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Case Report

Extramedullary hematopoiesis of the paranasal sinuses associated with moyamoya syndrome in sickle cell disease☆

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ABSTRACT

Sickle cell disease is a common blood disorder with well-known clinical presentations including extramedullary hematopoiesis and moyamoya syndrome. However, it is extremely rare for extramedullary hematopoiesis to occur in the paranasal sinuses. Here, we present a case of a child with known homozygous sickle cell disease who has extramedullary hematopoiesis within the maxillary and the sphenoidal sinuses associated with moyamoya syndrome.

1. Introduction

Sickle cell disease (SCD) is a common hemoglobinopathy resulting in abnormally shaped red blood cells [1]. A genetic anomaly causing an amino acid substitution in the beta-globin chain is responsible for the abnormal hemoglobin structure. A patient with only one abnormal hemoglobin gene is defined as sickle cell trait. This heterozygous form is usually asymptomatic. If two abnormal hemoglobin genes are present in a patient, homozygous SCD occurs [1]. Patients with SCD may have various well-known clinical manifestations including extramedullary hematopoiesis (EMH) and moyamoya syndrome.

Extramedullary hematopoiesis, a result of increased demand for blood production, is usually observed in the spleen, liver and lymph nodes [1]. Sinonasal tract involvement for EMH in a patient with SCD is a rare entity with only a few case reports in the literature [2,3].

Moyamoya disease is a progressive arteriopathy causing narrowing of distal internal carotid artery and proximal circle of Willis vessels with secondary collateralization [4]. This condition is defined as moyamoya syndrome if associated with other disorders such as SCD [4]. Moyamoya syndrome is a significant cause of cerebrovascular stroke in children. It is, therefore, important to make a prompt diagnosis of this vasculopathy in patients with SCD in order to improve long term prognosis [4].

In this case report, we present magnetic resonance imaging (MRI) and diffusion weighted imaging (DWI) findings of EMH within the maxillary and the sphenoidal sinuses in a patient with SCD who has concomitant moyamoya syndrome. To the best of our knowledge, this is the first report of EMH of the paranasal sinuses associated with moyamoya syndrome in a patient with SCD.

2. Case report

A 10-year-old male with known homozygous SCD presented to the pediatric hematology clinic of our institution for intermittent headache in the last two months. His past medical history was uneventful except a few acute crises. He had no epistaxis, nasal obstruction, rhinorrhea or visual changes. Physical examination including a detailed neurological evaluation revealed no abnormality.

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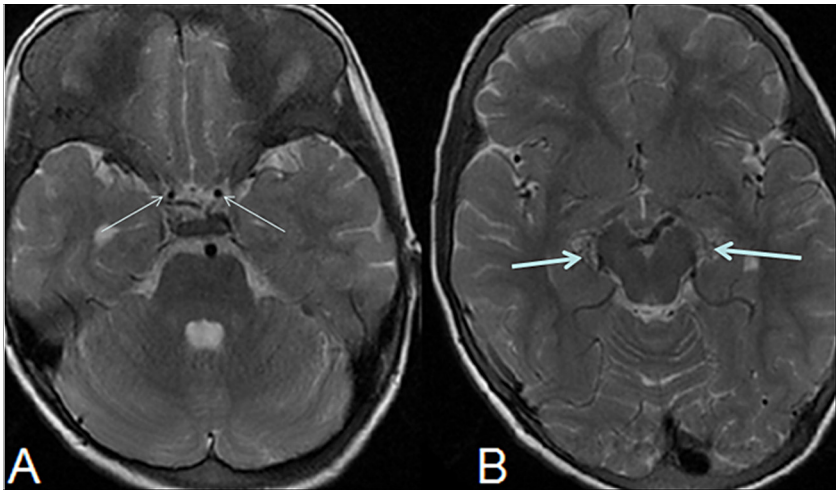

Fig. 1. Axial T2W image at suprasellar level (A) shows tapering of both distal internal carotid arteries (arrows). Axial T2W image at cerebral peduncle level (B) shows collateral vessels within the ambient cistern (arrows).

