CASE REPORT

Massive splenomegaly in rural Malawi: new wine, old wineskins and the importance of collaboration

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SUMMARY

During a voluntary placement in rural Malawi, we assessed a 21-year-old man who presented with dyspnoea and lethargy secondary to a chronic refractory anaemia associated with massive splenomegaly. He was initially treated at the rural hospital for a presumptive diagnosis of hyper-reactive malarial syndrome (HMS) with long-term malarial prophylaxis. There was inadequate provision of blood products and the availability of suitable donors was limited by the high local prevalence of blood-borne viruses. He was transferred to the district hospital for further investigations after transfusion of three units of blood. Unfortunately, he self-discharged without receiving appropriate investigations and medical treatment. Subsequently, his family sought help from the local traditional healer who performed scarification to attempt to treat him. Further efforts to emphasise the importance of hospital-based care proved unsuccessful, and sadly this man died at his family home 3 months after his initial presentation.

BACKGROUND

This case highlights the challenges of delivering healthcare in rural areas with limited resources and infrastructure. In addition to discussing the differential diagnosis and management of splenomegaly in the tropics, the article examines the uptake of medical and traditional treatments and the sometimes uneasy partnership between traditional healers and medical healthcare professionals. These experiences were eye-opening, challenging and occasionally frustrating. However, the benefits of UK physicians gaining knowledge of working in resource-poor settings are numerous and the important role that UK health professionals play globally in achieving the millennium development goals is well recognised.¹ Particularly as a trainee in infectious disease (NV), the opportunities to gain clinical experience and undertake research in this setting are limitless.

CASE PRESENTATION

A 21-year-old man was assessed in the community during a voluntary work placement in rural Malawi. He presented with dyspnoea, lethargy, anorexia and reduced exercise tolerance. He had previously been treated with artemisinin combination therapy (ACT), praziquantel and numerous blood transfusions for a chronic anaemia. In addition to medical care, the family had consulted the local traditional healer who had also offered treatments; the exact nature of which was unknown. He was not on any regular

medication and did not report any allergies. On examination, he was unwell with breathlessness on minimal exertion. His observations were as follows: heart rate 120/min, respiratory rate 28/min and temperature 37.9°C. He had marked pallor and mild jaundice; abdominal examination revealed gross splenomegaly and 2 cm hepatomegaly ([figure 1](#)). There were no stigmata of chronic liver disease and no detectable ascites. The remainder of his examination was largely unremarkable.

INVESTIGATIONS

After discussion with his family, he was admitted to the local community hospital—this required his brother to carry him on a bike for 5 km to reach the nearest road, from where public transport could be obtained. The resources at the local hospital were scant and the limited range of available blood tests was performed. His initial results were as follows:

Haemoglobin	5.0 g/dL
HIV antibody	Negative
Hepatitis B sAg	Positive
Sickle cell test	Negative
Blood film	No malarial parasites seen Abnormal red cell morphology

DIFFERENTIAL DIAGNOSIS

There is a wide differential diagnosis of hepatosplenomegaly in sub-Saharan Africa ([table 1](#)). In contrast to developed world settings, infectious agents play a more significant role. The gross enlargement of the patient's spleen in this case makes HMS and lymphoproliferative disorders the most likely diagnoses.

TREATMENT

On admission to the rural hospital, he was started on fansidar (sulfadoxine and pyrimethamine), the only available alternative antimalarial to ACT for a presumptive diagnosis of HMS. In addition, he was given iron supplements, folic acid and antibiotics. The next challenge was to obtain blood products. Despite many patients on the ward requiring this, there was no local system for provision. Members of the local community were requested to donate blood. Numerous people came forward; however, many of these were unsuitable due to the high local prevalence of blood-borne viruses. A few suitable donors were identified and he was transfused with three units. He was then transferred to the district

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Figure 1 A photograph showing (1) left-sided abdominal swelling secondary to splenomegaly; (2) palmar pallor and (3) scarification of the thorax and abdomen.

hospital to obtain further investigations including routine blood tests, chest X-ray and abdominal ultrasound.

