Persistent haemorrhagic ascites in generalised haemolymphangiomatosis: a therapeutic dilemma

We report a case of intractable, recurrent intraperitoneal bleeding secondary to visceral lymphangiomatosis, which we do not think has been reported before.

Case report

A woman was well until 1976, when, aged 24, she developed recurrent haemoptyses. Chest radiography showed multiple right cavities in the upper zone. Sputum screening for acid fast tubercle bacilli yielded negative results, and bronchoscopy was unhelpful. Right upper lobectomy was performed, and histological examination showed fibrous cavities lined with alveolar macrophages laden with pigment.

She presented in 1981 with massive splenomegaly. Haemoglobin concentration was 121 g/l, and a peripheral blood film showed macrocytosis and Howell-Jolly bodies. The histological appearance of a bone marrow trephine biopsy specimen was typical of myelofibrosis. At laparotomy a multicystic spleen weighing 4.2 kg was removed. The splenic pulp was replaced by large thin walled vascular spaces filled with proteinaceous material (figure). Postoperative chest radiography showed upper mediastinal widening, and a skeletal survey showed sclerotic deposits in the pelvis, scapulae, and femoral heads.

In 1983 she developed severe anaemia (haemoglobin concentration 55 g/l) and gross ascites. Aspiration showed heavily bloodstained fluid with an initial haemoglobin content of 77 g/l. Over the next three years she required abdominal paracentesis on several occasions and received 48 units of blood. In January 1985 a Levine shunt was inserted and drained via the right internal jugular vein; her ascites improved. Scanning after red blood cells had been labelled with technetium-99m showed an unusual vascular pattern in the inferolateral aspect of the liver. At repeat laparotomy the left hepatic lobe contained large bullas filled with blood, which were excised. The venous tip of the Levine shunt became

obstructed in October 1985 and required replacement with a similar shunt to the left internal jugular vein. Subsequent scanning showed retrograde flow around this shunt but not down the superior vena cava. In February 1986, aged 34, she collapsed with angiographically proved emboli in both main pulmonary arteries. Streptokinase was thought to be contraindicated and embolectomy was unsuccessful. Necropsy showed organised thrombus extending through most of both pulmonary vascular trees as well as lymphangiomatosis of the liver, mediastinal pleura, kidneys, pancreas, and heart. The right atrium contained friable adherent clot 3 cm long.

Comment

Splenomegaly due to cystic lymphangiomatosis is rare. A review of reports between 1940 and 1952 identified 27 cases.¹ Associated ascites is usually chylous and implies coexistent retroperitoneal lymphatic disease. Such ascites has been successfully treated with a Denver peritoneal venous shunt.² Severe haemorrhagic ascites in a patient with normal coagulation has to our knowledge not been reported before.

The blood in the ascitic fluid may well have obstructed the first shunt in our patient. The second shunt was obstructed by a fibrous sheath. This complication is well recognised,³ and research into non-silicone materials that cause less fibroplasia is now in progress. Caval thrombi are a recognised and dangerous complication of Levine shunts and require anticoagulation. Unfortunately, our patient's recurrent emboli were both silent and untreatable as long term treatment was contraindicated by continuing bleeding.

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Strongyloides stercoralis infection in Burma Star veterans

Infections with the nematode worm *Strongyloides stercoralis* may persist for many years after exposure, by autoinfection. The condition is well described among former second world war prisoners of the Japanese¹ and occurs in over a fifth of those who worked on the infamous Thai-Burma railway.² Most infections cause a "creeping eruption" or "larva currens" eruption—in itchy, serpiginous urticarial rash which occurs intermittently, usually over the trunk. The real danger of strongyloidiasis, however, is the potentially fatal hyperinfection syndrome, which may occur when infected subjects become immunosuppressed—particularly with corticosteroid drugs.³

Soldiers of the second world war Burma campaign fought in areas endemic for strongyloidiasis. They usually had reasonably adequate footwear, however, making them much less liable to infection (which is acquired by skin penetration of free living soil larvae). Nevertheless, we have recently described a case of strongyloides infection in a British Burma Star veteran,⁴ diagnosed 40 years after tropical exposure. We therefore attempted to determine the prevalence of strongyloidiasis among this group of men.

