was present. There were increased numbers of histiocytes and fibroblasts. No blast cells were seen. Masson trichrome stain revealed extensive fibrosis and andomori stain showed a diffuse increase in reticulin.

On his final admission to hospital his major complaint was dyspnoea. Physical examination showed the following: pulse rate 100 beats/min, regular; blood pressure 120/70 mmHg; respiration rate 21 breaths/min. Chest examination showed an area of dullness over the right lower lobe with presence of bronchial sounds and crepitations. A pericardial friction rub was also present. Abdominal examination showed hepatosplenomegaly and ascites. There were several erythematous macular lesions on the skin.

Laboratory investigations gave the following results: haemoglobin 8.4 g/dl, leucocytes 2.1 × 10^9/l with 66% neutrophils, 18% stab cells, platelets 66 × 10^9/l. Serum electrolytes were normal. Urea nitrogen was 45 mg/100 ml and creatinine 1.8 mg/100 ml. Serum enzyme studies showed normal lactate dehydrogenase, serum aspartate transaminase, and alkaline phosphatase activities. Serum iron concentration was 18 µmol/l, total iron binding capacity 22 µmol/l. Direct and indirect Coombs' test was negative. Serum folate and B12 were normal. Serum albumin concentration was decreased (18 g/l), γ globulins were increased (20 g/l). DNA binding was increased at 44%, antinuclear antibody positive at 1/40 dilution. Cold agglutinins were not present. Antibodies to platelets were not detected.

By the second day after admission the white cell count had fallen to 0.9 × 10^9/l and the subsequent course was marked by a rapid fall in haemoglobin, white cell count, and platelets. The patient also had fever, deterioration in renal function, increasing ascites, and hepatomegaly. All blood cultures obtained were negative. The day before he died his white cell count was 0.35 × 10^9/l and platelets were 1.0 × 10^9/l.

At necropsy the parietal pleurae of both right and left sides were thickened and covered with fibrinous exudate. The parietal pericardium was also thickened. There was massive ascites and a fibrinous exudate over the parietal peritoneum. The liver was considerably congested. The kidneys were mottled and diffusely granular.

Microscopy confirmed the presence of polyserositis. In the liver there was central fibrosis indicating chronic congestion. The kidneys showed changes consistent with systemic lupus erythematosus. The bone marrow sections confirmed the presence of myelofibrosis. There was no evidence of malignancy.

The above case report shows that myelofibrosis can be a cause of pancytopenia in systemic lupus erythematosus. The pathogenesis is not known but Dr Daly and Dr Scott have adequately reviewed the possible mechanisms by which myelofibrosis can occur in systemic lupus erythematosus. Unlike their patient, myelofibrosis was not reversed in our patient by corticosteroid treatment.

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Spiral organisms in endoscopic biopsies of the human stomach

We were particularly interested to read the article by Dr Rollason and his colleagues, in which they described spiral organisms, thought to be spirochaetes, in human gastric biopsies. We would like to draw attention to recent correspondence in the Lancet in which it is suggested that the Gram negative bacteria seen in gastric biopsies belong to the family Spirillaceae.

We are currently engaged in a prospective study of gastric biopsy material in an attempt to establish the nature and importance of the microbial flora in the normal and diseased human stomach. In our preliminary studies we have been able both to visualise and to isolate organisms which appear similar to those previously reported. We agree with Marshall, on the basis of morphological and cultural characteristics, that the organisms are Gram negative bacteria of the family Spirillaceae rather than "spirochaetes." In fact, our results suggest that these organisms are members of the genus Campylobacter, although they cannot as yet be assigned to any of the currently recognised species.

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Bedside biochemistry

The introduction of portable selective analysers has made the concept of bedside biochemistry a reality for many of the analyses now undertaken in the chemical pathology laboratory. Much debate has taken place both at meetings and in the published work about the advisability of this limited decentralisation. The disadvantages have been weighed against the advantages of clinical convenience, cost effectiveness, and efficiency. Priority must be given to safe guarding the best interests of the patient: it is important that analytical results generated outside the laboratory, by relatively unskilled staff, are accurate, precise, and reliable. These aims are included in the document issued jointly by the Royal College of Pathologists, the Association of Clinical Pathologists, and the Association of Clinical Biochemists.1

We would like to describe the cooperation, developed over a period of time, between the chemical pathology laboratory and the special care baby unit. The unit operates an American optical bilirubinometer (Ophthalmic Instrument Division of Reichert-Jung Limited, 820 Yeovil Road, Slough, SL1 4JB) as a side room analyser. The chemical pathology laboratory has established an advisory quality control service to the special care baby unit, by monitoring and improving the performance of the bilirubinometer and so providing a reliable bilirubin result for the patient.

