SHORT REPORT

Peripheral Arterial Lesions in Patient with Sickle Cell Disease

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Vascular occlusion in sickle cell disease (SCD) is often considered to be synonymous with occlusion of microvasculature by sickled red blood cells. However, other mechanisms are also involved. One of these is intimal hyperplasia in the macrovasculature. This creates irregular areas of endoluminal narrowing, which may promote thrombogenic occlusion. This process has not been documented in the peripheral arteries. We report a 14-year-old boy with SCD who developed critical ischemia right foot with absence of atherosclerotic risk factors. Assessment of the patient revealed wide spread arterial disease in both upper and lower extremities.

Keywords: Sickle cell disease; Peripheral arterial disease; Arterial calcification.

Case Report

A 14 year-old boy was referred to our vascular unit, with gangrene of the right foot. The condition started about 1 year prior to this referral with ulceration of the foot which was treated conservatively. The condition of the foot deteriorated until development of gangrene of the most of the foot. The boy is a known patient of SCD. His past medical history revealed right sided stroke when he was 8 years old. His parents have SCD. His brother had also SCD and died suddenly at the age of 5 years.

There were no identifiable risk factors for atherosclerosis and sickle hemoglobin (Hb S) was 72%.

On examination, there were no palpable pulses. He was found to have heavily calcified femoral and brachial arteries. Plain x ray of both arms (Fig. 1A&B), femurs (Fig. 1C) showed extensive calcifications of brachial, femoral and popliteal arteries. An X ray of his right foot showed (Fig. 1D) showed infarction and osteomyelitis of most of the bones. Plain CT of the Abdomen and pelvis showed calcification of splenic artery (Fig. 2 A) and both renal arteries (Fig. 2: B), a solitary gall stone (Fig. 2 B) and calcifications of both iliacs (Fig. 2 C) and inferior mesenteric artery (Fig. 2 D). Digital subtraction angiography showed occlusion of right external iliac artery (Fig. 3 A) and both superficial femoral arteries (Fig. 3B) with extensive collaterals. MRI & MRA of the brain showed left parietal wedge area of infarction with total occlusion of the supraclinoid segment of left internal carotid artery and multiple collaterals. The patient had a right below knee amputation and was discharged home on antiplatelets.

Discussion

Sickle-cell disease is inherited disorders of Hb structure and synthesis and is characterized by the presence of Hb S. Acute, painful vaso-occlusive crises are the most common, and earliest, clinical manifestations of SCD and is usually caused by sickle-shaped red blood cells that obstruct capillaries and restrict blood flow to an organ, resulting in ischemia, pain, and organ damage. A recent study of spleens resected from SCD patients, due to episodes of sequestration or infarction, has shown that there was consistent vascular lesion affecting large arteries. The same finding was also shown in studies of brains from SCD patients who developed cerebrovascular accidents. These

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EJVES Extra 14, 15–18 (2007)
doi:10.1016/j.ejvsextra.2007.04.001, available online at http://www.sciencedirect.com on

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lesions were attributed to the rigidity of sickled erythrocytes causing mechanical injury to the endothelial cells. The widespread distribution of the lesions was also suspected in other studies, in which it was suggested that the sickled erythrocyte-endothelial adhesion seen in the microvasculature could be occurring in large arteries and contribute to large vessel endothelial injury, vascular intimal hyperplasia and thrombosis. The end result of the intimal injury is diffuse or focal intimal proliferation and/or disruption of the internal elastic lamina with or without medial hypertrophy or fibrosis.

There is little information about peripheral vascular disease in patients with SCD. The lack of awareness

Fig. 1. Plain x ray of both arms (Fig. 1A & B), femurs (Fig. 1C) showed extensive calcifications of brachial, femoral and popliteal arteries and the right foot (Fig. 1D).
of this condition was probably responsible for the delay in the diagnosis of the present case. Management of the vascular lesion is difficult. Medications or change in life style e.g. exercise are unlikely to control or improve the condition. The diffuse calcified nature of the vascular disease may make revascularization not only difficult but perhaps unrewarding as well.

Fig. 2. Plain CT of the Abdomen and pelvis: Axial sections showed calcification of splenic artery (Fig. 2 A) and both renal arteries (Fig. 2 B), solitary gall stone (Fig. 2 B) and coronal reformatted showed calcifications of both iliacs (Fig. 2 C) and inferior mesenteric artery (Fig. 2 D).

Fig. 3. Digital subtraction angiography showed block of right external iliac (Fig. 3A) and both superficial femoral arteries (Fig. 3B) with extensive collaterals.
References


Accepted 20 April 2007