A case of Takayasu Arteritis presenting with young stroke

Seng Wee Cheo, MRCP[1], Haneesah Mohd Zamin, MBBS[1], Qin Jian Low, MRCP[2], Yee Ann Tan, MRCP[3], Yuen Kang Chia, MRCP[3]

[1]Department of Internal Medicine, Hospital Lahad Datu, Sabah, Malaysia, [2]Department of Internal Medicine, Hospital Sultanah Nora Ismail, Batu Pahat, Johor, Malaysia, [3]Department of Internal Medicine, Hospital Queen Elizabeth, Kota Kinabalu, Sabah, Malaysia

SUMMARY

Stroke is a debilitating disease as it carries significant morbidity especially when it affects the younger population. There are various etiologies of young stroke, namely arterial dissection, cardioembolism, thrombophilia, inherited genetic disorder and vasculitis. Young patient with stroke should undergo complete evaluation to identify the underlying etiology in order to prevent recurrence of stroke. Here, we would like to illustrate a case of Takayasu arteritis presenting as young stroke in a 17-years-old lady with no known medical illness.

INTRODUCTION

Stroke is defined as acute onset of neurological deficit attributed to focal injury of the central nervous system due to a vascular cause. It is the second leading cause of death worldwide with more than 5 million people who died from stroke annually. Broadly, stroke can be divided into ischemic and hemorrhagic types. Ischemic stroke can be further classified by TOAST (trial of ORG 10172 in acute stroke treatment) classification into large-artery atherosclerosis, cardioembolism, small vessel occlusion, stroke of other determined etiology and stroke of undetermined etiology. At present, there are more than 150 aetiologies of stroke.[1] On the other hand, stroke in young adults are uncommon where it only accounts for 10% of stroke cases. Here, we would like to illustrate a case of Takayasu arteritis (TAK) presenting with young stroke.

CASE REPORT

A 17-years-old woman with no known medical illness presented with sudden onset of right sided body weakness and loss of speech 1 hour prior to hospital visit. She denied headache, seizures, blurry of vision, chest pain, shortness of breath or fever. There was no altered consciousness. On arrival at the Lahad Datu hospital, her blood pressure (BP) was 108/75mmHg, pulse rate was 98 beats per min, temperature was 38°C. Her Glasgow Coma Scale (GCS) was E4V1M6. On examination, she had right carotid bruit, right sided power was 1/5 with expressive aphasia. Blood investigations showed hypochromic microcytic anaemia (Hemoglobin 9.7g/dl) with normal renal function and liver function. Her electrocardiogram showed sinus rhythm. Emergency Computed Tomography (CT) of her brain showed hyperacute left middle cerebral artery (MCA) infarction (Figure 1).

She was then admitted to the ward, treating the case as acute left MCA infarct with aspirin and statin. Young stroke workup showed negative anti-nuclear antibodies, C-ANCA, P-ANCA and normal C3 and C4. Her echocardiogram was also normal with no valvular lesions, shunts or clots. Subsequently, she was noted to have discrepant upper and lower limbs BP with upper limb BP read 89/65mmHg and lower limb 140/70mmHg. There was also absence of radial pulse. TAK was then suspected.

Subsequently, her computed tomography angiography (CTA) of aorta showed thickening of the thoracic aorta, right pulmonary artery and the large and medium size vessels of abdominal aorta causing varying degrees of stenosis, and occlusion of left common carotid, proximal left subclavian, common hepatic and proximal portion of superior mesentery arteries. Interval CT brain showed evolution of MCA infarction and on contrast, long segment left common carotid artery and ICA thrombosis seen (Figure 2). There were no features of vasculitis seen in the CTA of the brain. She was diagnosed to have TAK. She was treated with pulse intravenous methylprednisolone 1g daily for 3 days followed by prednisolone 1mg per kg daily. She was also started on methotrexate to control her disease activity. After intensive rehabilitation, she improved functionally over time. She was able to ambulate and tolerate orally and speak. Her Modified Rankin Scale after 6 months' post stroke was 2.

DISCUSSION

Stroke is uncommon in young adults and children. However, stroke has more significant impact on younger adults as it leaves them with significant morbidity in their most productive years.[2] Globally, there is no universal definition to delineate the age cut off for young stroke. Previous published article defined as those younger than 45-year-old. Young stroke has different etiologies compared to stroke in older adults. Arterial dissection, cardioembolism, atherosclerosis, inherited coagulation disorder, thrombophilias, vasculitis, autoimmune disease, genetic diseases are common causes of young stroke. Patient with young stroke should undergo complete evaluation such as echocardiography, thrombophilia screening, genetic testing to confirm the underlying etiology. Genetic testing that can be done include NOTCH-3 mutation for cerebral autosomal dominant arteriopathy with subcortical infarct and leucoencephalopathy (CADASIL) and mitochondrial DNA testing for mitochondrial diseases. It's important to conclude the underlying etiology as certain etiologies are treatable.