Cerebral MRI demonstrated narrowing in both distal internal carotid arteries and abnormal thin vessels within the ambient cistern (Fig. 1). Fluid-attenuated inversion recovery (FLAIR) image showed sulcal hyperintensities consistent with leptomeningeal vascular engorgement along the cerebral convexities (Fig. 2). Cerebral parenchyma, cerebellum and brain stem were normal. DWI demonstrated no parenchymal abnormality. Intracranial imaging findings were found to be consistent with moyamoya syndrome.

Cerebral MRI also demonstrated symmetric homogeneous soft tissue masses filling the entire cavities of both maxillary sinuses (Fig. 3). A sphenoidal mass with similar signal characteristics to those seen in both maxillary sinuses was also present (Fig. 4). The bone marrow presented low signal intensity on both T1-weighted and T2-weighted images compatible with the red marrow reconversion induced by sickle cell anemia. The masses demonstrated signal intensity similar to those of red marrow within the craniofacial bony structures on all pulse sequences. DWI showed restricted diffusion both within the masses and craniofacial bone marrow with similar signal intensity (Figs. 5 and 6). Ethmoidal sinuses and nasal cavity were normal. The masses within the maxillary and sphenoidal sinuses were diagnosed as EMH on the basis of MRI and DWI findings. No histopathological examination or surgical procedure was intended due to the presence of typical imaging findings and the lack of directly related symptoms.

3. Discussion

Clinical manifestations of SCD usually develop secondary to vaso-occlusion, chronic anemia and infection [1]. Chronic hemolytic anemia leads to an increased demand for blood production. EMH, increased cardiac output and high blood flow velocity develops to compensate the increased blood demand [1]. High flow velocity leads to increased flow turbulance especially in proximal portion of arteries and at sites of bifurcation. High flow turbulance associated with the adherence of sickled erythrocytes to the endothelium leads to the intimal damage in large intracranial vessels resulting in vascular stenosis and occlusion [5]. Therefore, both EMH and moyamoya syndrome may be attributed to the chronic hemolytic anemia in patients with SCD.

EMH most commonly occurs in chronic hemolytic anemias such as thalassemia and SCD [6]. This physiological compensatory phenomenon may also be seen in hematological conditions such as myelofibrosis, leukemia and lymphoma. Although EMH most commonly occurs in the spleen, liver and lymph nodes, all body sites may be involved mainly in the form of masses [6]. Paranasal sinus involvement by EMH is an exceptional entity which may be confused with the other masses or masslike conditions of the sinuses including tumors. EMH in the paranasal sinus is believed to occur secondary to the herniation of medullary tissue into the sinus lumen due to the underlying bone marrow hyperplasia [1].

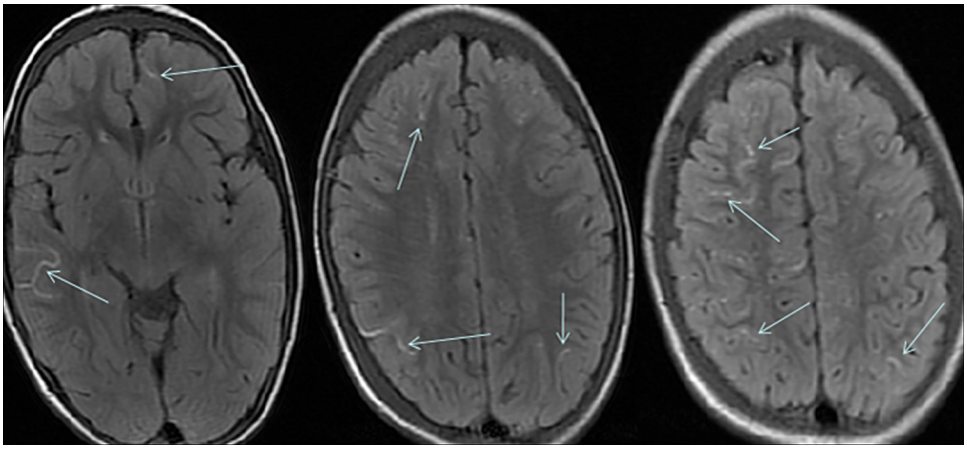

Fig. 2. Axial consecutive FLAIR images show bright sulci (arrows) consistent with slow flowing engorged pial vessels (ivy sign).

