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A review of cephalhaematoma in adult sickle cell disease patients

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Summary

Twelve patients with sickle cell disease (SCD), eleven males within the age range 21 to 24 years and one female, fourteen years old presenting with cephalhaematoma at the Haematology Day Care Unit (H.D.C.U.) of the University College Hospital, Ibadan during a three year period were reviewed. Only one patient was sickle cell haemoglobin SC (HbSC) while others were homozygous (HbSS). They all presented with a triad of symptoms consisting of fever, headaches and scalp swelling. Significant findings include the exclusive male preponderance and the almost exclusive presentation in HbSS patients. Treatment include analgesic and intravenous fluids.

Résumé

A l'Haematology Day Care Unit (HDCU) de l'University College Hospital, Ibadan, nous avons étudié, pendant trois ans, douze patients, onze hommes âgés de 21 à 24 ans et une jeune fille de quatorze ans, tous porteurs de la drépanolytose et souffrant de céphalhématome. Un seul patient accusant l' hémoglobine Sc (HbSc) de la drépanocytose les antres étant homozygotes (HbSS). Tous présentaient les trois symptomes de fièvre, maux de tête et enflement du cuir chevelu. Parmi less conclusions significatives, nous avons constaté que la maladie se manifeste surtout chez les hommes et presque tonjours des patients HbSS. Le traitement comprend les analgésiques et les fluides intraveneux.

Introduction

Cephalhaematoma has been reported by some workers as a relatively uncommon complication of sickle cell disease (SCD)[1,2] the cause of which was attributed to intravascular erythrocyte sickling which results in vaso-occlusion, thereby leading to ischaemia of the tissues and probably rupture of the blood vessels[2]. Cephalhaematoma formation has been attributed to infarctive process during crisis[3]. Infarctive crisis is the commonest of crises occurring in SCD. Bone is one of the most frequently affected tissues partly because its extreme vascularity provides a site for sickling and sludging of red blood cells within the microvascular system, resulting in infarction[4]. Recently at our Haematology Day Care Unit (HDCU) we reviewed subperiosteal skull bone infarct with haemorrhage in eleven patients with homozygous SCD and one male patient with sickle cell haemoglobin SC disease. The present report highlights the clinical presentation and sex predominance of this complication.

Patients and methods

We reviewed the case notes of all patients who presented to Haematology Day Care Unit with cephalhaematoma between 1st July 1987 to 30th June 1990. These patients have been attending our Sickle Cell Disease Clinics regularly since diagnosis. In events of crisis before their next clinic appointment, patients are seen in the Haematology Day Care Unit where all haematological emergencies are treated. Coagulation tests were routinely done in all the patients.

Results

Clinical presentation: Table 1 shows the demographic data on patients who presented with cephalhaematoma. Eleven patients were males in their early adulthood and one adolescent female. All patients presented within 72 hours of the onset of symptoms. The mean age of 22.1 ± 2.7 years.

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Table 1: Presentation of patients with sickle cell disease and cephalhaematoma

Patients	Haemoglobin genotype	Age/Sex (yr.)	Site of cephalhaematoma	Period before presentation (hr.)
*1.	SS	24/M	(R) Frontoparietal	24
2.	SS	21/M	(L) Frontoparietal	24
+3.	SS	23/M	Bilateral parietal	72
4.	SS	24/M	(R) Parietal	48
5.	SS	23/M	(R) Parietal	24
6.	SS	21/M	(L) Frontoparietal	48
+7.	SS	22/M	(R) Parietal	24
8.	SS	24/M	(R) Parietal	24
*9.	SC	23/M	(R) Parietal	24
*10.	SS	23/M	(L) Frontoparietal	24
11.	SS	23/M	(L) Frontal	48
12.	SS	14/F	(L) Parietal	24

Note

* Denotes patients with recurring cephalhaematoma.

-R- Right

-L- Left

\+ Denotes patients on whom fine needle aspiration was done.

Table 2: Frequency of presentation of the different symptoms in sickle cell disease patients with cephalhaematoma

Symptoms	Total no. of patients	%
Swelling of the scalp	12	100
Headaches	11	91.7
Fever	11	91.7
Bone pains	10	83.4
Generalised weakness	5	41.6
Blurring of vision	2	16.6
Neck stiffness/pain	2	16.6
Vomitting/nausea	1	8.3

Table 2 shows the presenting symptoms and the frequency of each. Most patients presented with headaches, swelling of the scalp and fever. One of the two patients who did not complain of bone pains at presentation had severe bone pains one week prior to presentation. None of the patients was on Aspirin or Aspirin-containing drugs routinely prior to development of cephalhaematoma. Coagulation tests were within normal limits in all the patients.

