Case Report

Non-traumatic spontaneous acute epidural hematoma in a patient with sickle cell disease

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\textbf{A B S T R A C T}

A 19-year-old female with sickle cell anemia (SCD) was referred to our hospital after two days of hospitalization at another hospital for a headache crisis. This headache crisis was due to a raised intracranial pressure; these symptoms were noted and included in her comprehensive list of symptoms. There was an acute drop in the hemoglobin and hematocrit levels. The cranial CT scan demonstrated a left fronto-parietal acute epidural hematoma (AEH) and a calvarial bone expansion, which was suggestive of medullary hematopoiesis. The patient underwent emergent craniotomy and evacuation of the hematoma. There were no abnormal findings intra-operatively apart from the AEH, except skull thickening and active petechial bleeding from the dural arteries. Repeated CT scan showed a complete evacuation of the hematoma. The possible underlying pathophysiological mechanisms were discussed. In addition to the factors mentioned in the relevant literature, any active petechial bleeding from the dural arteries on the separated surface of the dura from the skull could have contributed to the expanding of the AEH in our patient. Neurosurgeons and other health care providers should be aware of spontaneous AEH in patients with SCD.

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\textbf{Hematoma epidural agudo espontáneo no traumático en una paciente con enfermedad de células falciformes}

\textbf{R E S U M E N}

Una mujer de 19 años con anemia de células falciformes (anemia drepanocítica) fue remitida a nuestro hospital tras 2 días de hospitalización en otro centro, debido a una crisis de migraña. Dicha crisis de migraña fue debida a un incremento de la presión intracraneal; se anotaron dichos síntomas y se incluyeron en su amplia lista de síntomas. Se produjo un pico agudo en los niveles de hemoglobina y de hematocrito. El escáner craneal mostró un hematoma epidural agudo fronto-parietal izquierdo y una expansión del hueso de la bóveda craneal, que indicaba una hematopoyesis medular. Se sometió a la paciente a una
cranietomía de urgencia y a una evacuación del hematoma. Aparte de la anemia de células falciformes, los hallazgos intraoperatorios fueron: adelgazamiento craneal y petequia activa de las arterias durales. La repetición del escáner mostró la evacuación total del hematoma. Se analizaron los posibles mecanismos patofisiológicos subyacentes. Además de los factores mencionados en la literatura relevante, cualquier sangrado petequial activo de las arterias durales sobre la superficie separada de la duramadre, desde el cráneo, podría haber contribuido a la expansión del hematoma epidural agudo en nuestra paciente. Los neurocirujanos y demás facultativos sanitarios deberían tener conocimiento del hematoma epidural agudo espontáneo en pacientes con anemia de células falciformes.

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Introduction

An epidural hematoma is classically associated with head trauma. The spontaneous acute epidural hematoma (SAEH) without trauma is rarely reported in literature and is usually associated with infectious diseases. Other causative factors are the presence of coagulopathy, dural vascular malformations, hemorrhagic tumors, open heart surgery, hemodialysis, systemic lupus erythematosus, and sickle cell disease (SCD). SCD is a chronic progressive hemolytic disease, and its neurological complications are mostly due to ischemic events. Extraaxial hemorrhagic complications are very rare in SCD.1–4

We present a case of SAEH occurring in a patient with SCD requiring emergent surgical evacuation and discuss possible underlying mechanisms.

Case report

A 19-year-old female with SCD was admitted to another hospital in our city to receive treatment for a headache crisis. She was on vitamin B12, folate, and hydroxyurea. Her headache did not improve during the two days of hospitalization. Nausea and vomiting were added to her symptoms. She was referred to the hematology department of our university. On admission, her laboratory results were the following: white blood cell count (WBC) 4250 cells/mm³, hemoglobin (Hb) level 7.2 g/dL, hematocrit (Htc) 21.8%, and platelet count 142,000 platelets/mm³. Two units of erythrocyte suspension were transfused.

Her Glasgow Coma Scale score was 13 upon her arrival to our hospital. There was no history of any head trauma. A complete blood count (CBC), prothrombin time, activated partial thromboplastin time, international normalized ratio (INR), full chemistry panel, liver function tests, and a cranial CT scan were obtained. The coagulation profile was normal. Her WBC was 5070 cells/mm³, Hb level 9.48 g/dL Htc 27.6%, and her platelet count showed 107,000 platelets/mm³. The remaining laboratory tests were normal.

The CT scan revealed a left parietal AEH (Fig. 1A and B). The bone window of the CT scan revealed a calvarial bone expansion that was suggestive of medullary hematopoiesis (Fig. 2). An emergent craniotomy and epidural hematoma evacuation were performed. There were no abnormal findings intra-operatively apart from the epidural hematoma, except the thickening of the bone and active petechial bleeding from the dural arteries on the separated surface of the dura from the skull. The hemostasis was achieved using bipolar cautery on the dural surface. The bone flap was replaced because it had a normal color and consistency. A repeated CT scan showed the complete evacuation of hematoma (Fig. 3A and B). The patient was discharged with a normal neurological examination on the fifth postoperative day.

