May 2016

Interrupted aortic arch complicated with takotsubo cardiomyopathy mimicking aortic dissection

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Journal:	*BMJ Case Reports*
Manuscript ID	bcr-2017-219612.R2
Manuscript Type:	Reminder of important clinical lesson
Date Submitted by the Author:	29-Jul-2017
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Keywords:	Stroke < Neurology, Heart failure < Cardiovascular medicine, Hypertension < Cardiovascular medicine

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TITLE OF CASE

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SUMMARY

A 50 year old man presented to emergency with interscapular pain, diaphoresis and restlessness. Initial examination raised the possibility of aortic dissection; however the CT scan did not concur with the diagnosis. An ECG showed ST segment elevation in leads V1-V6 and echocardiography showed severe left ventricular systolic dysfunction. Coronary angiography through the right femoral artery was attempted but the diagnostic catheter could not be advanced to the ascending aorta. Radio contrast injection showed complete obstruction of the descending aorta. Coronary angiography through right radial approach showed mild LAD disease. The aortogram showed complete interruption of the ascending aorta with extensive collaterals network. LV gram was consistent with stress induced cardiomyopathy. We noticed intermittent confusion and agitation. MRI of the brain showed areas of deep white matter ischemia as well as micro haemorrhages, suggesting posterior reversible leucoencephalopathy syndrome. He unfortunately went in to cardiac arrest and could not be revived.

BACKGROUND

This case is an example of an unusual presentation of rare congenital anomalies in an uncommon age group. Approximate incidence of interrupted aortic arch (IAA) is reported to be 3/1000, 000 live births,[1]. It is usually associated with other congenital heart defects such as ventricular septal defect (VSD), patent ductus arteriosus (PDA), and truncus arteriosus (TA),[1]. Very rarely is it an isolated anomaly as in this case. It is the first cardiac anomaly which demonstrates a genetic pattern which is homozygous deletion 1.5-3 Mb region of chromosome band 22q11,[2-3].

In spite of being labelled as hypertensive in young age, unfortunately our patient was not investigated for the cause of hypertension in his age group. Anticipating rare causes of common disease may have an impact on the outcome of the patient. Interrupted Aortic Arch is a very rare cause of hypertension in adult population. It may be diagnosed early in young adults by anticipating on physical examination. Young patients presenting with uncontrolled hypertension should have a complete examination of the upper and lower limb pulses and blood pressure in all the four limbs. The diagnosis can be subsequently confirmed by relevant imaging.

CASE PRESENTATION

A 50 Year old man was brought in to the emergency department with history of tearing interscapular pain, diaphoresis and being restless for 20 hours. He was initially taken to a

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local hospital where his blood pressure was found to be significantly elevated. He was treated as a case of hypertensive urgency. A plain computed tomography (CT) scan brain was done to look for evidence of an acute cerebral insult, suspected due to his significantly elevated blood pressure and being restless but showed no abnormality. His past medical history was significant for hypertension since the age of 16 years. However he was not compliant with his antihypertensive medications.

On examination he was restless and diaphoretic but oriented to time place and person. He attributed his restlessness to chest pain. The Pulse was 100 beats per minute, regular and low volume in the right arm. Blood pressure in his left arm was 160/96 mmHg and 120/80 mmHg in the right arm. His lower extremities were cold and peripheral pulses were not palpable. Clinical examination of his chest and heart was unremarkable. He was electively intubated for further management.

