J Med Sci 2019;39(2):98-101
DOI: 10.4103/jmedsci.jmedsci_133_18

CASE REPORT

Primary Thoracic Endovascular Aortic Repair to Aortic Coarctation Complicating with Spontaneous Epidural Hemorrhage

Ching-Wei Lin[1], Yi-Ting Tsai[1]

[1]*Division of Cardiovascular Surgery, National Defense Medical Center, Tri-Service General Hospital, Taipei, Taiwan*

Development of collateral circulation belongs among the typical signs of aortic coarctation (CoA) and can lead to the growth of ectatic, fragile spinal neurovascular malformations. Spontaneous spinal epidural hemorrhage (SSEH) as a complication of CoA is exceptionally rare, with only few case reports proposed up to date. At our institute, we had two experiences of SSEH complicated by CoA. The first case had been published in 2016, with disappointing neurologic outcome. As for the second case, presenting with extremely sharp upper back pain followed by paraplegia and anal sphincter impairment, we chose immediate surgical intervention with thoracic endovascular aortic repair (TEVAR), aiming at amelioration of the pressure gradient across the juxtaductal coarctation in the aorta, to ensure the safety and the completeness of following surgical decompression of the spinal epidural hematoma. To our knowledge, this case is the first one featuring TEVAR to the CoA immediately followed by surgical decompression of SSEH in this kind of emergent setting. This case recovered satisfactorily without neurologic deficit. Conservative treatment for SSEH caused by CoA may not succeed at all times. Emergent TEVAR to CoA immediately followed by surgical decompression is achievable, making following surgical decompression of the epidural hematoma easier and safer.

Key words: Aortic coarctation, aortic stenting, paraplegia, spontaneous spinal epidural hemorrhage, spontaneous spinal epidural hemorrhage, thoracic endovascular aortic repair, thoracic endovascular aortic repair

INTRODUCTION

Aortic coarctation (CoA) accounts for about 6.8% of congenital heart disease[1] and is usually diagnosed and treated in childhood. Untreated adults may present with various symptoms of hypertensive cardiac disease and vascular complications such as intracranial aneurysm or hemorrhage. Complication with spontaneous spinal epidural hemorrhage (SSEH) is exceptionally rare, with only few case reports[2-7] proposed up to date. Fortunately, we had two experiences of SSEH complicated by CoA at our institute in recent 3 years. The first case[6] had been published in 2016, with disappointing neurologic outcome. This time, we report the case of a 44-year-old male, presenting with extremely sharp upper back pain followed by paraplegia and anal sphincter impairment developed in emergency department (ED), which was treated by immediate intervention with emergent thoracic endovascular aortic repair (TEVAR) with stenting, aiming at amelioration of the pressure gradient across the juxtaductal coarctation in the aorta, thus minimizing the vascular pressure of the patient's spinal artery, to ensure the safety and the completeness of following surgical decompression of the spinal epidural hematoma. To our knowledge, this is the first case featuring TEVAR to the CoA immediately followed by surgical decompression of SSEH in this kind of emergent setting. This case recovered satisfactorily without neurologic deficit.

CASE REPORT

A 44-year-old man, with a history of hypertension since his early adulthood, was sent to our ED due to sudden-onset extremely sharp upper back pain while playing Mahjong with

Received: September 03, 2018; Revised: September 25, 2018; Accepted: October 03, 2018
Corresponding Author: Dr. Yi-Ting Tsai, Division of Cardiovascular Surgery, National Defense Medical Center, Tri-Service General Hospital, No. 325, Sec 2, Chenggong Rd, NeiHu district 114, Taipei, Taiwan. Tel: +886-2-87923311; Fax: +886-2-87927376. E-mail: cvsallen@mail.ndmctsgh.edu.tw

How to cite this article: Lin CW, Tsai YT. Primary thoracic endovascular aortic repair to aortic coarctation complicating with spontaneous epidural hemorrhage. J Med Sci 2019;39:98-101.

