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Case Report

Are spontaneous epidural haematomas a rare complication in sickle cell disease? A report of two new cases

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Summary

Background. Sickle cell anaemia, an autosomal recessive disease relatively common among the black races, gives rise sometimes to neurological complications. Among these, spontaneous epidural haematoma constitutes a rare event that is not always easy to treat in the Third world conditions.

Methods. Two new cases are described and their pathology is compared with the five already described cases in the literature. A vaso-occlusive pathological process as in the orbital compression syndrome is thought to be implicated in the generation of the spontaneous epidural haematoma.

Results. When facing an epidural haematoma as a complication of sickle cell disease in a hospital of the Third world conditions, a cautious attitude towards surgery should be observed because of the high complication rate.

If the relation between the haematoma and the anaemia is not immediately apparent, we are in favour of starting treatment with antibiotics.

Keywords: Sickle cell disease; orbital compression syndrome; spontaneous epidural cerebral haematoma.

Introduction

Sickle cell anaemia or drepanocytosis is an autosomal recessive hereditary haemoglobin disorder characterised by the production of haemoglobin S, in which a substitution of a valine for glumatic acid as the sixth amino acid in the B-globin has occurred. Sickle cell anaemia has a well known spreading among the black race with an incidence up to 7,3 percent [1]. The prevalence of the S gene may reach up to 40 percent in the subtropical parts of Africa such as the Congo [2].

Not only do homozygous patients suffer from typical chronic haemolytic anaemia, elevated susceptibility to infections, painful vaso-occlusive crises, but also from neurological complications, which are present in 6 to 34% of cases according to the different series [1]. Cerebral lesions of ischemic origin account to 75 percent of the neurological manifestations, the remaining 25% have a haemorrhagic mechanism, mainly intracerebral and subdural haematomas. Epidural haematomas seem to be rare, as we have only found five cases [3, 5–7, 9] mentioned in the literature. During our 5-year (93–98) stay in Kinshasa we have encountered two more cases. These cases will be described and discussed with special attention to management of such cases in Third world conditions.

Case 1

I.B., a male 2 year-old black Congolese child, known in our paediatric department for having homozygous sickle cell disease, was admitted to our Intensive Care Unit in a state of pronounced asthenia, with abdominal distension, oedema and unilateral palpebral ecchymosis, which all appeared during the course of an attack of bronchopneumonia. There was no history of known trauma. That same day the child developed progressive deterioration of consciousness along with bilateralisation of the palpebral ecchymosis, despite symptomatic treatment, consisting in the administration of oxygen, analgesics, transfusion, intravenous rehydration and antalgic posturing.

Clinical examination showed an apathic, comatose child, with little response to pain stimuli, important mucocutaneous pallor, and a right-sided exophthalmic eye with anisocoria: the right pupil being more

dilated than the left, but still photoreactive. Furthermore paralysis of the right external eye muscle was present. An urgent blood analysis with coagulation tests (PTT: 70%) was done, which did not show any anomalies except for a leucocytosis of 22,000 wbc/mm^3. An emergency CT-scan revealed the presence of an important right-sided frontotemporal epidural haematoma with a small retro-orbital extension and a midline shift. Consecutively trepanation was done with evacuation of an epidural haematoma. The patient died nevertheless in the following twenty-four hours due to deficient coagulation with melena and petechiae. Bone imaging could not be done.

Case 2

L.M., a 12 year-old black Congolese boy, with known homozygous sickle cell disease, was admitted to our Paediatric Department because of the recent onset of headaches, fever and joint pain. An infarctive crisis of

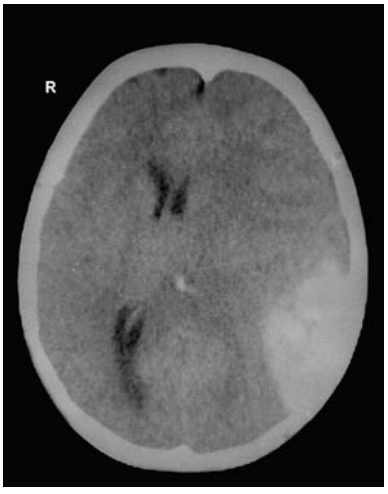

Fig. 1. Case no 2: CT-scan after 15 days. Picture of large left-sided epidural haematoma