INVESTIGATIONS

His first ECG showed ST segment elevation in leads V1-V6 but with no reciprocal changes [figure -1]. With a presumed initial clinical diagnosis of aortic dissection due to the inter-arm blood pressure difference and sharp interscapular pain, an urgent bedside transthoracic echocardiography was performed which revealed severe left ventricular systolic dysfunction (visual estimation, Basal segments were showing normal contraction while all the mid and apical segments were akinetic) [video-1A&1B]; however, a dissection flap could not be appreciated. An urgent contrast computed tomography (CT) of the chest was performed and did not show features of dissection. Lab investigation showed a WBC count of 25 x 10E9/L and Troponin-I value of 2.5ng/ml. A repeat 12 lead ECG showed new T wave inversions in lead V1-V6 [Figure-2]. On the basis of dynamic ECG changes, positive cardiac biomarkers and new left ventricular systolic dysfunction we decided to proceed to coronary angiography to rule out coronary artery disease. Femoral access was used. The J tipped guide wire could not be passed beyond the aortic arch. A radio contrast injection showed complete occlusion of proximal segment of descending aorta [figure-3]. The right femoral approach was switched to right radial approach. His aortogram with pigtail 6F catheter revealed complete occlusion of the aorta after origin of left subclavian artery associated with large collaterals arising from the arch of the aorta and subclavian artery and connecting to the descending aorta [Figure-4 &5]. His coronary angiogram revealed mild disease in mid LAD. The LV gram findings were consistent with stress induced cardiomyopathy. The repeat transthoracic echo after 72 hours showed normal left ventricular systolic function which further confirmed his diagnosis of Takotsubo cardiomyopathy [video-2]. During his wean from sedation, while he was kept off sedation, we noticed intermittent confusion and agitation in his behaviour which was not explicable with his metabolic and medication profile. Neurology advised an MRI brain which showed focal area of diffusion restriction at the corpus callosum and deep white matter along the occipital horn of right lateral ventricle [Figure-6 a-d]. The Sequential

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weighted images (SWI) showed evidence of signal drops out in the fourth ventricle and bilateral parietal sulci likely representing hemmorrhage [Figure-7].

DIFFERENTIAL DIAGNOSIS

Aortic dissection was the first differential diagnosis due to the clinical presentation. However as the initial CT aortogram did not show evidence of dissection flap, the next differential to rule out was acute myocardial infarction due to ECG changes and LV dysfunction. The coronary angiogram did not show obstructive coronary lesion. The complete occlusion of the aorta distal to the origin of subclavian artery confirms the diagnosis of interrupted aortic arch.

TREATMENT

His coronary artery disease was not compatible with his severe left ventricular systolic dysfunction. He was treated medically for his acute left ventricular failure secondary to stress induced cardiomyopathy and was successfully extubated.

Cardiothoracic team was involved for possible surgical intervention for interrupted aortic arch and surgical options were discussed but no immediate procedure was planned due to his fluctuating neurologic status.

OUTCOME AND FOLLOW-UP

He was extubated after 48 hours and was managed conservatively for his brain injury. On day 8 of his hospital stay, he developed apnoea followed by bradycardic cardiac arrest. The possible cause at that time was extension of his neurological injury up to the extent of acute cerebral compromise but he was not stable enough to be moved for another brain imaging. He was attempted for resuscitation but could not revive.

DISCUSSION

Interrupted aortic arch (IAA) is defined as the complete loss of luminal continuity between the ascending and the descending aorta. It is usually associated with other congenital cardiac defects like ventricular Septal defects, Bicuspid aortic valve and other aortopulmonary trunk anomalies,[1].

IAA is classified into three types based on the site of interruption. Type A which is interruption distal to the origin of left subclavian artery (LSA), Type B which is interruption between left common carotid and LSA and Type C which is interruption between brachiocephalic trunk and left common carotid artery,[4]

Majority of the cases are fatal in very early childhood due to the lack of extensive collaterals necessary to maintain the distal flow and closure of patent ductus arteriosus (PDA).

Diagnosis of IAA can be made by non-invasive and invasive techniques. It can be anticipated by doing a careful examination of young adults with hypertension. Non-invasive techniques

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include echocardiography, computed tomography and cardiac magnetic resonant imaging. Invasive diagnosis is made in cardiac catheterization laboratory by aortogram.

Clinical presentation in IAA varies from failure to thrive in neonates to hypertension (most common), chronic fatigue, nonspecific chest pain (due to collaterals) and heart failure (rarely) in adults. Presentation mimicking aortic dissection or acute MI has not been reported. On review of the literature we found few case reports on IAA in adult patients. Isolated IAA without other congenital cardiac anomalies in adults is even rarer,[5].