While waiting for further investigations, he was started on intravenous quinine and broad-spectrum antibiotics. It was not possible to recruit further blood donors and he remained frail. The patient self-discharged after a few days due to frustration with the hospital environment and because he was missing his family who were unable to visit, as his village was 70 km away from the district hospital.

OUTCOME AND FOLLOW-UP

At home, the family continued to engage with the local traditional healer who administered a treatment, which appeared on inspection to be burns or poorly healed scars to the thorax and abdomen (figure 1). We had a long discussion with the family about the importance of receiving hospital care and the serious consequences of not doing so. Unfortunately, this young man died 1 month after the authors' return to England.

DISCUSSION

We describe the case of a 21-year-old man presenting with a severe chronic refractory anaemia associated with massive splenomegaly. Hepatosplenomegaly is a common presentation in sub-Saharan Africa associated with significant morbidity and mortality.² In addition to lymphoproliferative disorders (LPD), a common cause of massive splenomegaly in the developed world, hyper-reactive malarial syndrome (HMS) and schistosomiasis are important differentials. Unfortunately, the exact

diagnosis in our patient will remain a mystery. What is known is that he had a refractory anaemia, most likely due to the increased pooling and destruction of red blood cells in a grossly enlarged spleen. Other potential contributing factors include haemolysis, bone marrow dysfunction, nutritional deficiencies and parasitic co-infection.

The limited diagnostics and treatments available at the rural hospital meant that HMS was the only condition, though presumptive, for which treatment could be offered. It is well recognised as one of the most common causes of splenomegaly in sub-Saharan Africa.^{3–4} The term 'tropical splenomegaly syndrome' was first coined in the 1960s, and since then it has been renamed as HMS.⁵ Despite the widespread distribution of HMS and associated morbidity and mortality, it is under-represented in the literature. Much of the data is historic and there are few recent studies. Fakunle⁶ first described criteria for its diagnosis, which was later refined by Bates *et al*⁷ in 1997 (box 1).

These criteria are of most value in resource-rich settings; however, a large proportion of patients with this condition live in rural areas with limited access to healthcare facilities. In a Ghanaian study of 221 patients with massive splenomegaly, HMS (41%) was the most common cause identified. It was a diagnosis of exclusion, and a 6-month trial of antimalarials resulting in at least a 40% reduction in the spleen size was used as a diagnostic test. This study showed that mean IgM concentrations were neither sensitive (64%) nor specific (57%) in distinguishing between HMS and LPD, which accounted for the other most important differential in this population.³ Similarly, it has previously been shown that mean IgM levels were not significantly different between patients with schistosomal splenomegaly and HMS.⁸ They also rightly question the value of histological changes on liver biopsy, which can be a non-specific finding, and most healthcare facilities are unlikely to have the expertise to perform safe biopsies and analysis. This study was set in starkly different healthcare settings to rural Malawi with significantly more advanced investigations available. However, the underlying diagnosis still remained elusive in 23% of patients. A Zambian study which analysed 344 cases of

Box 1 Diagnostic criteria for hyper-reactive malarial syndrome⁷

Major criteria include the following:

- ▶ Gross splenomegaly 10 cm or more below the costal margin in adults for which no other cause can be found.
- ▶ Elevated serum IgM level 2 SDs or more above the local mean.
- ▶ Clinical and immunological responses to antimalarial therapy.
- ▶ Regression of splenomegaly by 40% within 6 months after start of therapy.
- ▶ High antibody levels of *Plasmodium* species ($\geq 1:800$).

Minor criteria include the following:

- ▶ Hepatic sinusoidal lymphocytosis.
- ▶ Normal cellular and humoral responses to antigenic challenge, including a normal phytohaemagglutination response.
- ▶ Hypersplenism.
- ▶ Lymphocytic proliferation.
- ▶ Familial occurrence.