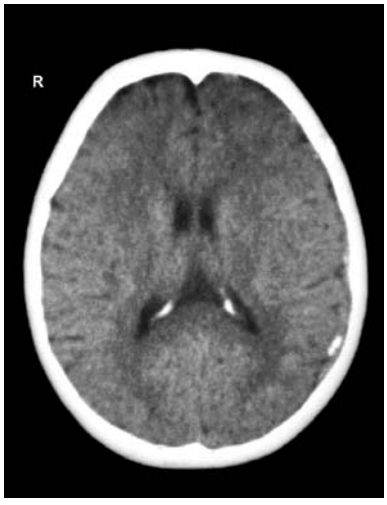

Fig. 2. Case no 2: CT-scan after 3 months. Nearly complete remission of the hematoma

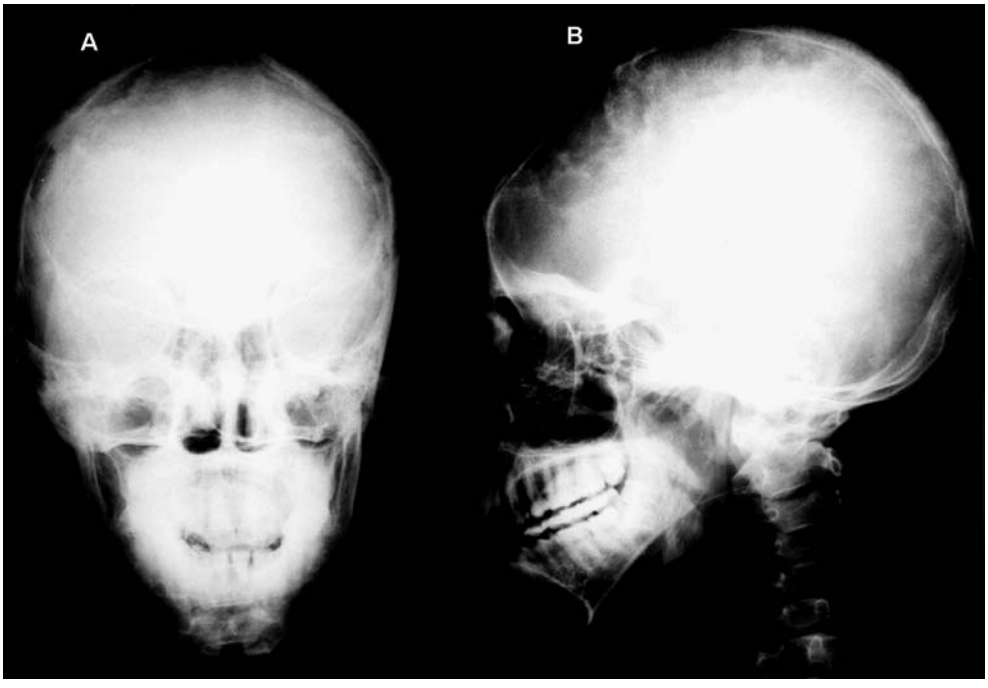

Fig. 3(A and B). Case no 2: RX 6 months later shows infarction of the bones in the fronto-parietal vault

Table 1. *Summary of reported cases of spontaneous epidural haematomas and sickle cell anaemia*

Authors & year	Sickle cell anaemia	Skull infarction	Orbital compression syndrome	CT/MRI	Epidural haematoma
Mallouh *et al*. 1987	+	+	+	+	+
Karacostas *et al*. 1991	+	?	+	+	+
Resar *et al*. 1996	+	+	−	+	+
Cabon *et al*. 1997	+	+	−	+	+
Naran *et al*. 2001	+	+	+	+	+
Kalala *et al*. 2003	+	?	+	+	+
Kalala *et al*. 2003	+	(+?)	−	+	+

the ''hand-foot'' type was anticipated; the clinical history did not reveal any trauma. The boy was hospitalised for observation, during which the headaches progressively increased. Physical examination revealed apathy and a right sided hemiparesis, scored 2/5, associated with a hyperreflexia. Except for anaemia with a haemoglobin level of 8.3 g/dl, the laboratory blood and coagulation tests did not show any abnormal values. Symptomatic treatment was initiated consisting in transfusion, rehydration, analgesics (Paracetamol) and anti-inflammatory drugs. A CT-scan was requested, but could not be performed due to practical problems at that time. After fifteen days of observation his paresis gradually disappeared. In the meantime a CT-scan proved the existence of a large left-sided epidural haematoma (Fig. 1). Due to the favourable evolution of the child's status, the parents refused surgical intervention. A control CT scan three months later confirmed the nearly complete resolution of the haematoma (Fig. 2). Because of recurrence six months later of the headaches with this time painful scalp-swelling, skull radiography was taken and revealed a large frontoparietal bone infarction (Fig. 3). Puncture of a subperiostal collection revealed pus. The patient died during the course of the week following the puncture due to septicaemia in spite of antibiotherapy and drainage of this collection.