Patients, methods, and results

We circulated a questionnaire at two reunion meetings of the Burma Star Association in 1985 in Bridlington and London. These questionnaires asked whether members suffered a creeping eruption type of rash, which was described in detail (colour photographs of the rash were also distributed). Those who responded positively were contacted further by post, and if their rash was considered typical investigations were arranged (three stool samples for microscopy and larval culture, blood eosinophil count, and an enzyme linked immunosorbent assay (ELISA) serum test for strongyloides). Of 566 who replied to the questionnaire, three men were finally found to have strongyloidiasis—a prevalence of 0.53%. These were all successfully treated with mebendazole.

Histological appearance of spleen. Top (low power): Multicystic spaces filled with proteinaceous material and lined by low endothelium. Bottom (high power): Detail of endothelium lining cysts.

Comment

In ex-Far East prisoners of war 84% of those with strongyloidiasis have the typical rash.¹ Given also the accepted limitations of our survey, this suggests that the prevalence of 0.5% for Burma Star veterans is an underestimate. Nevertheless, even this figure means that some 100 to 200 Burma veterans in Britain today have undiagnosed strongyloides infections. Although the infection rate in ex-Japanese prisoners is much higher, doctors should also be aware of the possibility of strongyloidiasis in veterans of the Burma campaign.

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β Endorphin: A factor in "fun run" collapse?

Over the past six years 38 entrants have collapsed near the finish of Tyneside's annual Great North Run (a half marathon "fun run"). Though they represent a very small proportion of the entry, which now exceeds 25 000 a year, we find it surprising that healthy men can run until they become confused, dehydrated, hyperthermic, and hypophosphataemic¹ without first experiencing intolerable discomfort. Because endogenous opioids suppress pain, have a possible role in temperature regulation, and may be responsible for "runner's high"^{2,3} the concentration of β endorphin—one of the most potent of these peptides—is worth considering as an important factor facilitating collapse during such runs.

Subjects, methods, and results

Blood samples were obtained from 11 runners who collapsed near the end of two consecutive Great North Runs and from a control group at both the start and finish of the race. Fortuitously, there were also 11 controls. Both groups comprised men of modest but previously undistinguished performance. The collapsers were aged 19-43 (median 27) and the controls 27-45 (37).

The plasma was separated immediately and stored at -20°C. As soon as was practicable after each race duplicate assays of plasma β endorphin were performed by radioimmunoassay, after immune affinity chromatography, using a method giving a cross reaction with β lipotrophin of less than 5%. The figure shows the results.

The control group had a mean β endorphin concentration of 8.4 (4.2) pmol/l before the run and 27.2 (9.7) pmol/l (SD) after the run. Two runners showed slightly increased initial concentrations, perhaps as a result of exercise or anxiety before the race. The highest control concentration after the run was 46.2 pmol/l. The collapsed group at the finish had a median β endorphin concentration of 110 pmol/l (range 66-414 pmol/l). The concentrations in the controls after the run were significantly higher than the corresponding starting concentrations ($p < 0.005$, Wilcoxon signed rank test) and were themselves considerably exceeded by the concentrations found in those who collapsed during or at the finish of the race ($p < 0.001$; Mann-Whitney rank sum test).

Comment

Though we cannot know the β endorphin concentrations of the runners before collapse, and they may reasonably be assumed to increase during the process of collapsing, the evidence from the control group is that the concentrations are already high before this happens. Janal *et al* showed in a double blind study that long distance runners experience hypoalgesia and "runner's high" and that these effects are associated with an increase in β endorphin concentrations and are inhibited by naloxone.³ We suggest that the unusually high concentrations of β endorphin in those who collapsed were probably responsible for the insensitivity to pain, enabling the runner to keep going. The sense of wellbeing produced by opioid peptides may be a

Changes in β endorphin concentrations (log scale) in those who completed the half marathon and those who collapsed near the finish. □ = Normal reference range.

factor that determines the competitor's enthusiasm for running. At times this seems to be extraordinary: of the men we studied, one had collapsed during a previous half marathon, and another, who spent considerable time in intensive care after his run, subsequently expressed the intention to continue participating in such events.

Treadmill exercise produces an increase in β endorphin concentration in both trained and untrained subjects.^{4,5} Gambert *et al* found this to be much greater in men than in women.⁵ Perhaps this accounts for the fact that over the past six years all of the runners who collapsed at the end of the Great North Run have been men.

Doubtless the potential collapser is caught up in the group enthusiasm that surrounds the run and is subjected to the pressures of self esteem and perhaps the knowledge that considerable sums of money given in sponsorship for charities are at risk. The important factor that enables such entrants to run until they collapse, however, is probably the high concentration of circulating endogenous opioids.

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