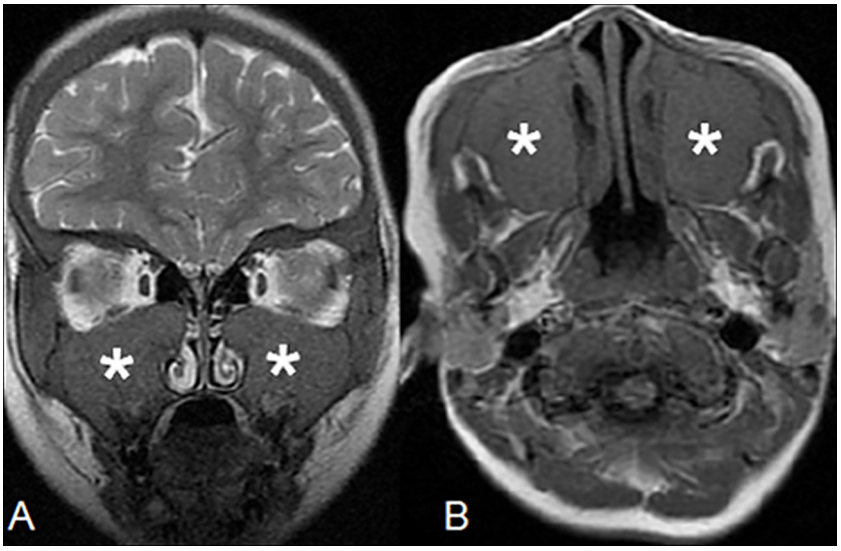

Fig. 3. Coronal T2W (A) and axial T1W (B) images demonstrate homogeneous masses (asterisk) occupying both maxillary sinuses with signal intensity similar to those of craniofacial bone structures.

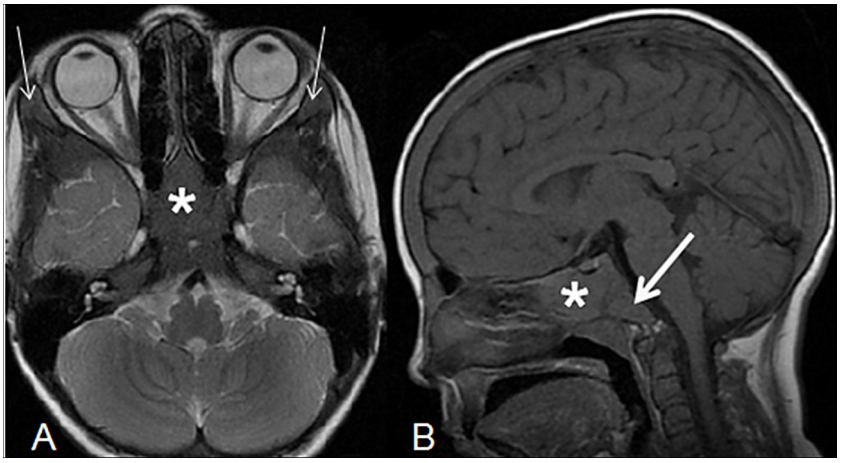

Fig. 4. Axial T2W (A) and sagittal T1W (B) images show a mass filling the sphenoidal sinus (asterisk). Note the mass has similar signal intensity with the orbital wall (thin arrows) and the adjacent clivus (thick arrow).

