CASE REPORT

Sternal Cleft With Coarctation of Aorta and Vascular Anomalies - Learning Point in Young Stroke

Koa Ai Jiun[1], Teh Yong Sim[2], Lim Ruwei[1]

[1] Radiology Department, Faculty of Medicine and Health Sciences, Universiti Malaysia Sarawak (UNIMAS) 94300 Kota Semarahan Sarawak
[2] Pusat Jantung Sarawak, Kuching –Semarahan Expressway 94300 Kota Semarahan Sarawak

ABSTRACT

Sternal cleft is a rare congenital anomaly that is known to be associated with other congenital abnormalities in particular coarctation of aorta and vascular anomalies. We present a case of a young teacher who presented with recurrent episodes of ischemic stroke. During the workup to look for the cause of stroke, it was discovered that this patient had complete midline sternal cleft with underlying coarctation of aorta and left subclavian artery aneurysm. We wish to highlight the relation between the presence of sternal cleft and coarctation of aorta as well as vascular anomalies as one of the rare cause of young stroke.
Malaysian Journal of Medicine and Health Sciences (2022) 18(6):359-362. doi:10.47836/mjmhs18.6.49

Keywords: Sternal cleft, Coarctation of aorta, Subclavian artery aneurysm, Young stroke

Corresponding Author:
Koa Ai Jiun, MRAD
Email: koaaijiun@yahoo.com
Tel: +60138259062

INTRODUCTION

Sternal cleft is a rare congenital anomaly that results from failure of sternal fusion early during embryological development (1). It is usually diagnosed based on clinical examinations and chest radiograph. However, Computed Tomography (CT) Thorax remains as the gold standard to ascertain and characterize the extent of the sternal anomaly as well as for the planning of surgical correction. Several literatures have reported the association between sternal cleft with other congenital abnormalities in particular vascular anomalies and coarctation of aorta. However, to date there is no literatures reporting the relationship between the presence of underlying sternal cleft with the higher risk of developing ischemic stroke.

CASE REPORT

We presented a case of 46 years old gentleman, who are from an indigenous group of Sarawak, and worked as a teacher in government school. He was referred to our center for recurrent episodes of ischemic stroke within a period of one-year. The first episode of stroke showed right total anterior circulation infarct, and was started on Tablet Aspirin 75 mg once daily and Tablet Atorvastatin 40 mg once daily. However approximately one year after, he suffered a second episode of stroke with posterior circulation infarction involving bilateral cerebellum and left pontine. On clinical examination his blood pressure ranged from 100-110/80-90mmHg with heart rate ranging from 70-80 beats per minute. He had mild left sided weakness with the power of 4 over 5 involving both the left upper and lower limbs. Normal tone and reflexes in the left upper limb with mild increased in the tone and reflexes in the left lower limb. No contractures of the joint documented. He was ambulating with walking stick when he came for his clinic follow up. Cranial nerves examinations are normal. He was a social smoker, however he had stopped smoking 3 months after the first episode of stroke. There was no family history of cardiovascular or cerebrovascular events.

CT Angiogram (CTA) of Brain, Neck and Thorax as part of workup for young stroke showed a narrowed segment of aortic arch after the origin of left subclavian artery, approximately 4cm in length suggestive of coarctation of aorta (Figure 1). The diameter of this narrowed segment measured 1cm. There are mural thrombi at the proximal part of this narrowed segment, immediately below the ostium of left subclavian artery. This narrowed segment of aorta is tortuous and curved superiorly, with evidence of post narrowing dilatation of the aorta (Figure 1). There is also fusiform aneurysm of left subclavian artery after its origin from the aortic arch (Figure 2). The aneurysm measures 3.6cm in diameter and 5.6cm in length. The aneurysm covers the ostium of left vertebral artery and left internal mammary artery. Distal left subclavian artery is normal in caliber. CTA of the neck also showed long segment non-opacification of proximal right vertebral

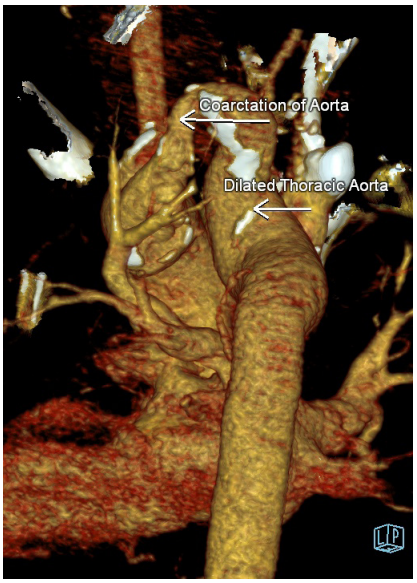

Figure 1: Volume Rendering Technique (VRT Image): Narrowed segment of aortic arch after the origin of left subclavian artery consistent with coarctation of aorta with post narrowing dilatation of the descending thoracic aorta

