A man with aplastic anaemia

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Medical History

Mr M. is 28 years old and has been a qualified architect for two years. Until he was 24 he had been free from any major illnesses, but in October 1966, while still an architectural student he presented to his doctor with a sinister list of symptoms which had been progressively worsening for several months. His main complaints were of bruising on minimal trauma and of bleeding gums whenever he brushed his teeth. He had been feeling generally weak for some time, but had put off seeing his doctor because he thought he might have just been feeling "out of sorts" due to the strain of his architectural examinations. When he did eventually consult his doctor, he was very weak indeed and was subject to muscle and joint pains on exertion, he had also suffered from a throbbing generalised headache for about three weeks. There had been a few episodes of epistaxis and his mouth and throat had been sore for several days.

On examination, he was found to have ulcers in the mouth and pharynx, and the fauces were inflamed. Also, he had several large bruises on his legs, trunk and forearms. His general practitioner made a provisional diagnosis of leukaemia and arranged for his immediate admission to hospital for investigation and treatment. This action was expedient, because his blood picture on admission showed a severe anaemia of all the blood cellular components. Two sternal marrow punctures were unsuccessfully performed, and eventually an iliac crest trephine biopsy produced a minimal amount of marrow, which microscopically pointed to a diagnosis of aplastic anaemia. He was treated immediately with a transfusion of whole blood and for a further four weeks his blood picture was monitored with transfusions of whole blood and packed cells being given as necessary. During his stay in hospital it was not possible to elicit any history of exposure to noxious industrial chemicals or to drugs which might have been responsible for causing the aplastic anaemia. Six weeks after admission he was discharged to the care of his family doctor when his blood picture had improved for a period of two weeks without any signs of further deterioration. The possible poor prognostic significance of the episode was explained to Mr M. and his family, but in view of the encouraging progress he had made, it was hoped that a spontaneous recovery might have occurred.

For four months, Mr M. remained symptom free and resumed work as an architectural student, but then he was admitted to hospital again having been overcome by paint fumes while respraying his car. This apparently led to an exacerbation of his aplastic anaemia and it was necessary to treat him again with transfusions of packed cells. However, he was discharged, having made a good recovery only eight days later.

For almost two years, he was reviewed regularly by his general practitioner, but only on a few occasions did he require to attend hospital for transfusions to boost his blood picture, and each of these occasions only required a single night's stay as an inpatient. In January 1970, he presented with acute abdominal pain which was diagnosed as bleeding into the mesentery. On this occasion, he experienced his first adverse reaction to the transfusions which he was given, with marked pyrexia and rigors. The opinion of another specialist in blood disorders was sought at this time, and a course of oxymethalone and prednisone was started. Phytohaemagglutinin was kept in reserve as a last resort.

Mr M.'s blood picture improved little during the weeks following his discharge from hospital. He managed to remain active at his work, and the transfusions he needed were given at week-ends, since he refused to take time off work or to be kept at home or in hospital. By the end of March 1970, he was becoming very upset by his condition, and had learned to know his own haemoglobin level from his appearance and physical state. His prednisone and oxymethalone had been continued and he became markedly cushingoid as a result, which naturally added to his displeasure. While examining his conjunctivae in the mirror one morning,

to try to assess his condition, he noticed a yellowish hue and this was also seen when he attended an outpatient appointment later that day. From the results of liver function tests, it was thought that he might be suffering from mild serum hepatitis resulting from the many transfusions he had received, but there was also the possibility that the hepatotoxic effect of oxymethalone (which is a methyl testosterone derivative) might be the cause. The withdrawal of androgen therapy was not thought necessary and his oxymethalone was continued. A course of phytohaemagglutinin was started as a last resort.

There was no dramatic improvement as a result of treatment with phytohaemagglutinin, but over a period of a few months, Mr M.'s blood picture did show signs that his marrow was responding and the haemoglobin and WBC levels rose very slowly, though platelets remained consistently undetectable. By June 1970, he was having great trouble with epistaxes and bleeding gums again, and was very conscious of his cushingoid appearance. Nevertheless, with support from his doctor, his family and his fiancée, he still remained active and in regular employment, and was married early in September, even though hospital treatment was by this time becoming necessary almost every week-end. A crisis point was reached two weeks after his wedding, when quite suddenly he developed a severe acneiform eruption over the greater part of his body which necessitated an immediate hospital admission. The extent of the reaction was such that it was impossible for him to lie in any position without severe irritation, and as infection supervened over almost all body surfaces he became virtually moribund. Topical and systemic antibiotic treatment was given and his steroid doses were progressively reduced as it was considered that the dermatological disorder was primarily due to his sustained high dosage steroid treatment.

