A woman with recurrent anaemia and the Malawian health system

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A 22 year old woman presented to the general medical clinic in 2005 with a six month history of generally feeling tired. She had been seen a number of times in the accidents and emergency department with similar symptoms and a diagnosis of anaemia was made at each visit. Iron tablets and albendazole were prescribed. She took the iron tablets for about two months but there was no improvement. It was at this time that she was referred to the general medical clinic. A review of her medical history showed that she had grown up well and had completed all her immunizations. She attained menarche at the age of 13 years and had a regular menstrual cycle. Her periods were not heavy and she was not sexually active. Her diet was normal. She had no other sites of blood loss that she could point to. A physical examination showed a well-developed 22 year old woman with normal blood pressure. The only abnormality was a pale conjunctiva. In the chest, heart, skin and abdomen, there were no abnormalities.

The womn brought with her a full blood count result which showed an HB of 4.9g/dL, MCV of 88fl, RBC count of 2.3 x 1012/L and a platelet count of 78,000/uL. The rest of full blood count report was normal. A peripheral blood film was requested and the patient was admitted to hospital for a blood transfusion. She received two units of blood and felt much better. A peripheral blood film was reported as showing no specific important features. An HIV test was negative. A bone marrow aspirate was reported as showing features of aplastic anaemia. A detailed drug history was taken which yielded no positive leads. She was treated with 60mg of prednisolone daily for one month. A month later at a predetermined clinic review, there was no improvement. Her HB at this visit was 3.8g/dL. She was readmitted to hospital for another two units of blood transfusion. Prednisolone was tapered to stopping over one month. She was transfused blood almost every four to six weeks for six months until her referral to a haematology unit in South Africa.

A review of her history and physical examination was done in South Africa and confirmed the findings in the referral letter. In the hematology unit, a direct antiglobulin test; and Anti Nuclear Factor (ANF) of 1/320 (speckled), were found to be positive. She had low C3 and C4 complement and a negative Epithalial Membrane Antigen (EMA). The treating team concluded that the woman had Evans syndrome Idiopathic Thrombocytocypenic Purpura(ITP) plus heamolytic anaemia) on a background of SLE. She was started on a combination of prednisolone and azathioprine. By the time she was being discharged, her blood count had normalised. She was sent back to Malawi with a six month treatment without any scheduled follow up visit to the heamatology unit in South Africa. After completing her course of treatment she was followed up in the general medical clinic until January 2011 when she was lost to follow up. All these years she had remained well.

Anaemia is a common finding in patients presenting and being admitted to the adult medical wards at the Queen Elizabeth Central Hospital (QECH). Data is hard to come by but estimates would suggest that almost 50% of medical admissions to the QECH would have low haemoglobin (HB) as an isolated presentation or as part of other medical conditions. Infections like tuberculosis and non typhi salmonella are some of the commonest causes of anaemia on the background of HIV infection[1,2]. Another significant proportion of patients may have anaemia as a result of nutritional deficiencies or worm infestation. A small proportion of patients after investigations would have a hematological malignancy. There is however a large minority of patients both HIV infected and uninfected who present with anaemia and whose investigations do not point to a specific treatable condition with current capacity at our hospital. The attending physician is at a loss as what to do next. The patient being presented here is a case in point. She is unlikely to be an isolated case. A young woman who looks healthy without other health issues presenting with a chronic anaemia whose cause was difficult to find. This patient was lucky that the government of Malawi allowed her to travel to South Africa and be evaluated in a haematology unit where a diagnosis of Evans Syndrome with a possibility of SLE was made.

In 1951 Robert Evans described 24 cases whose main characteristics were acquired haemolytic anaemia and primary thrombocytopenic purpura[3]. Later a syndrome was named after him that comprises a combination (either simultaneously or sequentially) of immune thrombocytopenia (ITP) and autoimmune haemolytic anaemia (AIHA) with a positive direct antiglobulin test (DAT) in the absence of known underlying aetiology. Thus, by definition true Evans syndrome is a diagnosis of exclusion and other confounding disorders should not be present[4]. Literature describes this syndrome to be rare. Literature suggests that patients who present purely with Evans syndrome are likely to manifest with SLE at some point[5]. In one study, the conditions were diagnosed at the same time in almost 92% of the patients.

Systemic lupus erythematous is a commoner syndrome but rarely diagnosed in our clinical practices in Malawi. We would not have made a diagnosis in this young woman even if we were to be suspicious of the disease because of lack of laboratory capacity to carry out tests that would confirm the diagnosis.

This patient has been lost to follow up. We do not know whether she has now developed full blown SLE. One only hopes she is still well.

What are the issues?

The aim of presenting this case is not to describe either Evans syndrome or SLE. There are many good reviews on these subjects in literature that would adequately describe both disease entities[6-10]. The main aim is to demonstrate how the present health care system fails many people. Malawi recently celebrated 48 years of independence and we still refer people to other countries for treatment even for the simplest of ailments only if the capacity was made available. Almost every month, the referral team at QECH

meets to decide which patients should be referred for further management. The majority of the patients are either children or adolescents with valvular heart disease. It would cost the country less if capacity for conducting such operations was made available at one of the hospitals in Malawi and experts brought once or twice a year to carry out these operations. Coming back to our patient, she illustrates availability of diagnostic tests (some simple e.g. the direct antiglobulin test and others complex) in some national unit may save the government of Malawi large sums of money while at the same time providing quality health care to its people. We are not privy to the amount of money the government of Malawi paid for this patient's care. It is likely to run into millions of Kwacha. However if a national laboratory (dedicated to the task) was available that could do all these tests (with quality control/ and quality assurance that is required) for the nation, we could save a lot of money. Many more patients like this young woman are not investigated to their logical conclusion because of logistical handicaps and end up either living long with disabilities or dying prematurely. It is imperative that antiglobulin tests be made available at the main referral hospitals and technologists trained to perform these tests to the highest quality possible. QECH and Kamuzu Central Hospitals being teaching hospitals, need to be provided with the capacity to carry out such tests for the nation.

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