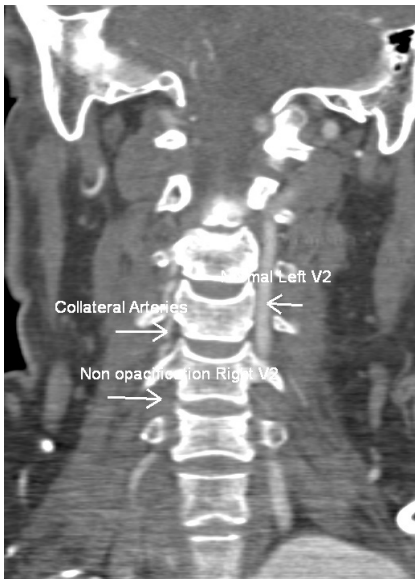

Figure 3: Coronal images of CT Angiogram neck. Long segment non-opacification of proximal right vertebral artery involving V2 segment with presence of multiple collateral arteries suggestive of chronic thrombosis. Normal opacification of left vertebral artery.

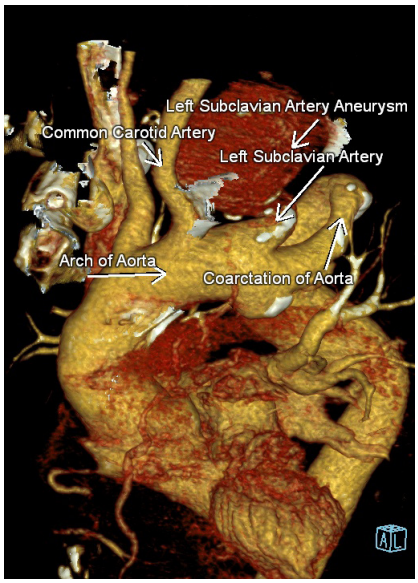

Figure 2: VRT Image. Fusiform aneurysm of left subclavian artery after its origin from aortic arch

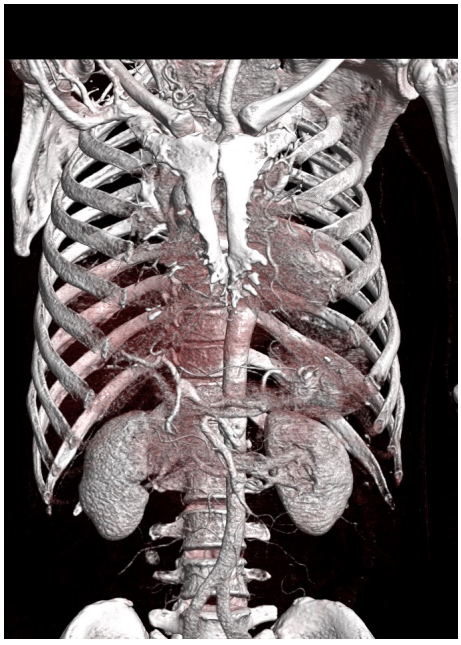

Figure 4: VRT Image. Complete midline sternal cleft

artery involving V1 to V3 segments with presence of multiple collateral arteries (Figure 3) suggestive of chronic thrombosis. Distal vertebral artery is small in caliber, however remained patent. Normal opacification of left vertebral artery (Figure 3). No abnormalities of the intracranial cerebral arteries and abdominal aorta.

Another interesting finding include presence of complete midline sternal cleft (Figure 4). Overall, the findings are consistent with coarctation of the aortic arch and left subclavian artery aneurysm with chronic thrombosis of right vertebral artery and underlying sternal cleft.

Echocardiograms showed coarctation of aorta with pressure gradient of 51mmHg. The left ventricle is normal in size with no thrombus and ejection fraction of > 55%. Right ventricle is normal in size and function. Both left and right atriums are normal with intact interatrial septum. The mitral, tricuspid, aortic and pulmonic valves are normal in structure and function.
CTA coronary was also done and the findings showed aneurysmal left anterior descending coronary artery with 30-40% stenosis. The left main stem coronary artery, left circumflex coronary artery and right coronary artery are normal and patent.

This patient was subsequently referred to another center for Thoracic endovascular aortic repair (TEVAR) for the left subclavian artery aneurysm. Unfortunately he

passed away from the complication of the procedure.

DISCUSSION

Generally stroke in population under 45 years old are less common compared to the older population and is usually categorized as young stroke. According to the epidemiology and etiology study of young stroke by Davna Griffiths et al, the causes of stroke in younger population differ from those of older population (2). One of the key findings in this study showed that although cardio embolic events remain as one of the most common causes of stroke in both the younger and older population; the cause of these cardio embolic events differ between the age groups (2). Screening for congenital defects or abnormalities, nonatherosclerotic vasculopathies, extracranial arteries dissection and autoimmune conditions should always be part of the workup for young patients who presented with stroke.

Our patient first presented with ischemic stroke at the age of 45 years old. Unfortunately, further investigations were only carried out when he presented with second episode of stroke a year later. A series of investigations which include CTA of Coronary vessels, Thorax, Neck and Cerebral showed evidence of coarctation of aorta, left subclavian artery aneurysm, long segment chronic thrombosis of right vertebral artery (V1 to V3 segments) and left anterior descending coronary artery aneurysm. Of note, one incidental findings during the imaging study revealed that this patient had a complete midline sternal cleft. Sternal cleft is a rare congenital anomaly that results from failure of ventral fusion of the sternal bars (1). Its etiology is uncertain, however literatures have reported cases of other congenital abnormalities that may co-exist with sternal cleft in particular vascular anomalies and coarctation of aorta. As seen in our patient who had a complete midline sternal cleft, he also had coarctation of aorta, left subclavian artery aneurysm left anterior descending coronary artery aneurysm.