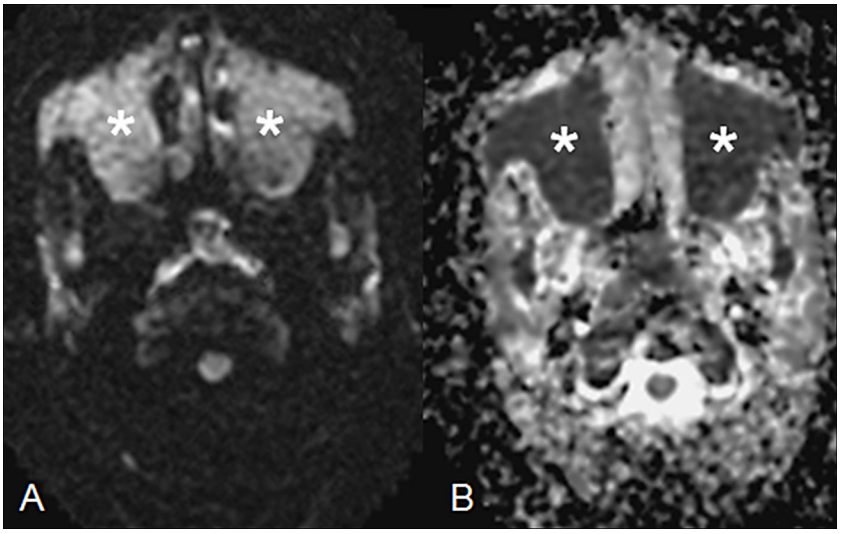

Fig. 5. DWI with trace image (A) and corresponding ADC map (B) shows diffusion restriction within the masses (asterisk) of both maxillary sinuses.

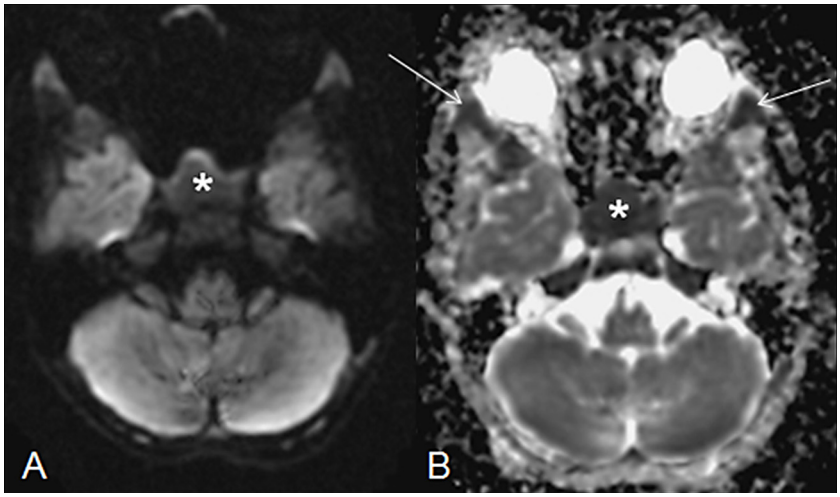

Fig. 6. DWI with trace image (A) and corresponding ADC map (B) also demonstrate diffusion restriction within the sphenoidal mass (asterisk). Note the mass has similar signal intensity with the orbital wall (arrows).

Maxillary sinus is the most commonly affected site in the sinonasal tract. Patients with EMH in the paranasal sinuses are often asymptomatic [1]. Therefore, the mass within the sinus lumen is usually discovered incidentally in patients with sinonasal EMH as it occurred in our patient.

Imaging findings are helpful in the diagnosis of EMH in the paranasal sinuses. On CT, EMH usually appears as an expansile mass involving the paranasal sinuses with attenuation similar to those of red marrow. Findings such as intralesional central high density or preserved septations within the sinus occupied by the mass have also been reported in the literature [3,7]. The mass shows signal intensity and diffusion pattern similar to those of red marrow on MRI and DWI, respectively, as it occurred in our patient [1]. On contrast-enhanced images, the enhancement of EMH is usually similar to that of red marrow and relatively more intense than that of normal bone marrow due the increased vascularity of red marrow [1].

Moyamoya syndrome is seen in up to 35% of patients with SCD at conventional angiography [5]. Although conventional angiography is the gold-standard technique to demonstrate moyamoya, a diagnosis based on MRI and magnetic resonance angiography without conventional angiography is recommended in children [4]. MRI findings include stenosis or occlusion of distal internal carotid arteries and proximal circle of Willis vessels, thalamoperforating and lenticulostriate collaterals presenting with thin vessels in cisterns, enhancing dots in basal ganglia and slow flowing engorged pial vessels consistent with leptomeningeal 'ivy sign'.

In conclusion, although the presence of EMH and moyamoya syndrome in SCD is well-known, EMH arising from paranasal sinus is a rare entity resulting in diagnostic dilemma. EMH should be considered in the presence of a homogeneous soft tissue mass in the paranasal sinus if the mass and the intramedullary red marrow have similar signal intensity and enhancement pattern.

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