Takotsubo cardiomyopathy (CMP) was initially recognized in japan in 1990 and is an emerging diagnosis since 1998. Majority of the patients are women. Events are more prevalent in afternoon when stressful triggers are more likely to take place due to adrenaline surge which results in consequences such as acute coronary syndrome and left ventricular dysfunction. Common clinical presentation is with chest pain, dyspnoea and palpitation,[6]. To our knowledge this CMP has not been reported with IAA.

Initial presentation of our patient raised a high suspicion of aortic dissection that turned out to be an Interrupted aortic arch with associated stress induced cardiomyopathy. Literature shows that Between 5 to 10 % of dissections do not have an obvious intimal tear on the imaging and these are due to the rupture of aortic vasa vasorum with clinical sign and symptoms similar to acute intimal dissection,[7].

The posterior reversible encephalopathy syndrome (PRES) is a clinic-neuroradiological diagnosis with various etiologies, severe hypertension being the one cause. It usually results in vasogenic edema of posterior cerebral regions however the literature shows up to 15% incidence of intracranial hemmorrhage (from minute hemmorrhage to hematoma) in PRES as in our patient. The outcome varies, being reversible in majority of patients but can results in morbidity and mortality,[8].

We think that his clinical presentation was a combination of more than one disease; he may have had a tear in one of his collaterals due to the vasa vasorum rupture and therefore did not have a classical flap on CT scan. As he had high blood pressure since the age of 16 years, if he could have a comprehensive workup for secondary hypertension at young age, this diagnosis of IAA could have potentially been made and treated a long time before he presented in such an emergency situation.

This case emphasizes the role of detailed history and physical examination in diagnosing diseases with high mortality which are rare, such as IAA in adults which join the two factors.

LEARNING POINTS/TAKE HOME MESSAGES

1. Stress induced cardiomyopathy is a mimicker of myriad cardiac conditions

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2. Detailed work up should be done for secondary hypertension before starting antihypertensive medications, especially in young patients

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FIGURE/VIDEO CAPTIONS

Video 1A: PSAX view at the level of apex showing akinetic apical segments

Video 1B: PSAX view at the basal level showing contraction of all the segments

Video 2: PSAX view at the level of apex showing normal contraction of all segment

Figure 1: 12 lead ECG showing ST segment elevation in leads V1-V6

Figure 2: 12 lead ECG showing new T- wave inversion in leads V1-V6

Figure 3: Radiocontrast injection across the descending aorta showing complete occlusion of the proximal part

Figure 4: Radiocontrast injection across the ascending aorta showing complete occlusion of distal part with luminal discontinuity

Figure 5: Image of the CT aortogram showing extensive collateral circulation

Figure 6: ADC and DWI images of MRI brain showing focal area of diffusion restriction along the occipital horn of lateral ventricle (a-b) and at the splenium of corpus callosum(c-d).

Figure 7: SWI showing signal drop out in fourth ventricle and bilateral parietal sulci likely representing hemmorrhage