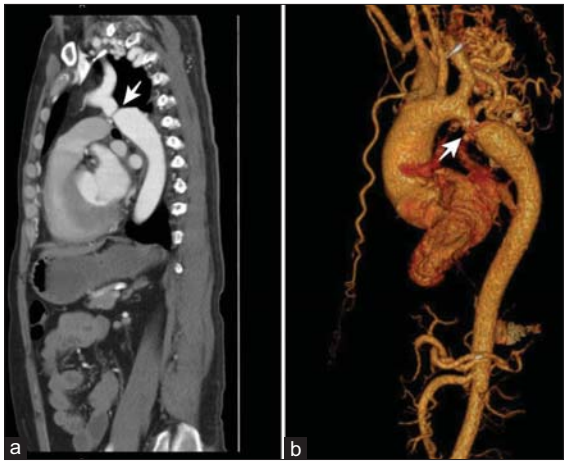

Figure 1: (a) Sagittal computed tomography angiography at the emergency department showing juxtaductal coarctation of aorta. (b) Computed tomography angiography (three-dimensional reconstruction image) demonstrating coarctation of aorta and abundant collateral vessels

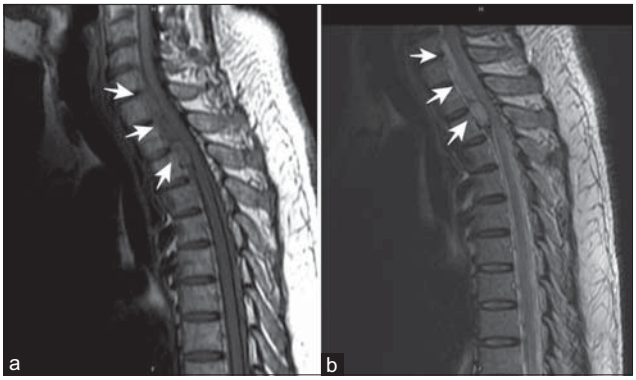

Figure 2: (a) Sagittal magnetic resonance T1-fluid-attenuated inversion recovery image showing isointense (to spinal cord) epidural hematoma (arrows) at the C6–T2 level of spine. (b) Sagittal magnetic resonance T2-fast spin-echo image demonstrating heterogeneous signal cord-compressing hematoma (arrows)

his friends. There was no history of injury to his back or chest wall. On arrival, the patient was alert and oriented. Blood pressure measurement was 260/184 and 258/178 mmHg taken from his both upper arms, while relatively low blood pressure of 126/88 and 134/90 mmHg was taken from his both lower limbs. Electrocardiography showed sinus tachycardia with left ventricular hypertrophy. Chest computed tomography angiography was immediately arranged and the presumptive diagnosis of aortic dissection was excluded. However, a juxtaductal CoA with extensive network of collateral vessels was incidentally found [Figure 1]. In the following 3 hours, the patient developed symptoms of paraplegia and paresthesia below the T4 dermatome in the ED and lost his anal sphincter function. Then, an emergent spinal magnetic resonance image was arranged, showing a long, thin, fusiform isointense epidural hemorrhage causing compression to the spinal cord in the spinal canal at the level of C6–T2 in T1-phase image, whereas the epidural hematoma was heterogeneously intense in T2-phase image [Figure 2].

With regard to the long-term neurologic outcome, emergent surgical evacuation of the epidural hematoma is considered, but high pressure in the spinal artery secondary to the CoA might interfere with the surgical decompression procedure. We chose immediate intervention with TEVAR [Figure 3], aiming at amelioration of the pressure gradient across the juxtaductal coarctation in the aorta, to ensure the safety and the completeness of following surgical decompression of the spinal epidural hematoma. Then, the cervicothoracic spinal epidural hematoma was evacuated with the procedure of laminectomy (C6–T2) plus internal fixation with transpedicular screws immediately after the TEVAR. The patient regained his muscle power of the lower limbs soon postoperatively after extubation in the intensive care unit, as well as the anal sphincter function. His blood pressure of upper limbs significantly lowered after the surgery. The patient was transferred to the general ward and was discharged 2 weeks later.