Management: All patients were hospitalised for the period of treatment. Routine blood counts, bleeding time, coagulation screening tests, sepsis work-up and radiological examinations were performed on all patients. Patients were placed on intramuscular analgesics and intravenous fluids at the time of admission.

Outcome: The cephalhaematoma in all the patients resolved within two weeks of presentation. None of the patients developed osteomyelitis of the skull following cephalhaematoma. Three patients had recurrent cephalhaematoma. In patient No. 1, recurrence occurred on the same site two years after initial presentation while in the other two recurrence was on the contralateral side, presenting within six months of the first episode. These patients were managed as previously stated.

Discussion

Many of the complications of sickle cell disease resulting in pathological and clinical manifestations

are caused by the sickling process which leads to sludging and stasis in the capillaries and arterioles resulting subsequently in ischaemia and infarction[5,6]. Sometimes, these manifestations are in form of inflammation and or haemorrhage. In the patients reviewed the resultant swellings were found mostly in the parietal region of the head (see Fig. 1). We believe that these swellings arose as a result of pathology in the periosteal area of the skull since they did not cross suture lines. The fact that screening coagulation tests were normal in all our patients suggest that the underlying pathology is not a coagulation defect. Radiological examinations in these patients showed soft tissue swellings of the affected area of the scalp with no evidence of infection.

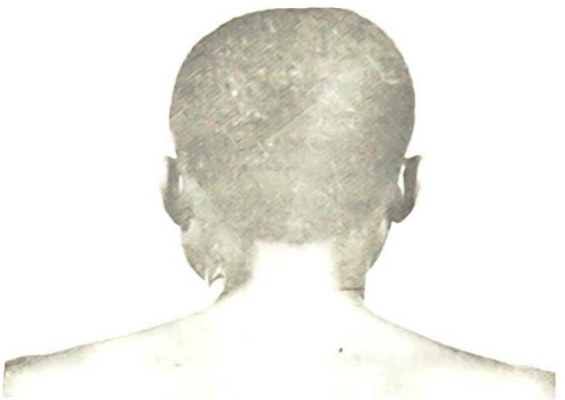

Fig. 1

Fine needle aspiration performed on two of our patients (Patient 3 and 7) yielded bloody aspirate. However needle aspiration could not be done on all our patients because of the risk of introducing infection or even precipitating osteomyelitis.

In a previous study, this complication was linked to the use of aspirin which is known to cause functional platelet defect. This was not the case in patients studied since none of them had history of aspirin ingestion. The most logical pathogenesis is therefore that of haemorrhagic infarction following the sickling process in the capillaries and arterioles supplying these areas of the head. The parietal region is the most frequently involved in cephalhaematoma in this series of patients (10/12), the reason for this is not clear. Also it is not understood why there is a male predominance. The present report also highlights the occurrence of cephalhaematoma in haemoglobin SC disease. The relative rarity of this complication in SC patients may be related to the lower incidence and milder clinical course in this category of patients. Generally, homozygous sickle cell patients have a higher concentration of sickle haemoglobin in their red cells, higher red cell turnover and require less anoxic insult to suffer an infarct than HbSC patients.

It is thus important for clinicians involved in the management of sickle cell disease patients to have a higher index of suspicion for cephalhaematoma especially in those who present with a triad of symptoms consisting of fever, headaches and scalp swelling. Our experience has also shown that these patients will do well with conservative management consisting of analgesics and intravenous fluids.

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References

1. Adedeji MO, Okafor LA, Ejele OA, Monu JUV. Sickle cell cephalhaematoma in Benin-City, Nigeria: A report of three cases. Central Afr. J. of Med. 1988; 34: 33-37.
2. Ndugwa CM, Mukidi P. Sickle cell cephalhaematoma East Afr. Med. J. 1976; 53: 545-548.
3. Konotey-Ahulu FID. The usual and unusual in clinical haemoglobinopathy. In Szabo G, Papp Z eds. Medical Genetics, Amsterdam: Excerpta Medica, 1977.
4. Attah B ed. Pathology of sickle cell disease. In Fleming AF. Sickle cell disease. A handbook for the general clinician. Edinburgh: Livingstone 1982: 42-56.
5. Fleming AF. Sickle cell disease. Postgrad. Doc. (Afr.) 1981; Oct.: 324-300.
6. Diggs LW. Sickle cell crises. Am. J. Clin. Pathol. 1965; 44: 1-15

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