PATIENT'S PERSPECTIVE

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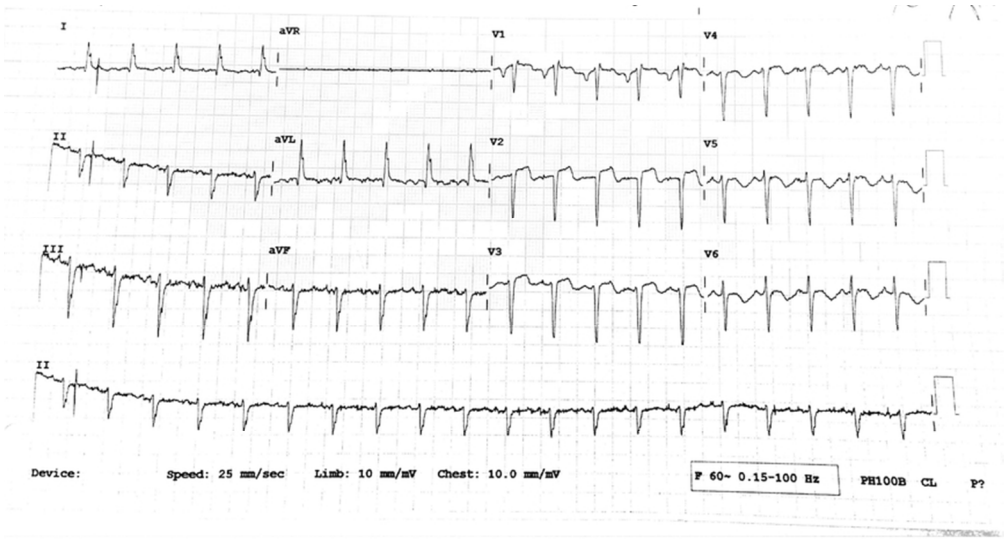
Date: 29 -07-2017

Farhala Baloch_ 29 July _ 2017



12 lead ECG showing ST segment elevation in leads V1-V6

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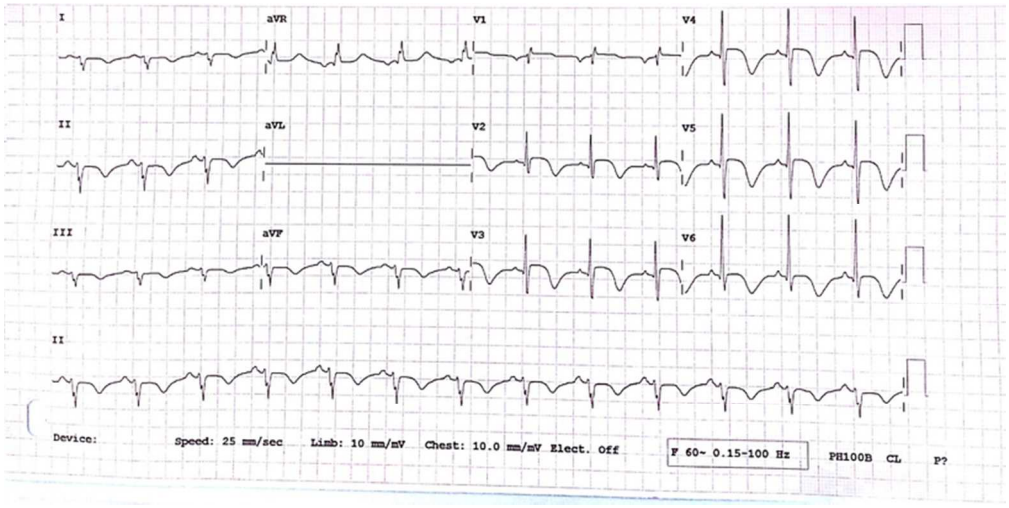