Following this last severe episode, Mr M. was discharged from hospital and has not required further admissions. He has however, required continued therapy with topical oxytetracycline which has been given in association with hydrocortisone as a spray (terra-cortril) from his general practitioner, and he is now (January 1971) almost ready to return to work.

Social History

Mr M.'s medical history over the last four years has been presented in some detail as an indication of the chronicity of his condition and of the need for persistent, often unpleasant treatment. This provides a basis for understanding the severe social problems which his illness has raised. I was fortunate to be able to spend a few hours with Mr M. once in his home, and we discussed in detail the problems he has faced because of the poor prognosis of his illness. The history above was almost entirely given to me by Mr M. who understands the nature of his disease, and who has taken a keen interest in his therapy and in each change in his condition. He has read widely and discriminately the literature about his condition and he knows the likely outcome of his almost inevitably fatal disorder. He appeared tired and unhappy when we first met, wearing dark glasses and he took great pains to try to conceal the disfiguring scars which covered his face and hands. He sat in an armchair which he had bought specially to give minimal contact with his tender skin, and chain-smoked an evil smelling type of cigarette which he rolled himself with Turkish tobacco. His condition was obviously causing him abject misery but during the course of our interview it became obvious that the basis of this was not just his physical state. His speech was slow and cautious, and for some time he was unwilling to discuss his feelings about the effects of his illness, but he gradually settled down and began to pour out the problems which had beset him, but had remained unknown to all save his closest friends and his doctor.

Effects on marriage prospects.

When the diagnosis of aplastic anaemia was first made Mr M. still had two years of his architecture course to complete and he was considering getting engaged. He was an athletic person, being particularly fond of ski-ing, and driving sports cars which he serviced and maintained himself. Ironically, he had taken out a valuable life insurance policy not long before falling ill. The severe prognosis which confronted him and his family did not leave him undaunted, and he did at one stage try to convince his intended wife that marriage was unlikely to be possible. However, when his illness seemed to remit in its early stages, he went ahead and became engaged and an early wedding was contemplated. His fiancée knew the situation entirely and the prospects were explained to her by the hospital physicians responsible for Mr M.'s treatment, and also at length by his general practitioner. Firm marriage plans were

not made and possible dates for the wedding were repeatedly put back each time exacerbations of his illness occurred. A firm date was eventually set for April 1970, and bookings for the church, reception and honeymoon were made, but following an episode of hospitalisation in February, the wedding had to be postponed and eventually the couple were married in September. They had to forego any elaborate wedding plans because they were forced to arrange the wedding quickly during a period of remission from illness, and it was not possible to take their honeymoon abroad as they had hoped. It was soon after the wedding that Mr M'.s skin condition became really severe, and this has naturally adversely affected the close mutual relations of the recently married couple, since Mr M. could hardly stand any skin contact at all.

From the time of the engagement, when Mr M.'s fiancée decided to continue with the wedding plans it was obvious to his medical advisers that no man could set the couple asunder even if an argument could be put forward that this might be desirable. The burden of support, advice and encouragement fell fairly and squarely on the family doctor who must at times have found himself in a very difficult position trying to reconcile the logical, unemotional need for cautious progress with the couple's irreversible desire to go forward with their plans, no matter what the consequences in this emotionally charged situation.

The problem of starting a family.

The decision whether or not to have children has naturally been raised by Mr M. and his wife. Although he is so very ill, he desperately wants to have children, but questions the wisdom of this in view of his present state. For some time, he has been expecting to die, despite the unusual relapsing course his illness has taken. His expectation of death has been reinforced as he has watched the progress of other patients who he knows to have suffered from the same condition, many of whom have died while in hospital with him. He reads the obituary columns of the local paper every day to follow the people he met in hospital. He firmly believes he will die within a few years, so he wants to have children as soon as possible. He would like to watch them grow up for as long as he can before he dies. However, he is postponing starting a family until he has discussed with his doctor the possibility of his illness being an inherited disorder. He admits that he and his wife do not intend to be very careful about their contraceptive precautions when it becomes possible for them to resume sexual relations, and if his wife should become pregnant neither of them would be unduly alarmed, and would not consider an abortion.