Table 1 Differential diagnosis of splenomegaly in the tropics

Infection	Haematological
Hyper-reactive malarial syndrome (HMS)	Lymphoma (non-Hodgkin's lymphoma and Hodgkin's disease)
Schistosomiasis (<i>Schistosoma mansoni</i>)	Tropical splenic lymphoma
Chronic hepatitis B with portal hypertension	Leukaemia
Miliary tuberculosis	Myelodysplastic syndrome
Brucellosis	Thalassemia
Splenic abscess	Polycythaemia rubra vera
Visceral leishmaniasis	
Other	
Splenic metastasis	
Splenic cyst	

splenomegaly also found similar aetiological factors, with HMS accounting for about 40% of the cases.⁴ Similarly, HMS was the most common cause (31%) of massive splenomegaly in Nairobi, Kenya.⁹

The use of long-term prophylactic antimalarial therapy in the management of HMS was first described in the 1960s. Agents including proguanil, chloroquine and primaquine have been shown to reduce spleen size and reduce associated morbidity and mortality.^{10–11} Unfortunately, none of these antimalarials were available locally, and therefore Fansidar was used. The proposed mechanism of action is to prevent further exposure to malarial antigens, thus removing the stimulus for the immune response, thought to be propagating the chronic splenomegaly. Splenectomy is of little value in these patients due to the high risk of postoperative complications and subsequent risk of exposure to life-threatening infections.¹²

We were significantly limited in exploring other important differentials, particularly lymphoproliferative disorders. B-cell LPD and tropical splenic lymphoma have been reported as significant causes of massive splenomegaly in sub-Saharan Africa.³ His blood film showed abnormal red cell morphology, which was non-specific, and no other abnormalities of diagnostic value were reported. Definitive histological investigations such as a bone marrow examination would have been of great value, but this was unavailable in this rural setting. Abdominal ultrasound was available at the district hospital, but unfortunately the patient took his own discharge before this could be completed.

Schistosomiasis, a parasitic infection caused by trematodes, is endemic in Lake Malawi with *Schistosoma haematobium* and *Schistosoma mansoni* being the two species responsible for the high prevalence of infection. *S. mansoni* can result in hepatosplenomegaly due to chronic periportal fibrosis and portal hypertension. In contrast, *S. haematobium* results in urinary schistosomiasis. Both species are present in Lake Malawi; however, *S. haematobium* is predominant, which rarely results in hepatosplenic involvement.¹³ Our patient received praziquantel to treat schistosomiasis and multiple courses of artemether, which has also been shown to have schistosomidal activity. It is also well recognised that the interplay between schistosomiasis and HMS is important and that their coexistence accelerates the underlying immune responses.¹⁴ Interestingly, treatment of *S. haematobium* infection in coinfecting Kenyan children resulted in regression of splenomegaly.¹⁵

It is also worth noting that he was hepatitis B sAg positive on admission. It is possible that portal hypertension associated with chronic hepatitis B contributed to splenomegaly, but he did not have any clinical stigmata of chronic liver disease and it is unlikely to have been the sole contributor to massive splenomegaly.

He was transferred for further investigations and management to the local district hospital, 70 km away from his home. While waiting for these, he remained weak and frail. He became frustrated by the delay in obtaining blood products and felt very homesick, which led him to self-discharge. Before our return to the UK, we had a long and frank discussion with his mother to emphasise the importance of hospital care in the investigation and management of his condition. During this visit, it became apparent that they had sought help from the local traditional healer and there was evidence of scars on his thorax and abdomen. Scarification is a common practice among traditional practitioners and one cross-sectional study analysing the health beliefs, knowledge and practices of Nigerian communities in the management of splenomegaly showed that over 45% of the population would prefer to consult the local traditional healer.¹⁶

Healthcare in Malawi is predominantly government-funded and aims to provide free access to basic medical care for all Malawians. However, 80% of the population lives in rural areas and the lack of infrastructure limits access.¹⁷ The use of traditional medicine in Malawi and many other African countries is widespread and a way of life, with up to 80% of the population depending on it for their primary healthcare.¹⁸ The WHO defines it as the sum total of knowledge, skills and practices based on the theories, beliefs and experiences indigenous to different cultures that are used to maintain health, as well as to prevent, diagnose, improve or treat physical and mental illnesses.¹⁸

Experience in Malawi showed persistent preference for traditional African medicine over medical treatment. This was clearly demonstrated during a local measles outbreak, where the majority of children receiving immunisation already wore necklaces acquired from traditional healers perceived to confer immunity. The use of traditional medicine has been shown to result in delays to accessing medical care; it therefore remains important for healthcare workers to encourage uptake of appropriate medical services in a prompt and timely manner.¹⁹ Diverse and complex factors are likely to result in an interplay in determining the choice of treatment including accessibility, economic and environmental constraints, understanding of disease and local societal preference. Some of these factors are reflected in our case as the patient self-discharged due to the distance from home and the impossibility of family visits.

