Spontaneous intracranial haemorrhage in haemophilia A

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Sri Lanka Journal of Child Health, 2001; **30**: 46-7

(Key words: Spontaneous intracranial haemorrhage, haemophilia A)

Introduction

Spontaneous intracranial haemorrhage is rarely seen with clotting disorders. Haemophilia A, an X linked recessive clotting disorder, usually presents with haemarthrosis or prolonged bleeding from cut injuries. Intracranial bleeds are rare and when a child presents with this, it's a dilemma for both the paediatrician and parents. Treatment regimens differ, and this child was treated with a combination of cryoprecipitate and factor VIII concentrate in the acute phase for 14 days and at present is on prophylactic factor VIII concentrate once a week.

Case report

C, a diagnosed haemophiliac, presented initially with vomiting, headache and right sided focal convulsions. He was admitted to Nagoda Hospital, Kalutara and transferred to National Hospital of Sri Lanka for a CT scan, which showed a small left subdural collection of blood (Figure 1). This was seen by the neurosurgeon who decided not to intervene and the child was managed conservatively with cryoprecipitate and factor VIII concentrate for fourteen days.

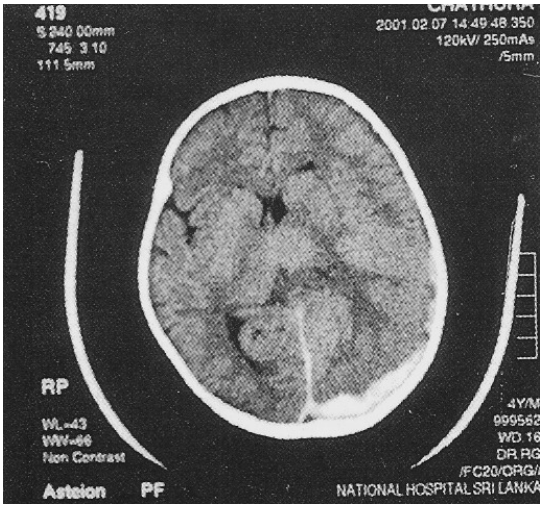

Figure 1. *Left subdural haemorrhage*

During this period he was transferred to our unit for control of convulsions. After discharge he was readmitted on three occasions within a space of a month, the first being for headache and the next time for his second dose of Hepatitis B vaccine and the third being for persistent headache with no obvious neurological signs.

The first episode of headache was treated with cryoprecipitate, where a hundred percent correction was done for three days and headache was better. But he presented with persistence of this headache and a neurological consultation was sought and it was decided that a CT scan was not indicated. However we decided to go ahead with a CT scan as his symptoms appeared to be significant. This showed a moderate sized subacute subdural bleed in the right fronto-parietal region (Figure 2).

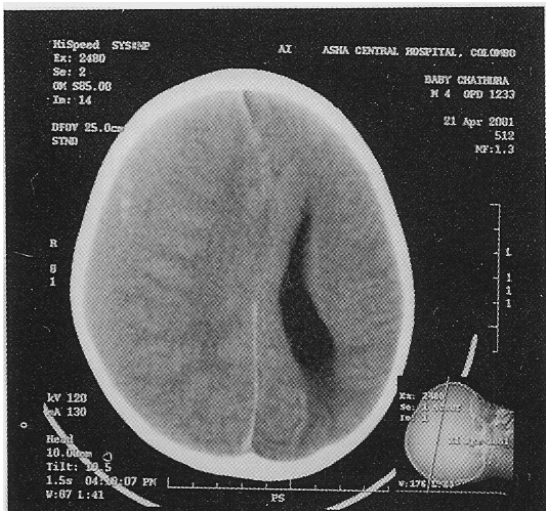

Figure 2 *Right frontoparietal subdural haemorrhage*

We were surprised with the result as the initial CT scan had a collection on the left side and this time on the right side. Then came the decision making on how to treat this problem. Following discussions with the haematologist we decided to treat him with cryoprecipitate and factor VIII concentrate for a total of 14 days. Another dilemma was the prevention of further internal bleeding and at present he is on prophylactic factor VIII concentrate one day of the week. The present plan is to continue the regime for another six months.

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Discussion

Intracranial bleeds are rare with Haemophilia A. It is even rarer to see these without any joint involvement other than in neonates. Intracranial bleeds are life threatening mainly due to the closed compartment into which blood collects and minimal intervention is only possible other than replacement therapy. Surgical intervention has been carried out in the past for evacuation of intracranial haematomas[1]. Underlying vascular malformation in a haemophiliac may be one of the causative factors[2]. Diagnosis of intracranial bleeds clinically is possible if obvious neurological signs are present. In the absence of this computerized tomography has to be done to make the diagnosis[3]. This case is an example where the bleed has been probably slow to develop with absence of neurological signs.

Replacement therapy is the present goal and when cloned genes for factor VIII production can be added to target cells via viral vectors this disease could be considered curable[4]. Since the first use of blood transfusion to treat haemophilia in the 1840's it took almost another century to separate fresh frozen plasma and cryoprecipitate to treat them. But in the 1980's transmission of hepatitis B and HIV necessitated stringent testing methods and purer plasma derived or recombinant proteins being used for replacement therapy[4].

Our patient was treated with cryoprecipitate and later with factor VIII concentrate as he developed anaphylaxis to cryoprecipitate. At the same time factor VIII inhibitor levels were checked, as his bleeding was unusual. He has long acting inhibitors too. At present he is on prophylactic factor VIII concentrate one day of the week and according to published regimes this is well below what he needs4. Availability of factor VIII and financial constraints have limited our options and we hope he recovers with minimal damage.

Acknowledgements

We thank Dr. I N A Gooneratne Consultant Radiologist and Dr. S Vidyathileke Consultant Haematologist Lady Ridgeway Hospital for their invaluable help.

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