One study observed that ischemic stroke occurred at a significantly younger age group in people with underlying coarctation of aorta (3). The earlier onset of ischemic stroke is presumed to be due to hypertension as the primary risk factor (3). However, in our patient his blood pressure was consistently within the normal range throughout his admission as well as during the clinic visits. Another hypothesis of stroke in younger patients with coarctation of aorta are due to arteropathies that are contributed by atherosclerosis or underlying vascular abnormalities; which is plausible in our case. In our patient, there is evidence of thrombus within the proximal part of the narrowed segment of the arch of aorta as well as evidence of chronic thrombosis of the right vertebral artery. This is likely to cause an embolic event to the cerebral arteries resulting in stroke.

Subclavian artery aneurysm per se is uncommon. Reported cases of subclavian artery aneurysm are often associated with trauma, thoracic outlet syndrome, infection and some genetic syndromes like Marfan syndrome and Turner's syndrome. Since there are no reported history of trauma or infection; as well as absence of syndromic facies; we are postulating that the left subclavian artery aneurysm and coarctation of aorta seen in this patient are congenital and are part of the vascular anomalies that are associated with sternal cleft. Several literatures reported about the association of sternal cleft in children with PHACES (posterior fossa abnormalities, hemangiomas, arterial lesions, cardiac abnormalities/coarctation of aorta, eyes abnormalities and sternal defects) syndrome.

PHACES syndrome is defined by presence of hemangioma larger than 5cm, and associated with 1 major or 2 minor criterias (4). Unfortunately, there are no reports or clinical notes on the presence of hemangioma in our patient thus he did not fulfill the criteria and guidelines for the diagnosis of definite PHACES syndrome. However he had three major criteria which are arterial anomalies (left subclavian artery aneurysm and left anterior descending coronary artery aneurysm), coarctation of aorta and midline sternal cleft. Accordingly, this lead to a possible PHACES diagnosis.

As there are involvement of the large arteries (aorta and left subclavian artery), Takayasu arteritis (TA) is another differential diagnosis that is worth considering. TA is an inflammatory disease that primarily involves large vessels such as aorta and its major branches. However, this patient was completely well and did not present with any episodes of malaise, fever or fatigue prior to the event leading to his hospitalization. Moreover, the typical findings for TA on CT which are concentric mural thickening with transmural calcification of the arteries causing luminal stenosis (5) are not depicted in this case. On top of that, TA is more prevalent among women under 40 years old and this patient does not fulfill the Sharma criteria for the diagnosis of TA (5).

CONCLUSION

Since the incidence of the sternal cleft itself is rare, there are hardly reported cases of association between sternal cleft with vascular anomalies and coarctation of aorta. In this case report we illustrate the relation between sternal cleft and coarctation of aorta as well as vascular anomalies as one of the rare cause of young stroke. This is to create awareness among the clinicians that the presence of sternal cleft is a strong indication to look for other asymptomatic vascular anomalies.

ACKNOWLEDGEMENT

The authors would like to thank the Universiti Malaysia Sarawak (UNIMAS),. Cardiothoracic Unit, Sarawak Heart Center and Cardiology Unit, Sarawak Heart

Center for their support.

REFERENCES

1. Paras Kothari, Prashant S. Patil, Geeta Kekre, Ravi Kamble, Kiran Vishesh Dikshit. Complete Sternal Cleft – A Rare Congenital Malformation and its Repair in a 3-month-old boy: A Case Report. Journal of Indian Association Pediatric Surgeons. 2016 Apr Jun; 21(2):78-80. doi: 10.4103/0971-9261.126961.
2. Davna Griffiths, Jonathan Stum. Epidemiology and Etiology of Young Stroke. Stroke Res Treat. 2011; 2011:209370. doi: 10.4061/2011/209370.
3. Ryan J Felling, Richard E Ringel. Mind the Brain: Stroke Risk in Young Adults with Coarctation of the Aorta. Journal of American Heart Association. Volume 7(11); 2018 Jun 5. doi:10.1161/JAHA.118.009461.
4. Anita Rotter, Luciana Paula Samorano, Maria Cecilia Rivitti-Machado, Zilda Najjar Prado Oliveira, Bernardo Gontijo. PHACE Syndrome: clinical manifestations, diagnostic criteria, and management. Brazillian Society of Dermatology. 2018 May-Jun; 93(3): 405-411. doi: 10.1590/abd1806-4841.20187693.
5. F P Zhu, S Luo, Z J Wang, Z Y Jin, L J Zhang and G M Lu. Takayasu arteritis: Imaging Spectrum at multidetector CT Angiography. Br J Radiol. 2012 Dec; 85(1020): e1282-e1292. doi: 10.1258/bjr/25536451.