This article was accepted: 31 August 2020
Corresponding Author: Seng Wee Cheo
Email: cheosengwee@gmail.com

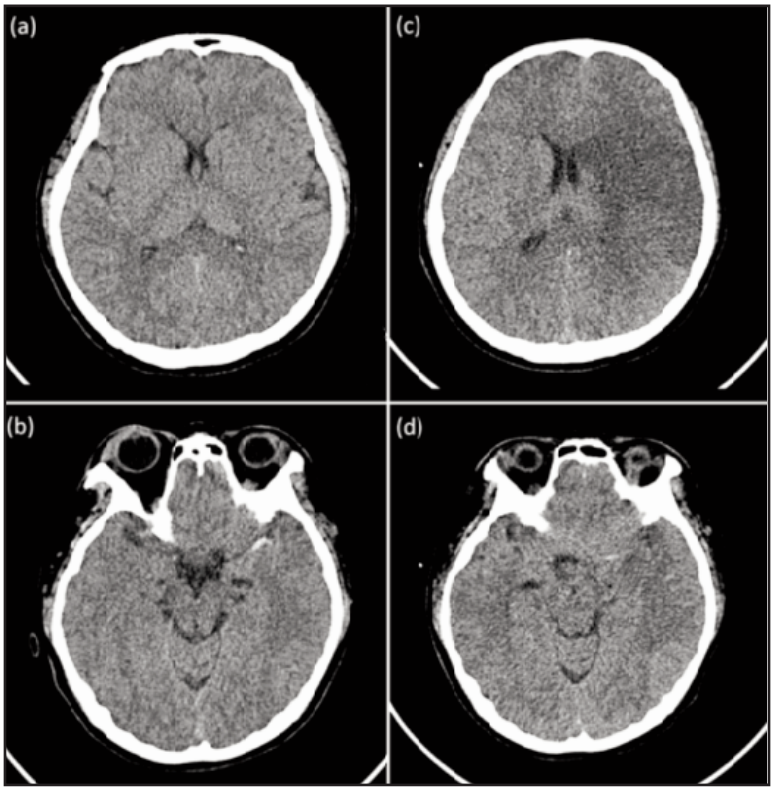

Fig. 1: (a)(b): CT Brain on admission showed hyperdense MCA sign suggestive of early MCA infarct, no focal brain edema. (c)(d): CT brain at 48 hours showed evolving changes of acute left MCA infarct with focal cerebral edema and mass effect.