12 lead ECG showing new T- wave inversion in leads V1-V6

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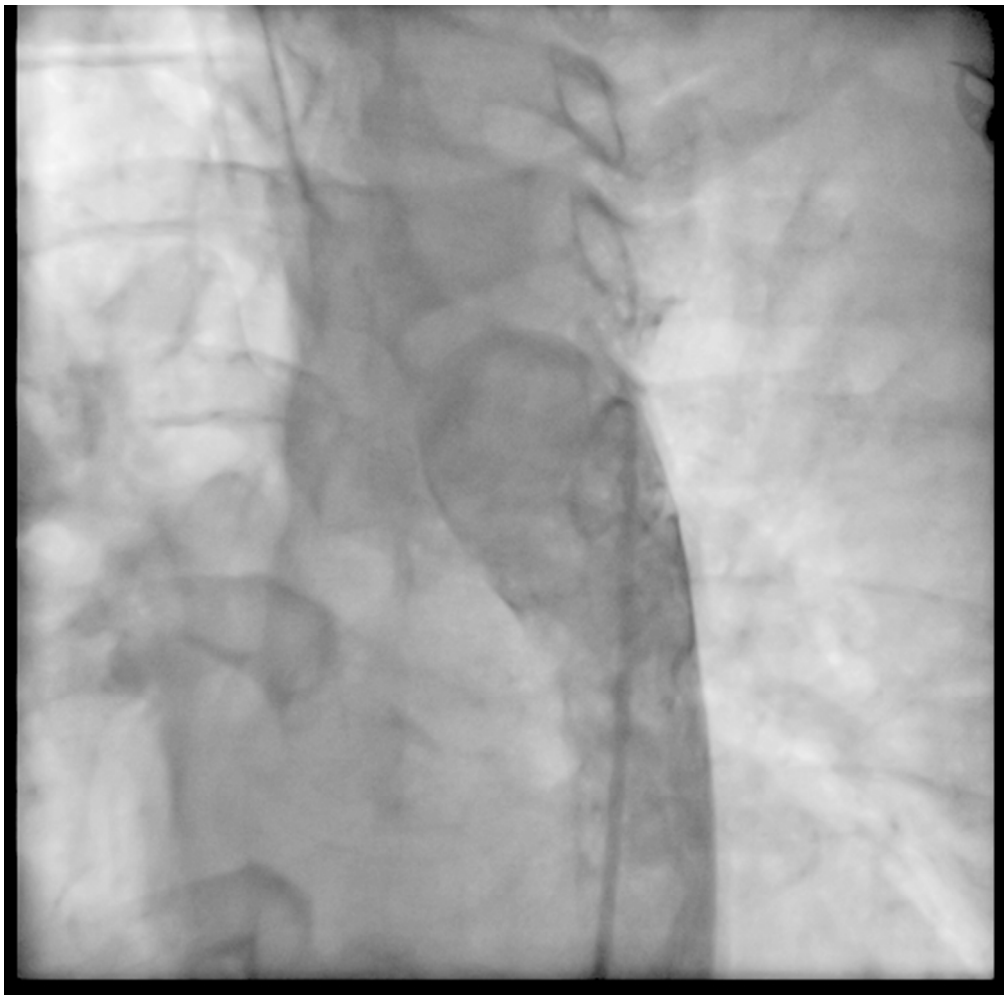
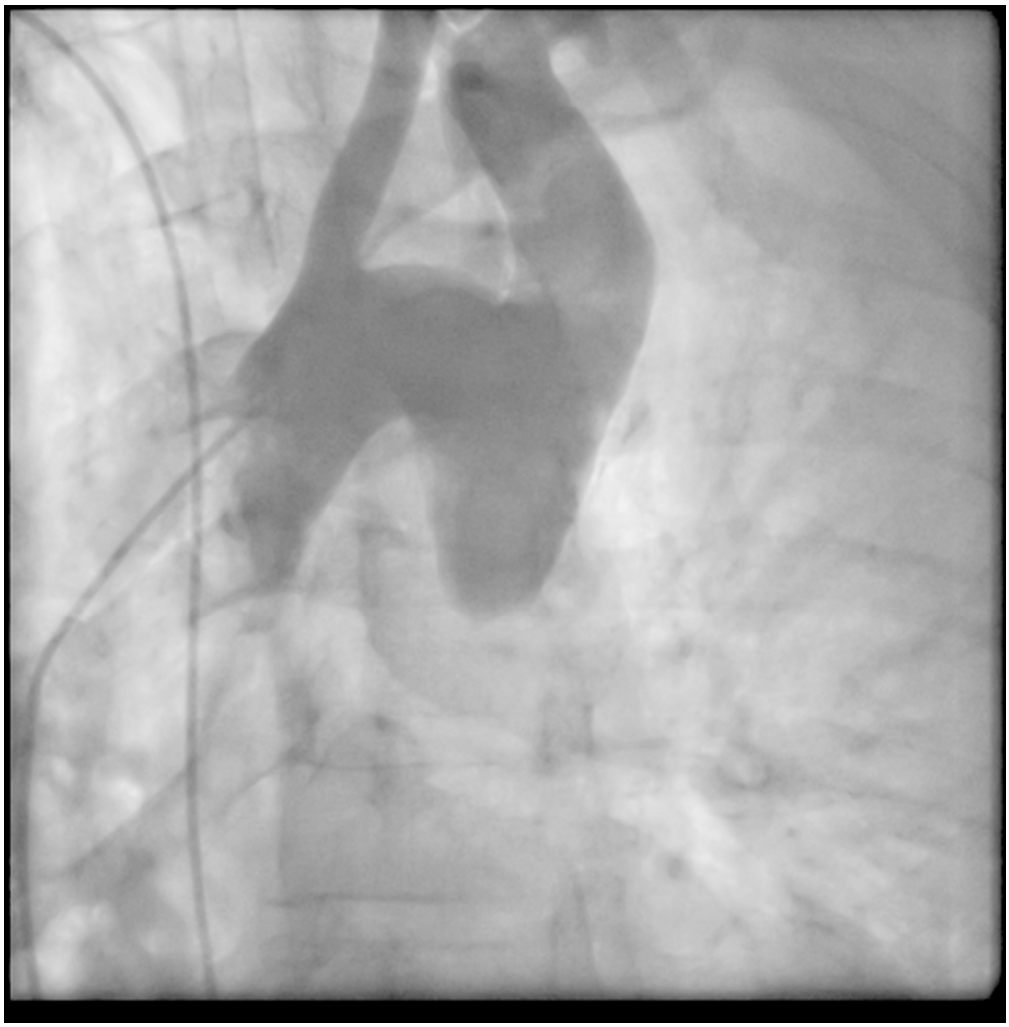