Discussion

Epidural hematomas are generally due to skull fractures. The hemorrhage arises from the middle meningeal artery or one of its branches, the fracture edges, torn venous sinuses or diploic veins. SAEH have been rarely reported in the literature, and one of the reasons for the occurrence of SAEH is SCD.1,3,5

Kotb et al.2 reported the brain MRI and CT findings in patients with SCD. Of the 36 patients with SCD, only one patient (a 10-year-old boy who presented with a headache, left hyperreflexia, and a fever) had an epidural hematoma associated with skull bone infarctions and scalp edema.

Arends et al.6 reported an adult man who presented with a relapsing severe headache caused by an uncommon complication of SCD. The brain CT scan was normal on his first presentation. Eight days later, the patient presented a second time with a relapsing severe headache. This time, the MRI showed an infarction located in the parietal skull bone with a small adjacent epidural hematoma. He was successfully managed without any surgical intervention. The authors argued that the first headache episode was thought to be due to the initial skull bone infarction because no epidural hematoma had been present initially. The second headache episode was thought to be due to the development of the epidural hematoma.

There are a few reports of spontaneous epidural hematomas (SEH) occurring in patients with sickle cell anemia.5,7–13 To explain the underlying mechanisms of SEH, the authors emphasized the necrosis of the bone marrow, infarction of calvarial bones, the tearing of small vessels and the resulting hemorrhage into the epidural space during the vaso-occlusive crises in these patients. The authors based their theory on evidence that was suggestive of the infarction of the skull (a devascularized, discolored, edematous and deformed skull). Additionally, an insufficient venous
drainage could be the inciting event that results in edema and bleeding.³

Dahdaleh et al.¹ presented a case of multiple spontaneous epidural and subgaleal hematomas in a patient with SCD. The patient underwent emergent surgical evacuation because of the rapid deterioration of the patient’s neurological condition. In their patient, it appeared that the hematomas developed concurrently with an acute and dramatic drop in the hematocrit level and platelet count. The authors asserted that the hematomas unlikely resulted from an ischemic event because no intraoperative evidence of skull bone infarction was observed. They proposed an alternative pathophysiological mechanism based on the histopathological evidence of the hematopoietic tissue within the patient’s hematoma. Their theory was that the anatomy of the patient’s skull was abnormal due to a pathological chronic medullary hematopoiesis. In the patient, the acute drop in the hematocrit level may have triggered such a reaction and resulted in the expanding hematopoietic tissue that disrupted the internal and external skull border; this led to the extravasation of blood and hematopoietic tissue into the subgaleal and/or epidural space. The violation of the skull margins may have occurred at the microscopic level if the bone flap appeared during the gross intraoperative examination. The coagulopathy due to the drop in the platelet count may also have been a predisposing or aggravating factor.¹ Bölke and Scherer¹⁴ agreed with Dahdaleh et al.¹ about the underlying mechanism of SEH, i.e., it was due to a space-occupying epidural hematoma that was most likely caused by an expansion of hematopoietic tissue and the disruption of the bony margins with bleeding into the epidural space.

Our patient had a similar clinical condition to the patient in the case report by Dahdaleh et al.¹, differing only in the observed rapid deterioration of the patient’s neurological condition. In our patient, a drop in the Hct and Hb levels were observed in the initial admission to the hospital. Two units of packed red blood cells were transfused. The second CBC revealed a drop in the platelet count; however, it did not reach hazardous levels. The platelet count was within normal limit and the transfusion of packed red blood cells may have protected the patient from neurological decline. This condition may be evidence of coagulopathy secondary to a drop in the platelet count contributing to bleeding into the epidural space.

We did not detect discoloration or deformation of the bone flap. Its consistency was not suggestive of an infected or infarected skull. Our findings and observations were similar to those in the case report by Dahdaleh et al.¹ and endorse their theory. In addition, active petechial bleeding from the dural arteries on the separated surface of the dura from the skull could be a facilitating factor to the expansion of the epidural hematoma in our patient.

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Fig. 1 – (A) and (B) An axial and a coronal brain CT showing a large left parietal, acute epidural hematoma that includes a mixed CT density clot, effacement of cortical sulci and the left lateral ventricle. There is no evidence of subgaleal hematoma or skull fracture.

Fig. 2 – An axial CT with a bone window that shows diffuse marrow proliferation that affects the calvarium and the widening of the diploic space.
Fig. 3 – (A) and (B) The postoperative axial and coronal CT that shows the complete evacuation of the epidural hematoma, the reappearance of the cortical sulci and the left lateral ventricle.

Conclusion

Neurosurgeons and others medical care providers should be aware that a patient who has a severe headache in SCD may have SAEH and may thus require surgical evacuation, especially during an acute drop in Hct and Hb levels.

References