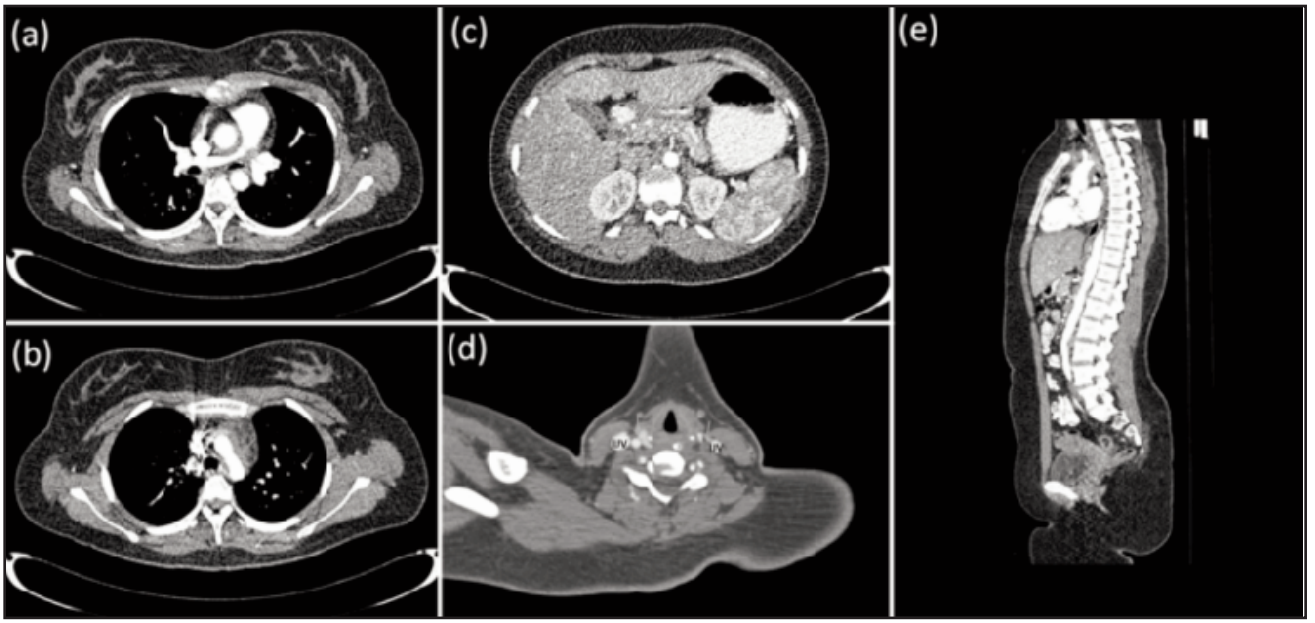

Fig. 2: (a): CTA showed moderate circumferential wall thickening of right pulmonary artery with moderate luminal narrowing. (b): moderate arterial wall thickening at the arch of aorta. (c): proximal superior mesenteric artery is not well opacified in keeping with occlusion. (d) left internal carotid artery is not well opacified. (e)sagittal view of aorta showed luminal narrowing of celiac trunk and superior mesenteric artery.

TAK is a rare large vessel vasculitis that predominantly affects the aorta and its major branches, the coronary arteries and pulmonary arteries. It is a rare vasculitis with incidence of 1-2 per million populations and predominantly affecting young female patients. Common clinical presentation includes claudication, absence of pulses, carotid bruit, hypertension, headache, difference in blood pressure and fever. Neurological symptoms are common but it is rare to have stroke as the initial presentation.[3] Other common neurological symptoms are headaches, blurring of vision and

etc. The possible mechanisms of stroke in TAK include embolism of stenotic or occlusive lesions of the aorta and it branches, hypertension, cardioembolism and cerebral hypoperfusion.[4]

In most cases, the diagnosis of TAK can be obtained by clinical evaluation paired with imaging. Imaging either CTA or magnetic resonance angiography, typically shows tapered luminal narrowing or occlusion with thickening of vessel wall. Diagnostic criteria by the American College of Rheumatology can further improve diagnostic accuracy. Our patient was a young lady who presented with acute ischemic stroke. Examination showed absent radial pulse, unequal blood pressure and bruits. CTA findings were consistent with TAK.

Management of TAK can be divided into acute or long term. Acute stroke can be treated with thrombolytic therapy and interventional revascularisation. Long term treatment aims to reduce inflammation of vessels. Steroids remains the mainstay of treatment for TAK.[5] Cytotoxic agents such as cyclophosphamide, azathioprine and methotrexate can be used as steroid sparing agents. Endovascular stenting or bypass should be considered in patient with severe structural damages.

CONCLUSION

In conclusion, young stroke is a unique entity as it affects patients at their most productive period of life. Patient with young stroke should undergo complete evaluation to exclude dissection, cardioembolism, thrombophilia, vasculitis and genetic disease. Takayasu arteritis is a rare cause of young stroke and a potentially treatable cause of stroke.

FINANCIAL DISCLOSURES

The authors of this manuscript confirm that they have no affiliations with or involvement in any organisation or entity with any financial interest in the subject matter or materials discussed in this manuscript.

CONFLICTS OF INTEREST

The authors have no conflicts of interest to declare.

INFORMED CONSENT

Written informed consent was obtained from the patient for publication of this manuscript.

REFERENCES

1. Amarenco P, Bogousslavsky J, Caplan LR, Donnan GA, Hennerici MG. Classification of stroke subtypes. Cerebrovasc Dis 2009; 27(5): 493-501.
2. Smajlović D. Strokes in young adults: epidemiology and prevention. Vasc Health Risk Manag 2015; 11: 157-64.
3. Gao S, Wang R. Takayasu arteritis presenting with massive cerebral ischemic infarction in a 35-year-old woman: a case report. J Med Case Reports 2013; 7: 179.
4. Gouda W, Alsaqabi F, Alkadi A, Amr HAE-A, Moshrif A, Mahdy ME. Ischemic stroke as the first presentation of takayasu's arteritis in young male. Clin Case Rep 2020; 8: 258-61.
5. Wen CY, Chang CJ, Hsieh CT. Stroke as an initial presentation of Takayasu's arteritis. Neurology Asia 2015; 20(2): 177-80.