Nontraumatic Extradural Haematoma: A Rare Complication Of Sickle Cell Disease, Report Of Two Cases In Togo

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Abstract

Sickle cell anaemia, an autosomal recessive disease relatively common among the black races, gives rise sometimes to neurological complications. Non traumatic spontaneous extradural haematoma is a rare complication of sickle cell disease. We report two cases of that spontaneous extradural haematoma with good evolution after surgery.

INTRODUCTION

Sickle cell disease is a qualitative hereditary hemoglobinopathy due to the presence of hemoglobin S.1 Its incidence is high in Africa and in African American populations.1,2 The prevalence of the S gene may reach up to 40 percent in the subtropical parts of Africa.3 In Togo, sickle cell is a real public health problem with 16% of incidence and 10% of mortality.4

According to differents series, 6 to 34%, homozygous patients suffer from typical chronic haemolytic anemia, elevated susceptibility to infections, painful vaso-occlusive crises, and neurological complications.5 Cerebral lesions of ischemic origin account to 75 % of the neurological manifestations, the remaining 25% have a haemorrhagic mechanism, mainly intra cerebral and subdural hematomas.6,7,8 Spontaneous intracranial extra dural hematoma (EDH) is a rare complication of sicle cell disease. We report two cases of spontaneous EDH with special attention to management of such cases in non developed countries.

CASE 1

Female patient, 22 years old, togolese , known for having homozygous sickle cell disease, was admitted to intensive care unit, with seven days of frontal headache and alteration of consciousness. These symptoms were preceded by an episode of vaso-occlusive crisis. There was no history of trauma. Clinical examination showed a Glasgow coma scale at 11 (E:3-4; V:2; M: 6), a left side hemiparesis scored 2/5. An emergency CT scan revealed a right frontal EDH. There was no skull fracture [Figure 1]. The patient underwent a right frontal craniotomy and evacuation of the EDH. In post-operative, the patient had a good recovery with complete resolution of headache and of the hemiparesis. Post-operative CT-scan showed complete evacuation of hematoma [Figure 2].

Figure 1

Brain CT scan showing frontal acute extradural haematoma (case 1)

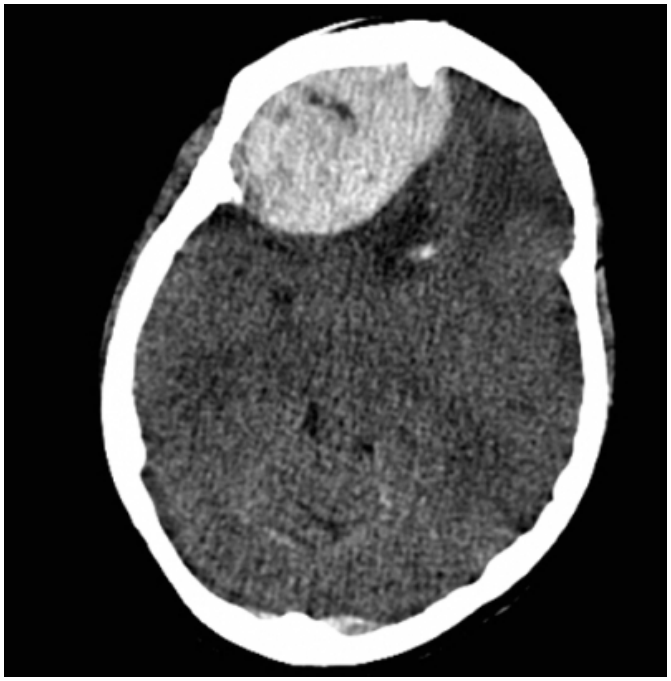
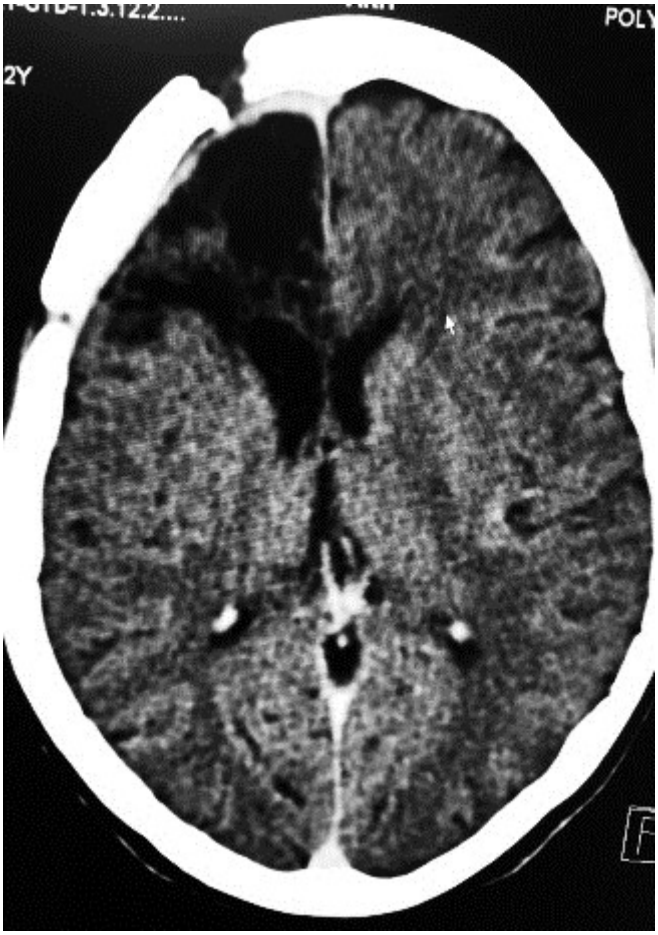