DISCUSSION

CoA, derived from the Latin word coarctatio meaning stricture, is a tight narrowing of the aortic lumen that can occur at any point in its course but is most often found at the level of the isthmus.[1] CoA may occur as an isolated defect or in association with various other lesions, most commonly bicuspid aortic valve and ventricular septal defect. Development of collateral circulation belongs among the typical signs of CoA and can lead to the growth of spinal neurovascular malformations due to the altered hemodynamic parameters and to a rich network of ectatic, fragile collaterals. The combination of these two disease entities creates unique and complex diagnostic and therapeutic challenges. Several case reports[2-7] had been proposed with similar clinical presentation. In the case reported by Dauphin *et al*.,[3] surgical evacuation of the spinal epidural hematoma was performed before neurological complication developed and the coarctation was surgically corrected thereafter. In the case reported by Harrer *et al*.,[4] spontaneous regression of neurological symptoms was achieved after aggressive strict control of blood pressure with medication, and the CoA was surgically repaired 1 week later. In the case[2] reported by Aoun *et al*., a spinal artery aneurysm at the C7 level of spine as well as extensive network of collateral vessels was discovered.

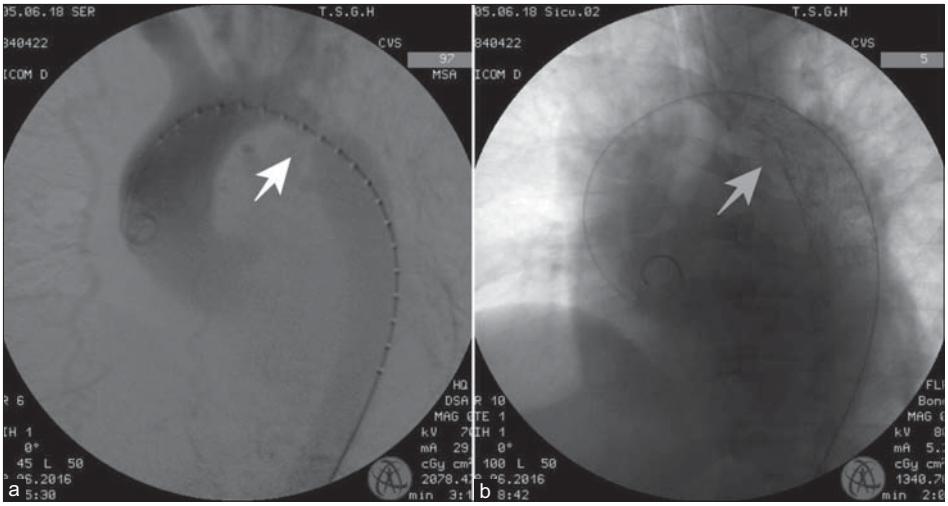

Figure 3: (a) Aortography showing coarctation of aorta (arrow) before endovascular stenting. (b) Perioperative fluoroscopy showing image after stenting (gray arrow) to the coarctation of aorta

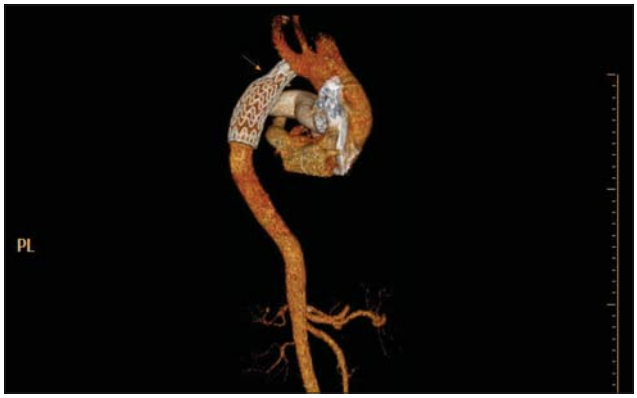

Figure 4: Computed tomography angiography (three-dimensional reconstruction image) of postoperative follow-up after thoracic endovascular aortic repair showing improved diameter of aorta at the juxtaductal coarctation (arrow) and significant regression of the collateral vessels

