Exchange transfusion for intrahepatic cholestasis due to sickle beta thalassaemia

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(Index words: intrahepatic cholestasis, sickle cell disease)

Introduction
Intrahepatic cholestasis is a rare but potentially fatal complication of sickle cell disease [1]. Sickle cell intrahepatic cholestasis (SCIC) is characterised by right upper quadrant pain, hepatomegaly and progressive hyperbilirubinaemia. To date 15 adult patients have been reported in the literature and only six survived [2]. We present a case of haemoglobin S/Beta thalassaemia with intrahepatic cholestasis successfully treated by partial exchange blood-plasma transfusion.

Case report
A 34-year old patient with haemoglobin S Beta thalassaemia presented with a three week history of jaundice and right hypochondrial pain. He did not complain of fever, anorexia or pruritus. There was no evidence of liver failure or chronic liver disease. Vital signs were normal. Sickle beta thalassaemia had been diagnosed at the age of 18 years and the patient had several episodes of haemolysis requiring blood transfusions. Investigations showed: haemoglobin 9.9 g/dl, WCC 10.8 × 10^9/l, and platelets 407 × 10^9/l, AST/ALT 131/170 IU/l, alkaline phosphatase 294 U/l, total bilirubin 215 μmol/l (102 μmol/l conjugated), reticulocytes 17%. An ultrasound examination showed gall stones without evidence of cholecystitis or dilatation of intra- or extra-hepatic bile ducts. Serology for hepatitis B and C was negative. Over the ensuing week he developed nausea, anorexia and pruritus. Bilirubin rose to 632 μmol/l (513 μmol/l conjugated). Initial management included vitamin K, blood transfusions and empirical broad spectrum antibiotics. Liver biopsy was performed. Biliary canaliculi and sinusoids distended with bile, and ballooning degeneration of hepatocytes and focal necrosis were noted. Over the next two weeks the patient became progressively more icteric and drowsy, developing asterixis and coagulopathy. He had blood stained vomiting and melaena. Investigation at this stage showed: total bilirubin 992 μmol/l (889 μmol/l conjugated), INR 3.4, haemoglobin 7.5 g/dl, WCC 35 × 10^9/l, blood urea 37 mmol/l, AST/ALT 171/81 IU/l. Partial blood-plasma exchange transfusion was carried out.

Discussion
A review of liver disorders associated with sickle cell disease has identified five specific syndromes i.e., hepatic crisis, viral hepatitis, gall stones, cirrhosis and intrahepatic cholestasis [3]. SCIC has been reported in homozygous HbS sickle cell patients as well as in patients who are heterozygous for HbS and beta thalassaemia [2]. SCIC is rare but carries a grim prognosis. The aetiology remains unclear and the pathophysiology is poorly understood. SCIC is histologically characterised by intracanalicular cholestasis [4].

Only six of fifteen previously reported adult patients had survived [2]. All six had undergone exchange transfusion. However, there are three reports of SCIC with a fatal outcome despite exchange transfusion [2,5,6]. In one report the patient was relatively older (48 years) and had hepatitis C and cirrhosis [2]. In the second report the patient expired two months after discharge from hospital and he had undergone two ERCPs and a liver biopsy prior to exchange [6]. The other fatal outcome was reported in a patient with advanced hepatic fibrosis who had a complicated trans-jugular liver biopsy which required surgical intervention [5]. All of them had advanced liver disease or had undergone an intervention or both prior to exchange transfusion. This may have contributed to the bad outcome in these patients. A series from England reported a 28% incidence of death following liver biopsy in patients with sickle cell disease associated with acute hepatic disease [7]. All forms of surgery including cholecystectomy, carry a high complication rate in these patients [8].

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Case reports

The treatment of this potentially fatal complication is not universally accepted. Early exchange transfusion seems to be the most accepted and effective form of therapy. The aim of this therapy is to reduce and maintain the haemoglobin S level at less than 30%. Clinical improvement may result from the exchange transfusion of RBC or FFP or both [9].

References

Uncommon presentation of hypereosinophilic syndrome

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Introduction

The hypereosinophilic syndrome is a group of diseases characterised by persistent blood eosinophilia, defined as more than 1500 cells per micro liter [1], with end-organ involvement and no recognised secondary causes such as skin diseases, parasitic infection or allergy (Table). The sustained overproduction of eosinophils causes eosinophilic infiltration and release of mediators causing damage to multiple organs [2]. Cutaneous involvement occurs in more than 50% of patients, but it usually appears late in the disease and is of less importance than cardiac and other organ involvement [1]. We describe a patient with the hypereosinophilic syndrome (HES) in whom the initial manifestation of the disease was recurrent, severe mucosal ulcers involving the mouth and genitalia.

Case report

A 39-year old man was well until January 2007 when he developed persistent oral ulcers. Two months later, the oral ulcers increased and he also developed erosive lesions on the glans penis and ulceration of the skin of the scrotum (Figures 1 and 2). Clinical examination of the heart, lungs and abdomen were unremarkable and there were no other skin lesions. His white cell count was 35×10\(^9\) /l with 54% eosinophils. He underwent investigations including stool tests for amoebae, ova and cysts, and bone marrow examination which did not show a cause for his illness. He was treated with sequential courses of intravenous antibiotics, albendazole, di-ethyl carbamazine and a short course of steroids without improvement in his symptoms or eosinophil count. He was discharged from hospital and was followed up in the clinic.

In August 2007 he was re-evaluated because of persistent orogenital ulcers and eosinophilia without other skin lesions. His white blood count at that time was 23×10\(^9\) /l with 60% eosinophils. The haemoglobin and platelets were within normal range. Eosinophilic leukaemia (myeloid cells that demonstrate a clonal chromosomal abnormality [4]) was excluded by blood picture, bone