Figure 2

Post operative CT scan showing complete evacuation of haematoma



CASE 2

Female patient, togolese, 14 years old, known in pediatric department for having homozygous sickle cell disease, was admitted in pediatric intensive care unit, for recent headaches, fever, and joint pain. An infarctive crisis of the “hand - foot” type was anticipated. There was no history of trauma. Physical examination revealed apathy and no neurologic deficit. During the observation in the intensive care unit, headaches increased progressively and a left sided hemiparesis scored 3/5 appeared. A CT-scan was requested and proved a right frontal EDH [Figure 3]. Patient underwent right frontal craniotomy with evacuation of the hematoma. Evolution was good, with resolution of the headaches and hemiparesis. Post-operative CT-scan showed complete evacuation of hematoma [Figure 4].

Figure 3

Brain CT scan showing frontal acute extradural haematoma (case 2)

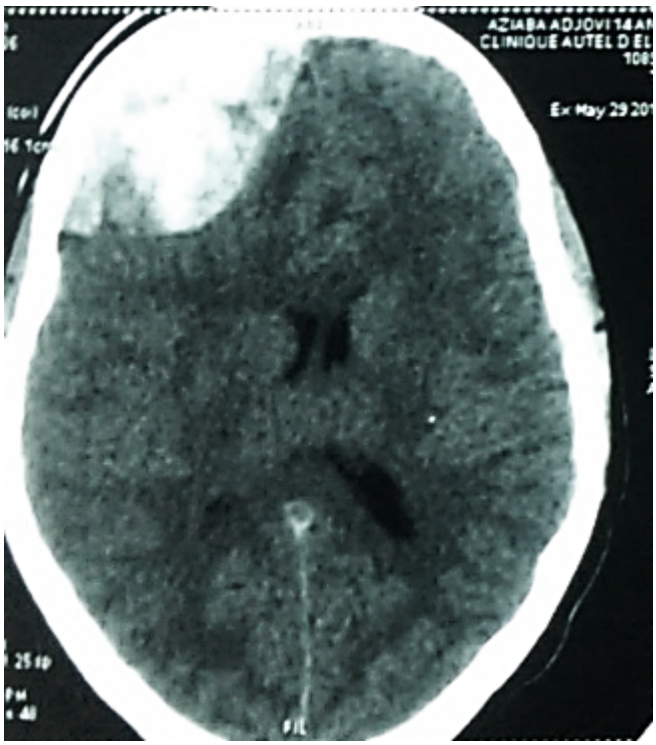
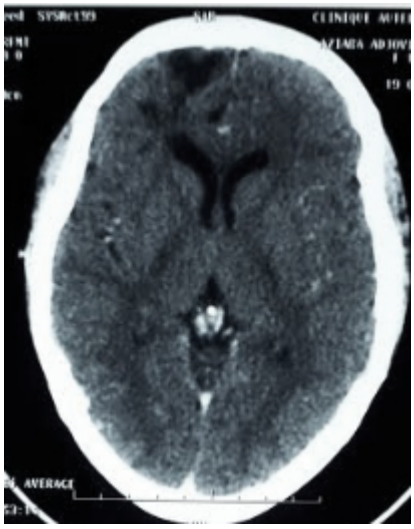


Figure 4

Post operative CT scan showing complete evacuation of haematoma (case 2)



DISCUSSION

Spontaneous EDH is rarely reported and its incidence is not known. Spontaneous EDH in a patient with sickle cell disease represent a rare and frequently unmentioned complication of this haemoglobinopathy.9 Few cases of spontaneous extradural hematomas due to sickle cell have been reported.2,3,6,10

Spontaneous EDH in patients with Hb SS occurred along with skull infarctions.2,7,10,12,13,14 Osseous infarctions may be a complication of drepanocytosis but occur mostly in the long bones. An infarction of the cranial bones is an unusual localization by itself.15 A microvascular diploic vaso-occlusive mechanism associated with occlusion of an emissary vein found in a sickle cell disease patient was suggested to be responsible for a spontaneous EDH.2,16 Another theory proposes that insufficient venous drainage is the inciting event that leads to oedema and haemorrhage.2

The clinical presentation of EDH in sickle cell disease patients is different from the classic description of post traumatic extradural hematoma which is usually characterized by a lucid interval.9 Our patients presented with persitent headache without any trauma.

CT scan and MRI help to establish the diagnosis. They delineate more preciously the necrotic territory and the relationship with underlying brain. In our developing countries, where MRI and CT scan are not usually available, it is impossible to come to a correct diagnosis of bone infarction. For our patients, there was no evidence of skull infarction on CT scan, and during the craniotomy, the skull was normal. Bankole O et al. described the same observations in a boy of 18 years old.9 The exact cause of the EDH in our patients is unclear.

Our patients underwent standard craniotomy and evacuation of EDH. Surgical evacuation remains the standard of treatment for this condition of EDH, and the presence of sickle cell haemoglobinopathy should not serve as a contradiction.9

Despite the high prevalence of sickle cell disease in Togo (16%), our patients are the first cases described. This is because CT scan machines are not available all over the country and it is not affordable for many togolese patients.

CONCLUSION

Spontaneous EDH is rare in patients with sickle cell disease. A high suspiscion is required among clinicians, in our developing countries where sickle cell disease prevalence is high, to make prompt diagnosis of this rare condition. Any sickle cell disease patient with symptoms and signs of raised intracranial pressure should be referred for cranial CT scan which will diagnose EDH in all cases where it is present.

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