The bilirubinometer is sited in a side room attached to the special care baby unit. Junior medical staff collect blood samples from their patients and measure the plasma bilirubin. Where possible, duplicate analyses are done on each specimen. The results are recorded in a book kept adjacent to the instrument. The chemical pathology laboratory is two floors below the special care baby unit. A senior member of chemical pathology was designated liaison officer to ensure efficient communication between the laboratory and the special care baby unit. After discussions, an initial monitoring service was established in October 1980 and continued during 1981. Aliquots of reconstituted Versatol Paediatric quality control material were taken to the special care baby unit by a member of the laboratory staff at varying intervals. Similar aliquots were also analysed in the main laboratory at the same time. The special care baby unit was, in addition, provided with a quality control data sheet for recording results. This sheet was designed to provide a graphical as well as a numerical record.

Some initial difficulty was experienced in obtaining a complete record of quality control analyses on the sheets provided. At the beginning of 1982, the service was formalised by providing a monthly quality control sample. This was extended to a weekly service for the first seven months of 1983. In May 1983 the DHSS issued a *Safety Information Bulletin*, SIB (9) 10, (2), concerning the "measurement of bilirubin for the jaundiced neonate" and the use of quality controls. It recommended that users of bilirubinometers should: (a) be made aware of the factors which affect analytical performance; (b) perform daily quality control checks; and (c) use a suitable quality control material, such as Versatol Paediatric, and be able to obtain specific advice on aspects of quality control material, procedures, and calibrants from the pathology laboratory. With this in mind our service was further extended to provide a daily service, which started in August 1983. The problems encountered in setting up and maintaining the service have been remarkably few; they can be attributed either to personnel or to the instrument. Personnel problems from both departments were predictable as on some occasions the quality control material although provided was either not assayed or assayed and the result not recorded. On other occasions quality control material was not provided. Both problems were overcome by personal communication with those concerned.

It was not our intention to undertake an evaluation of the instrument, which had already been purchased by the special care baby unit, as a full evaluation had previously been commissioned by the DHSS.2 It became apparent, however, that results obtained by the bilirubinometer were lower than those produced in the main laboratory using batches of Versatol Paediatric with consensus values between 325 and 358 µmol/l. Investigation revealed an inherent non-linearity in the bilirubinometer, which became significant for bilirubin values over 225 µmol/l. This concurred with a previously reported retrospective evaluation of the instrument.3 The main laboratory offered to provide extra back up facilities for the bilirubinometer, and the special care baby unit decided to send samples above 250 µmol/l, estimated on the bilirubinometer, for repeat analysis. The purchase of another suitable bilirubinometer is being considered by the special care baby unit to overcome the linearity problem. Other problems related to the instrument in which the main laboratory has been involved have included cuvette alignment, filter checks, and bulb replacements.

The day to day use of the instrument involves medical staff who change duties at regular intervals. Despite this, considerable care and skill has been achieved in the measurement of bilirubin by the special care baby unit staff. This is reflected by good precision, with between batch coefficients of variation between 2-5%. Regular preventive maintenance checks by the main laboratory and servicing by the manufacturers should circumvent the occasional instrument failure, which in the past has caused the instrument to be down for several days. Two important factors have contributed to the successful cooperation between the chemical pathology laboratory and the special care baby unit side room. These are the introduction of a laboratory liaison person to ensure efficient communication and the provision of a reliable back up service. A daily quality control service is now well established with the special care baby unit and data from the unit are compared with results obtained in the main laboratory. Similar procedures could be adopted for other ward side rooms which use stat instruments for the measurement of analytes including sodium, potassium, urea, creatinine, and glucose.

The main laboratory has the expertise to advise side room operators on analytical techniques, quality control methods, the purchase and evaluation of instruments and their suitability to ward use, as well as advice on health and safety aspects and clinical interpretation. The type of cooper-
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