Radiocontrast injection across the descending aorta showing complete occlusion of the proximal part



Radiocontrast injection across the ascending aorta showing complete occlusion of distal part with luminal discontinuity

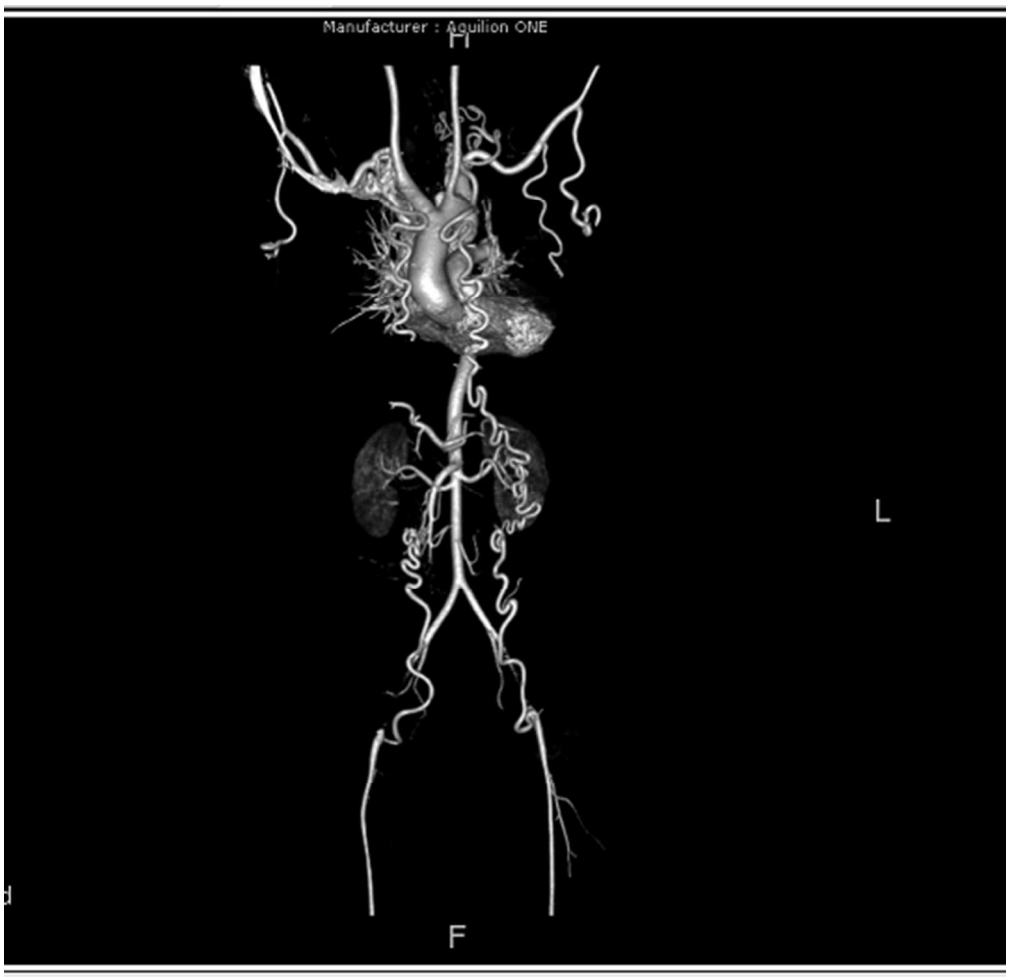
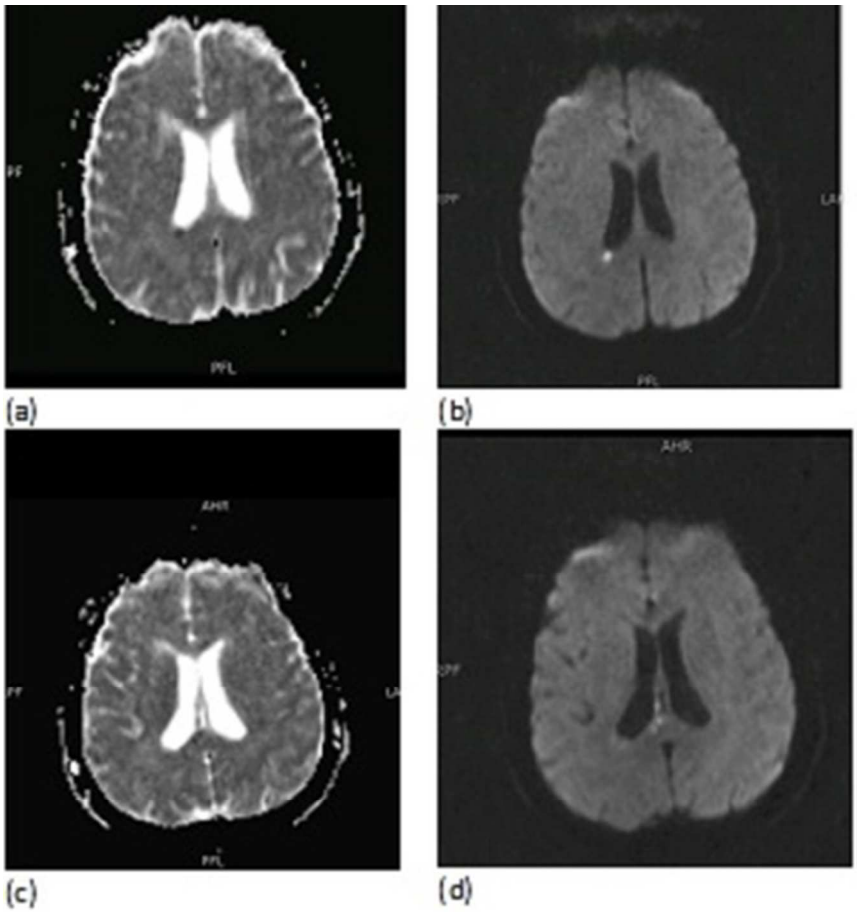