Financial considerations.

Financially, Mr M.'s condition has not been a problem until now. Being in a salaried profession, his income is not affected by short periods of absence from work, but he is worried about the prospect of long periods of absence in case he might lose his job. His wife works full-time as a secretary, and earns enough to keep both of them should the need arise, but this would not be possible if they started a family. In 1968, Mr M. was refused admission to his firm's superannuation scheme because of his medical state. As he had taken out a substantial life insurance before his condition was diagnosed, he is not troubled about his wife's financial prospects when he dies, and money has been set aside from this for boarding school education for any children they may have.

Another family problem for Mr M. is that his widowed mother may need to rely on him in future years. Although his wife has promised to take care of her mother-in-law if it becomes necessary, he feels that it is not fair to burden her with more responsibilities after she has already been confronted with so many.

Psychological problems.

Despite Mr M. suffering from a serious, potentially fatal condition, his spirits have generally remained remarkably high. He has been supported well during exacerbations of his illness and has shown a mental resilience which might seem remarkable. However, he does sometimes become depressed about his prognosis and admits to having been in very low spirits during the time of his severe skin reaction, when he seriously considered suicide as a relief from the severe pain he was suffering. He has been unable to work since his wedding because of this episode, and has been recovering at home since his discharge from hospital, which has added to his misery. He feels he is just rotting away, being left alone all day while his wife is at work, and his self-deprecation is made worse by the feeling that his wife is having to support him in

every way, emotionally, physically and materially. His psychiatric symptoms of severe depression have been recognised by his doctor and treated rapidly and vigorously.

The problem of social isolation.

One of the most distressing long term aspects of the illness from Mr M.'s point of view is the way in which it restricts his social life. Although he can accept not taking part in the sports and pastimes he enjoyed, he dreads the social isolation which he thinks would come if his friends knew the true nature of his condition. He never tells anyone at work about his problems, because he fears he will be labelled as a ' cripple ' and so he always makes light of his illness if it causes him to take time off work. Although his employers naturally know of his condition from the details given on medical certificates, he always tells his friends a different story. For instance, during his recent prolonged absence from work because of his skin reaction, he told his friends only that he had been suffering from an infection. He dreads the prospect that he might be singled out and, in his own words "wrapped in cotton wool", by his colleagues if they knew the true nature of his illness.

The family doctor's therapeutic task with Mr M. has been truly universal in providing for physical, psychiatric, and social support. This has been challenging and sometimes very time consuming. As an additional factor, the doctor has even had to bear in mind the long term cost of treatment as some of the drugs used are very expensive (e.g., 'terra-cortril' spray 60ml costs 87p; Mr M. used two every day).

The future role of the family doctor

The circumstances of Mr M.'s marriage can hardly be described as the best basis for a young couple to begin their married life. Yet throughout his illness his fiancée refused to change her intention to become his wife no matter how great the difficulties that beset her. She met Mr M.'s doctor early in his illness and has had frequent contacts with him since. Following her marriage she has transferred to his list. Since her wedding day she has not had cause to consult her new doctor on her own behalf, but it is easy to foresee possible emotional problems which may beset this young wife in attaining a full married association with her sick husband. The family doctor is likely to be called upon often to provide support for flagging spirits in times of difficulty.

Summary

For four years, Mr M. has suffered from aplastic anaemia, which has proved to be a chronic, progressively debilitating condition, with a probably fatal prognosis. This illness has presented persistent management problems for both his family doctor and for the hospital physicians who have collaborated closely in treatment and in providing support for the patient and his family during recurring periods of difficulty. The future course of the illness is unpredictable but it is likely to involve the family doctor in providing continued support for a young man with excellent prospects with normal health, during a protracted, demoralising illness with all its associated physical, psychiatric and social consequences. The role of the family doctor can be foreseen in providing support for a young woman who seems likely to be widowed at an early age, and continued care for the family which the young couple are determined to have. Such a task of continuing care for a family tenuously surviving on bonds of mutual love and understanding is not an easy one. This patient has known the nature of his condition throughout. The relationships between him and his family and doctor have been frank, patient and understanding. This has meant that the burden, for the whole family group, of having to cope with the situation has been eased by full discussion, and planning for the future has been easier despite the patient's uncertain prospects.