A number of studies have shown that knowledge and beliefs were the critical factors causing barriers to medical care and despite adequate medical provision, patients continue to access traditional healers.^{20–22} Chukweneke *et al*²³ found that under-utilisation of healthcare services was often due to a community finding that the model of illness was unacceptable; they preferred traditional healers who have similar health beliefs to their own. It is believed by many that some symptoms are supernatural in origin, the handiwork of neighbours or vengeance from an offended god, therefore indicating traditional healing to be appropriate.²⁰ This was demonstrated to us while attending a clinic of a local traditional healer—his method of working was to identify, via a spiritual diagnosis, a person wishing ill on his client and to confront them. If a person could not be identified, a mixture of herbs would be administered by varied routes including ingestion and insertion into small incisions in the skin. Our patient appeared to have had this treatment or a variation of superficial burns, also an accepted practice in this area of Malawi. This illustrates the conflict that can arise between traditional beliefs and medical healthcare. His family preferred to seek help from the local traditional healer instead of re-engaging with hospital-based care.

The importance of information exchange and collaboration between medical healthcare and traditional medicine is well recognised and the benefits of this approach are well documented in the management of HIV.²⁴ On reflection, it is possible that collaboration with the traditional healer may have improved concordance with medical treatment, as he had an existing trusted relationship with the family. However, there was no history of joint working in this area, with a degree of mistrust on both sides, and the practicalities of harmonising two models of healthcare so starkly different can be met with significant difficulty.²⁵

In summary, this case highlights a number of important factors concerning the challenges of medical care in rural Africa. Limitations in accessibility to appropriate investigations and treatment made accurate diagnosis difficult and resulted in the

loss of faith in hospital-based care by the patient and his family. Personal beliefs and concepts of illness were significantly divergent from that which we are accustomed to and optimising mechanisms to encourage collaboration and information exchange between patients and different models of healthcare are pivotal in delivering healthcare in this setting. Gaining 'hands-on' experience proved invaluable not only in increased diversity of clinical experience, but also provided insight into a community working hard to overcome their own problems and live life to the fullest.

Patient's perspective

- ▶ The patient's view was often difficult to discern, as we were only able to communicate through a translator and he was frail due to severe anaemia. Some indication of his family's perspective was gleaned from two main discussions between them and the authors.
- ▶ An initial discussion took place at his home prior to hospital admission, where the traditional healer was present. It was clear during this interaction that the patient and his family had a trusted relationship with him. The family found him easier to talk to as he was using their own language and his methods seemed familiar to them. The traditional practitioner appeared uneasy of our presence, possibly due to a history of criticism of their practice by western doctors.
- ▶ The second main communication occurred after the patient's self-discharge from hospital. He had left due to being a long distance from home, making it impossible for his family to visit. He was also frustrated at the long delays in treatment caused by limited resources. His family were aware of the gravity of his condition and the need for hospital-based care. However, they consistently favoured the traditional approach due to their familiarity with his methods in addition to the physical barriers described. It is possible that improved understanding and communication between medical practitioners and local traditional healers may have resulted in a better outcome in this case.

Learning points

- ▶ There are a number of diagnostic challenges while practising medicine in rural settings including accessibility, economic and environmental constraints and different models of health beliefs.
- ▶ Traditional medicine is an integral part of healthcare in African nations.
- ▶ Collaboration and information exchange between patients and practitioners from different models of healthcare is likely to result in improved health.
- ▶ Working in rural, resource-poor settings is of significant value for UK healthcare workers.

Competing interests None.

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