Image of the CT aortogram showing extensive collateral circulation



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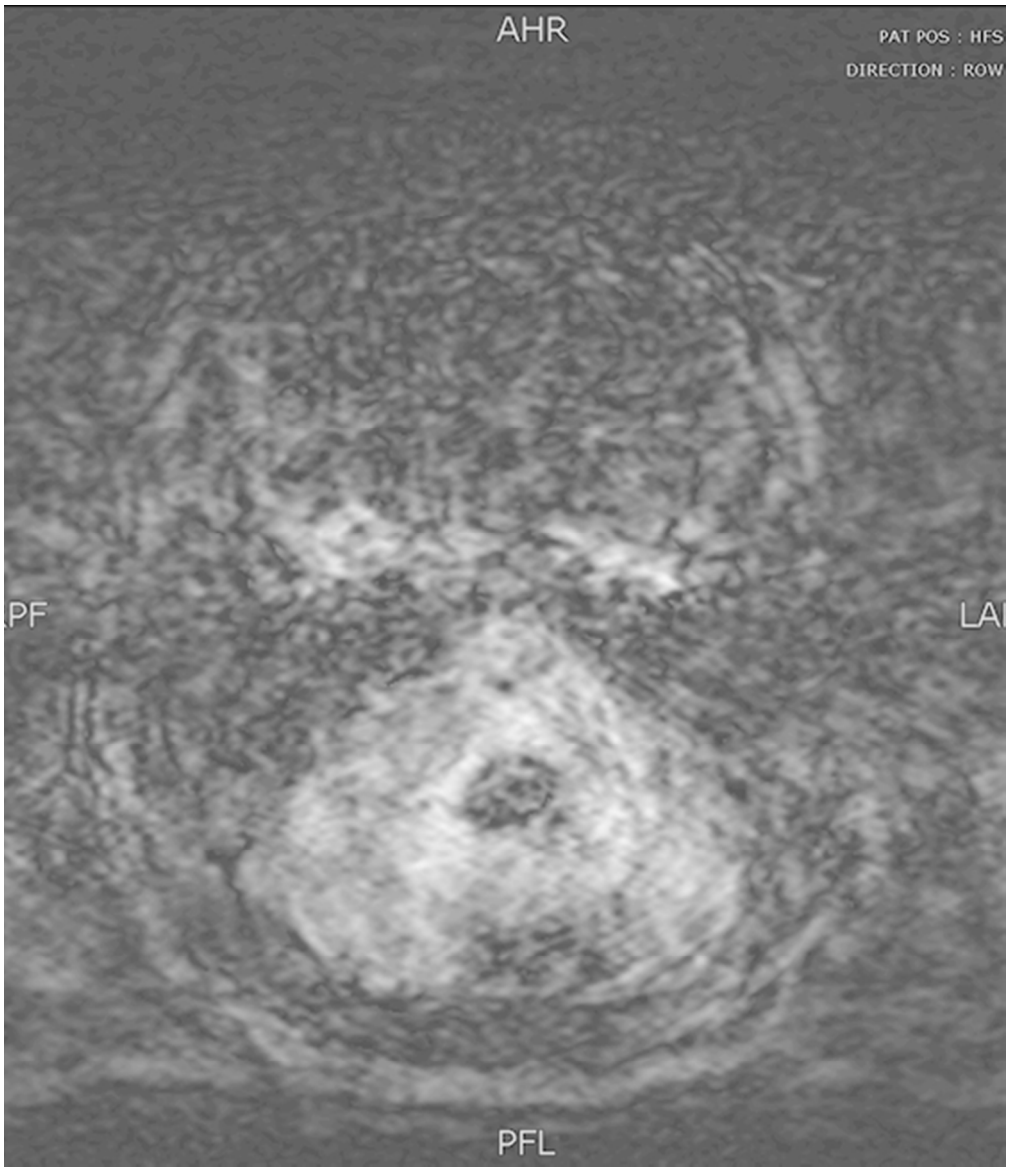

SWI showing signal dropout in fourth ventricle and bilateral parietal sulci likely representing hemorrhage