Aoun *et al.* performed embolization to the spinal artery aneurysm, followed by surgical correction of CoA 3 months later. Aoun *et al.* considered that treating the CoA could potentially induce severe hemodynamic disturbances and sudden increase of the pressure of aorta proximal to the stenotic lesion, as well as the pressure in the spinal artery aneurysm, leading to re-bleeding episode which may worsen the neurologic outcome. In the case[7] reported by Park *et al*., the patient showed gradual improvement in the neurological deficit of the upper extremities following anti-edematous therapy including intravenous corticosteroid injections, but not fully recovered, with some extent of neurologic sequelae. The first case of our institute[6] was treated with TEVAR several days after the date of onset of the paraplegia, with disappointing neurologic outcome.

In this case, we aimed at reducing the pressure in the spinal artery prior to surgical evacuation of the cord-compressing hematoma at the C6–T2 level of spine. We chose endovascular stenting technique to dilate the juxtaductal CoA of thoracic aorta, avoiding the relatively long time of cross-clamping at thoracic aorta, which is essential for the traditional surgical technique to treat the coarctation. To our knowledge, this is the first case featuring primary TEVAR to the CoA immediately followed by surgical decompression of spinal epidural hemorrhage in the emergent setting. Moreover, the subsequent neurologic outcome was excellent.

CONCLUSION

CoA with spinal complications is exceptionally rare, but can be diagnosed through modern imaging modalities. Rapid diagnosis of spinal epidural hematoma and adequate surgical decompression may lead to improved neurologic outcome. The concepts we want to share with readers are listed as follows: (1) Conservative treatment for SSEH caused by CoA may not succeed at all times; (2) TEVAR to the CoA can avoid the relatively long time of cross-clamping at thoracic aorta, which is essential for the traditional surgical technique to correct the coarctation as the cross-clamping may increase the arterial inflow of the collateral vessels; and (3) Emergent TEVAR to CoA immediately followed by surgical decompression of epidural hematoma is achievable and may provide lower pressure in the tortuous collateral vessels of spinal artery secondary to the CoA [Figure 4], therefore making following surgical decompression of the epidural hematoma easier and safer.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Abbott ME. Coarctation of the aorta of the adult type: Ii. A statistical study and historical retrospect of 200 recorded cases with autopsy, of stenosis or obliteration of the descending arch in subjects above the age of two years. Am Heart J 1928;3:574-618.
2. Aoun SG, El Ahmadieh TY, Soltanolkotabi M, Ansari SA, Marden FA, Batjer HH, *et al.* Ruptured spinal artery aneurysm associated with coarctation of the aorta. World Neurosurg 2014;81:441.e17-22.
3. Dauphin C, Lusson JR, Legault B, Perez N, Motreff P, Langlade S, *et al.* Medullary extradural hematoma revealing a coarctation of the aorta. Arch Mal Coeur Vaiss 2001;94:513-7.
4. Harrer J, Dominik J, Varvarovský I, Zizka J, Harrerová L. Paraplegia as an unusual manifestation of aortic coarctation. Thorac Cardiovasc Surg 2001;49:186-7.
5. Zizka J, Eliás P, Michl A, Harrer J, Cesák T, Herman A, *et al.* Extensive spinal epidural hematoma: A rare complication of aortic coarctation. Eur Radiol 2001;11:1254-8.
6. Huang YF, Tsai YT, Tsai CS, Ke HY, Hsu PS, Lin YC. Endovascular repair for primary adult coarctation of the aorta complicated with acute epidural hematoma leading to paraplegia: A case report. J Med Sci 2017;37:19-22.
7. Park CB, Jo DJ, Kim MK, Kim SH. Paraplegia due to acute aortic coarctation and occlusion. J Korean Neurosurg Soc 2014;55:156-9.