Brain Hemorrhage as Presenting Feature of Takayasu's Arteritis in an African Girl

Aaron Ngamolane MBBS[1], Ludo Taboka Molobe MuDr[3], Kabo Mojela MBChB[3], Canuto Silava MD DPBR FUSP FCT-MRISP[2], Francesca Cainelli MD[1,3] and Sandro Vento MD[1]

[1]Department of Internal Medicine, University of Botswana, Gaborone, Botswana
[2]Village Imaging, Gaborone, Botswana
[3]Department of Medicine, Princess Marina Hospital, Gaborone, Botswana

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A 13 year old, HIV-negative African girl presented to Bamalete Lutheran Hospital in Ramotswa, Botswana with sudden-onset headache followed by generalized tonic-clonic seizures with foaming at mouth; blood pressure was 180/118 mmHg, pulse rate 85/min, and the patient was afebrile, comatose (Glasgow Coma Scale 7/15) and had a poor right conjugate gaze. She was transferred to Princess Marina Hospital in Gaborone where hypertension (192/118 mmHg) was confirmed on arrival and examination revealed bruits over the abdominal aorta. A brain computed tomography (CT) scan showed a large hemorrhage in the left parieto-occipital region and an electrocardiogram (ECG) was suggestive of left ventricular hypertrophy. White blood cells were 24,790/µl, platelets 488,000/µl, erythrocyte sedimentation rate 40 mm/hr, C-reactive protein 69.3 mg/L. The patient was treated with spironolactone 50 mg daily, nifedipine 90 mg per day, hydrochlorothiazide 12.5 mg per day, captopril 75 mg daily, mannitol 150 ml, phenytoin 200 mg daily, cefotaxime 3 g daily. Two days later the patient was no longer comatose and a bilateral abducens nerve palsy was noted; an echocardiogram did not show any abnormalities.

A CT scan showed a thoracic aortic aneurysm (2.9 x 3.2 cm) and right kidney reduced in size, with a possible stricture in the abdominal aorta. Magnetic resonance angiography confirmed a large hematoma in the left parieto-occipital region [Figure 1] and segmental stenoses in the descending thoracic and proximal abdominal aorta, with right renal artery stenosis and chronic renal changes [Figure 2]. Treatment with daily prednisolone (50 mg) and weekly methotrexate (2.5 mg) was started, and 2 days later the patient was able to ambulate with no focal deficits apart from 6th cranial nerve palsy. She is presently doing well on low dose prednisolone.

Takayasu's arteritis is a rare disease of unknown etiology characterized by medium and large vessel vasculitis which generally involves the aorta; it occurs more often in Asian women under the age of 40 years. The diagnosis can be made reliably, based on the criteria listed below [1]. The diagnosis can be made if at least three criteria are met; our patient met criteria 1, 5 and 6.

1. age at disease onset < 40 years
2. extremity claudication
3. decreased brachial artery pulse
4. difference of > 10 mmHg in systolic blood pressure between arms
5. subclavian or aortic bruit
6. arteriographic evidence of arterial narrowing or occlusion of the aorta, its primary branches, or large arteries of the upper or lower extremities not

Figure 1. Hematoma in the left parieto-occipital region of the brain

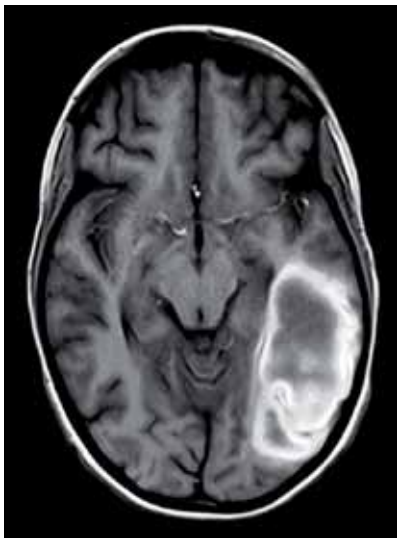
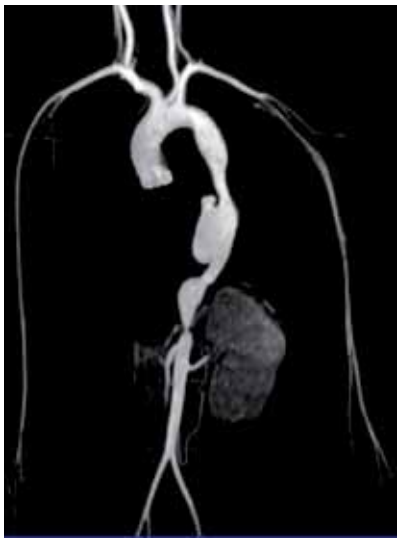


Figure 2. Segmental stenoses in the descending thoracic and proximal abdominal aorta with right renal artery stenosis and chronic renal changes



due to arteriosclerosis, fibromuscular dysplasia, or similar causes.

Renal artery involvement occurs in over 50% of cases [2,3] and can lead to renal failure and/or renovascular hypertension which is often resistant to medical therapy. Treatment is based on steroids and the effects are usually dramatic. However, the appropriate dose and duration of steroid treatment have not been established since no controlled clinical trials have yet been reported. In addition, it is unknown whether steroid therapy alters the outcome of the disease. Oral weekly methotrexate added to glucocorticoids induces remission in the majority of patients with glucocorticoid-resistant or relapsing Takayasu's arteritis [4]. Irreversible, symptomatic steno-occlusive vascular lesions are treated with reconstructive vascular surgery, whose long-term outcome seems to be better in children [5].

Correspondence
Dr. S. Vento
Dept. of Internal Medicine, University of Botswana, Private Bag 00713, Gaborone, Botswana
Phone: (267-3) 554-190
email: ventosandro@yahoo.it

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