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 glutamic 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/mm³. 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