Discussion

Epidural haematomas are mostly due to a lesion of the middle meningeal artery or one of its branches due to a skull fracture. In some cases venous bleeding due to the fracture or damage to a dural sinus can cause the epidural haematoma. The cases here described have in no way a traumatic origin but seem to be related to drepanocytosis.

Osseous infarctions may be a complication of drepanocytosis but occur mostly in the long bones. An infarction of the cranial bones is an unusual localisation by itself. In recent years some cases of skull infarcts have been described [10] in sickle cell syndromes. Among these a more precise entity concerning the orbits, initially described as orbital apex syndrome, now more often described as orbital compression syndrome (OCS), has been studied [4]. It is supposed to be a vaso-occlusive process in the marrow space around the orbits which results in bone infarction accompanied by frontal headache, fever, eyelid oedema all leading eventually to an orbital compression syndrome. A similar process with an irrigation fault related to ''sickling'' and venous drainage insufficiency leading to oedema and haemorrhage such as in the OCS could be considered in the cranial vault bones as demonstrated by our two cases. This could be at the origin of epidural haematomas, although the mechanism of the epidural bleeding is not so evident in the five cases described in the literature.

Plain radiography, bone and bone marrow scans, CT-scan, and MRI help to establish the diagnosis. In the early stage plain RX are of no help in the absence of a trauma history though they may serve as starting point for the further follow-up. Bone and bone marrow scanning, particularly the combination of both of them, are helpful and sensitive in distinguishing bone infarction from osteomyelitis [10], even when no abnormality is seen on physical examination [9]. This is of vital importance due to the fact that if infarct events are self-limiting and just require supportive therapy, on the other hand infection would call for aggressive treatment with antibiotics and eventually surgery. CT-scan and MRI delineate more precisely the necrotic territory and the relationship with the underlying brain.

In our third world conditions, where MRI and very often CT-scan are not available, it is impossible to come (quickly) to a correct diagnosis of bone infarction. Our first case had probably this pathology but the second case had subperiostal pus. This case had a biphasic

history and it is possible that here a primary infarction became with time infected considering the elevated susceptibility for infection in patients with sickle cell anaemia.

In our third world working conditions it seems reasonable to treat all these cases with antibiotics ab initio. When an epidural hematoma is present conservative treatment is preferable as those hematomas can absorb spontaneously; in addition the rates of complications of surgery are high. When signs of intracranial hypertension are present, then surgery becomes mandatory.

In reviewing the literature we have found only five descriptions of epidural haematomas in sickle cell patients and this was intriguing for somebody who has been working in an area where sickle cell anaemia is so frequent. Among the seven described cases at present three of them are from the Congo. The more likely explanation would be the fact that in these areas a CT-scan (and MRI still more) for diagnosis is accessible to less than 5% of the population. Apart from plain RX, CT-scan MRI and bone scans are most of the time not available. Furthermore the patients come from the poorest social class where antenatal diagnosis is not practiced and the families do not permit autopsies. Probably the diagnosis of this complication will become more common in the future with diagnostic facilities becoming more available.

The life expectancy of these patients is per se already lower than in the normal population [8]. In this context the occurrence of an epidural haematoma can prove fatal. The importance of antenatal diagnosis and counselling has to be emphasized too.

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Comments

This paper is original in reporting an unusual complication of sickle cell disease and will add to the literature of this rather common disorder in certain countries.

Khalaf Al Moutaery

I must confess that I was not entirely aware of this rare disease which is so frequent in the subtropical part of Africa. The occurrence of epidural haematomas in Sickle Cell disease is extremely rare, with only 7 cases reported so far.

This brief and interesting report describes this rare association. Even if the presence of the epidural haematoma did not change the course of the disease, there are some important findings in this report:

1) In some parts of the world access to CT scan is limited to less than 5% of the population. Still skull x-ray has a role to differentiate bone infarction from bone infections. I think that for us as Europeans it is a great lesson that we need to consider whenever we need more and more sophisticated tools for our diagnosis.
2) The epidural haematoma reported, Figs. 1 and 2, is, to my knowledge, the largest epidural collection ever published to reabsorb spontaneously. The diagnosis was made late in a state of clinical amelioration so surgery was not performed. It is interesting to see how minimal was the residual collection on follow up CT (see Fig. 2). We may ask ourselves how many epidural haematomas (both spontaneous and post traumatic) are treated in this way without the knowledge of the haematoma itself in the areas of the world where access to radiology is limited.

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