Stroke - a rare initial manifestation of Takayasu’s arteritis

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Abstract:
Stroke as an initial manifestation is rare in Takayasu's arteritis, a condition affecting young females. Here we present a 28 year old female with acute onset of right sided weakness as an initial presentation. She had severe uncontrolled hypertension and angiogram revealed normal aortic arch vessels, occlusion of abdominal aorta and narrow renal arteries. Her CT brain showed moderate infarct in left capsuloganglionic region. Possible mechanisms of stroke in Takayasu's arteritis are discussed. Importance of screening for Takayasu's arteritis in young stroke is highlighted.

Keyword: Takayasu's arteritis, stroke, intracranial arterial stenosis

Takayasu's arteritis (TA) is a granulomatous vasculitis of unknown etiology that affects the aorta, its major branches and the pulmonary arteries leading to stenosis, occlusion or aneurysm. At the time of diagnosis, 10% to 20% of patients with Takayasu's arteritis are clinically asymptomatic. The remaining 80% to 90% of patients present as a result of symptoms that are systemic or vascular. Stenosis or occlusion of the carotid and vertebral arteries or the vessels proximal to their origin can be completely asymptomatic or present as transient ischemic attacks, stroke, dizziness, syncope, headache, or visual changes. Stroke at the onset is present in less than 10% of patients. Here we describe an unusual case of Takayasu’s arteritis in a young woman who presented with stroke as the initial manifestation.

CASE REPORT:
28 years old unmarried female presented with acute onset of weakness of right upper limb and deviation of angle of mouth to left side for one day before admission. She gave no history of loss of consciousness, seizures or sensory disturbances. There was no history suggestive of involvement of other cranial nerves or disturbances of higher functions. In the past 3 years the patient had suffered from headache which occurred 2-3 times in a week and was holocranial. There was no other significant past history.
and no history of constitutional symptoms, claudication pain, skin rashes, oral ulcers, bleeding tendencies or joint pains. At admission she was conscious, oriented and afebrile. All pulses were equally felt with bilateral renal artery and abdominal aortic bruit. Her BP in right upper limb was 250/160 mm Hg and that of left upper limb was 270/180 mm Hg. On auscultation there was an ejection systolic murmur in aortic area with loud A2. Central nervous system examination revealed normal higher functions with right UMN type of facial palsy and pyramidal weakness of right upper limb. Fundus examination showed bilateral grade III hypertensive retinopathy. A diagnosis of young stroke was made. Her investigations were as follows: Hb :10.6 gm%, urine r/e : normal, RBS: 87mg%, s.creatinine : 0.9 mg%, T.cholesterol :162 mg %, TGL :126 mg %, HDL :49 mg%, LDL: 98mg %, ESR :15mm/hr, CRP: negative, s. homocysteine 6.1µmol/L (3.3-7.2), anti nuclear antibody and anti cardiolipin were negative.

Since there was hypertension and bilateral renal arterial bruit, we proceeded with imaging of the vessels. Renal arterial Doppler could not make out the origin of the renal arteries on both sides with contracted right kidney and multiple collaterals noted in both hila. Bilateral intrarenal arteries also showed ‘parvus et tardus’ pattern. CT angiogram of abdominal aorta and renal arteries showed diffuse wall thickening of abdominal aorta with complete occlusion of superior mesenteric artery origin. Renal arteries appeared narrowed for the entire course on both sides. Flow till the popliteal artery was normal and filling of vessels distal to politeal artery was delayed because of the abdominal aortic occlusion. CT angiogram also showed normal aortic arch vessels with luminal narrowing and complete occlusion of right proximal brachial artery with normal distal brachial artery and total occlusion of left distal axillary artery. Her cardiac function as assessed by echocardiogram was normal with moderate concentric left ventricular hypertrophy and grade II diastolic dysfunction. CT brain revealed moderate infarct in left capsuloganglionic region and adjacent corona radiata. DTPA scan revealed normal perfusion and function of left kidney with reduced size, perfusion and function of right kidney.

CT ARCH VESSELS AND UPPER LIMB ANGIOGRAM:
CT angiogram showing multifocal narrowing of bilateral upper limb arteries
CT RENAL ANGIOGRAM
CT Renal angiogram showing bilateral narrowing of renal arteries and smaller right kidney

CT BRAIN- PLAIN
CT BRAIN showing moderate sized infarct in the left capsulo ganglionic region

DIAGNOSIS: With the above picture, a diagnosis of Takayasu’s arteritis presenting as stroke was made as the patient satisfied the American College of Rheumatology criteria.

COURSE AND MANAGEMENT:
Patient was treated with antihypertensives, aspirin and physiotherapy. Her weakness improved. Low dose steroids and immunosuppression with methotrexate was started for disease activity. Since her hypertension was uncontrolled with drugs and as acute phase reactants were normal she was planned for renal artery revascularization and was referred to vascular surgery department.

DISCUSSION:
Neurological involvement is uncommon in TA and neurological symptoms as the first manifestation of the disease are extremely rare. The neurological manifestations that can possibly occur are TIAs, cranial nerve palsies, dizziness, syncope, headache or visual changes. Stroke at the onset is seen in less than 10% of cases. Few cases have been reported in the literature with stroke as the initial manifestation of TA. Sikaroodi H et al in 2007 reported a case of stroke as the initial presentation of TA in a 50 year old woman. Similarly Khelani and SM Baig in 2002 reported two cases of TA with stroke as the initial manifestation. From India M V Krishna in 2004 reported a case of stroke as the initial presentation in a 22 year old female.

Recently Hwang J et al discussed the possible mechanisms of occurrence of stroke in patients with TA. Strokes developed at an younger age in patients with TA than observed for atherosclerotic stroke. Stroke in TA patients may develop due to conventional risk factors like hypertension, dyslipidemia and diabetes or arterial occlusion. Lobar infarcts in TA are due to embolism from carotid plaque rupture while borderzone infarcts and deep subcortical infarcts occur due to hemodynamic compromise from extra cranial occlusion or intracranial arterial stenosis (ICAS). Conventionally TA is described as a large vessel vasculitis involving aorta and its major branches. But in this study (Hwang J et al 2012) greater than 50% patients with stroke had intra cranial arterial stenosis (ICAS). In three patients ICAS occurred with normal aortic arch vessels.

Regarding the cause for left capsuloganglionic stroke in our patient, hemorrhage due to uncontrolled hypertension is ruled out as the CT brain did not show any bleed. Also as other conventional risk factors like diabetes and dyslipidemia were absent the cause for ischemic stroke could be due to hypoperfusion due to carotid occlusion or intracranial arterial stenosis (ICAS). As carotid angiogram was normal in our patient the
most likely explanation for left capsuloganglionic infarct could be intra cranial arterial stenosis (ICAS). ICAS is normally diagnosed by angiogram of cerebral vessels which was not done in this patient due to financial constraints.

CONCLUSION: Stroke is a rare initial manifestation of TA. If suspected, young stroke patients should be evaluated for TA.

REFERENCES:


