

Case Study

Title: "Imaging of congenital subclavian artery to subclavian vein fistula in a newborn"

Abstract:

The purpose of this paper is to report a congenital subclavian artery to subclavian vein fistula in a three weeks old male infant who presented with congestive heart failure and pulmonary hypertension. He had been scanned by various imaging modalities including echocardiography, cardiac computed tomography, diagnostic cardiac catheterization, cardiac magnetic resonance angiography and cranial computed tomoangiography. He had successful surgical ligation after failure of device occlusion trial at age of 9 months.

Keywords: Congenital Fistula, Subclavian artery, Subclavian vein, Imaging Modalities

Introduction:

An arteriovenous fistula (AVF) is an abnormal connection between an artery and a vein that results in shunting of blood between the two vessels, therefore bypassing the high-resistance capillary system. This anomaly may be congenital or acquired.

It was first recognized by Hunter in 1764 (1). In general; AVFs are rare but correctable causes of hyperdynamic circulation and congestive heart failure as well as pulmonary hypertension when it develops in utero (2). Congenital systemic AVFs are generally rare anomalies in infants and children. Congenital AFVs appear in the majority of cases in early childhood in absence of any traumatic factor, and the communications between the arteries and veins are usually multiple instead of single (3). AVFs in the head and neck region are also rarely encountered. The majority of cases involving the major vessels of the neck are of traumatic origin, either blunt or penetrating. Other causes of these fistulas include ruptured aneurysms, collagen deficiency syndromes, arterial dissections, fibromuscular dysplasia, and extensive infection of the neck (4). Congenital arteriovenous communications involving the subclavian artery and jugular veins are rare (5).

Acquired subclavian AVFs which can result from trauma or iatrogenic causes, are rare because these vessels have good protection by the overlying bony skeleton, surrounding muscular structures and sheath (6). Iatrogenic subclavian arteriovenous fistula most commonly occurs after vascular access in catheterization procedures and can cause heart failure and be limb-threatening (6). Although many arteriovenous malformations (AVMs) are asymptomatic, they can cause severe pain, hemorrhage or can lead to other serious medical problems as pulmonary embolism. In subclavian arteriovenous fistula, the initial symptoms and signs may be congestive heart failure or continuous murmurs (7).

CASE REPORT:

We are reporting a three week old male who was referred to our hospital with the diagnosis of subclavian artery to subclavian vein fistula. He was delivered via a spontaneous vertex at 34 weeks gestation with a birth weight of 1.8 kg. APGAR at 1 and 5 minutes were 6 and 8 respectively. He developed respiratory distress immediately after birth. He was diagnosed and managed as neonatal respiratory distress syndrome. He was treated with endotracheal surfactant, high frequency oscillatory ventilation and received nitrous oxide for pulmonary hypertension (PHTN) for some time. His family history was unremarkable and specifically negative for cardiopulmonary disorders. When admitted, he was mechanically ventilated; with oxygen saturation of 98% on fraction of inspired oxygen of 100% from the right arm; blood pressure of 81/43 mmHg; and pulse rate of 160 /min. Physical examination showed a bounding peripheral pulses in both upper and lower limbs, and a continuous machinery murmur at the left upper sternal border. The examination of chest, central nervous system, skin and mucosa did not reveal any abnormality. Abdominal examination showed enlarged liver.

Chest roentgenogram demonstrated cardiomegaly. Electrocardiogram (ECG) showed normal sinus rhythm, normal QRS axis, and no right ventricular hypertrophy. Other laboratory findings showed hemoglobin levels of 16 g/dl, normal liver function tests and renal profile with mildly elevated Partial Thromboplastin (PTT) and International Normalized Ratio (INR). Two-Dimensional echocardiographic examination showed situs solitus, levocardia, D-looped ventricles, and normally related great arteries. It also showed a dilated right side of the heart with moderate tricuspid valve regurgitation with peak systolic pressure gradient (PSPG) of 75 mmHg. There were 2 mm AVF between the subclavian artery and vein with PSPG of 50 mmHg with dilated superior vena cava as

well as subclavian vein and artery with continuous flow in the subclavian artery and diastolic run off in the descending aorta (Figure 1 & 2 Video 1).

In order to outline the lesion, cardiac computed tomography (CT) was performed the next day of admission and showed AVF connecting the left subclavian artery to the large subclavian vein at the level of the middle half of the clavicle (Figures 3-6). Cranial ultrasound was normal initially with no hemorrhage or hydrocephalus.

The patient underwent cardiac catheterization with an attempt to coil occlude the fistula. The cardiac catheterization confirmed the presence of AVF (Videos 2 A & B). However due to the close proximity of the subclavian artery and vein, presence of vital branches and the concern that any type of device would occlude some of these vital branches and can cause progressive intra vascular thrombosis; the intervention was not performed and the case was referred to the surgical team. Due to his small size, the operation was postponed and the child was followed conservatively with anti-heart failure measures and the surgical team planned to do ligation at age 7-8 months if clinical condition permits. Cardiac magnetic resonance angiocardiography (CMRA) was performed at the age of 7 months and showed evidence of left subclavian artery to the left subclavian vein fistula (Figure 7). Because the infant had delayed neurological development; brain CT angiography was performed at the same time and showed abnormally dilated venous varix in the medial aspect of the middle cranial fossa (Figure 8).

Surgical ligation of the fistula was done at age of 9 months with excellent results with complete anatomic correction and rapid resolution of right sided cardiomegaly and pulmonary hypertension. The dilated intracranial venous system (which resulted likely from chronic venous hypertension) was significantly improved following fistula ligation. He continued however to have brain atrophy with extra axial fluid collection.

Discussion:

AVF is an abnormal congenital or acquired direct vascular channel between an artery and a vein that results in shunting of blood between the two without an intervening capillary network (4). The congenital type is frequently more anatomically complex and is often too widespread for device occlusion and often need surgical intervention. Congenital fistulas are composed of the embryonic mesoderm that result from an arrest in the embryological differentiation during the development

process of the vascular system with failure of the primitive plexiform structures to differentiate normally into mature capillary and venous networks. This leads to presence of abnormal arteriovenous channels; which could be single or multiple, local or diffuse, and visceral or cutaneous (7). It was also noted that there is higher incidence of AVF involving right subclavian artery more than the left which could be related to the complexity of the embryologic development of the right subclavian artery (8).

Despite congenital AVF are rare but they are important causes of heart failure in newborns, infants and young children. In our case the infant presented with high cardiac output heart failure which denoted significant shunting of blood from the high pressure subclavian artery to the low pressure subclavian vein through AVF. The volume and velocity of blood flow shunted through the fistula will depend on size of a fistula and its proximity to the heart and the pressure gradient. The nearer to the heart and the wider is the fistula, the more the increase in cardiac output and the more will be the cardiac work. For example, a small fistula from a branch of the aortic arch may result in more cardiac work than a larger one in the leg or forearm (9). In our case, the fistula was close to the heart and involving a major artery; so it presented with neonatal respiratory distress and heart failure.

There are few reports about AVF involving the subclavian artery. The fistula may connect the subclavian artery with subclavian vein, innominate veins or internal jugular vein (10, 11). These fistulae have a wide range of clinical manifestations from being asymptomatic to present with frank heart failure. Peripheral pulses are bounding due to hyperdynamic circulation like what was found in our case. Local examination may reveal a continuous bruit in the upper chest, loudest at the right or left infra-clavicular area (12). Congenital AVF usually present with heart failure in newborn (as in our case), infants and young children. Pulmonary hypertension can result from giant AVF (13). However, congenital AVF involving subclavian artery and vein was previously reported in 2 adults (14, 15).

There are different diagnostic modalities that help in diagnosing these types of fistulae. X-ray may show variable degrees of cardiomegaly (as that encountered in our case) with possible increased pulmonary vascular markings, widened mediastinum and prominence of the superior vena caval shadow. The cardiac silhouette may be similar to the "snowman" or "hourglass" configuration seen in older patients with total anomalous pulmonary venous return (15). However, the uncommon location of the fistula may produce other radiologic findings and can be misleading. ECG was normal in our

case. ECG may be normal or show right axis deviation, right and left atrial enlargement and right ventricular hypertrophy pattern. Sometimes ECG shows left ventricular hypertrophy (11, 15).

In our case, Two-dimensional echocardiography succeeded to visualize the fistula and detected the cardiac changes related to the effects of the fistula including cardiomegaly and increased pulmonary artery pressure. Two-dimensional echocardiography with color Doppler imaging may aid the initial diagnosis. Various echocardiographic modalities can help in evaluation of AVF. M-mode may show dilated right atrium, right ventricle, left atrium, and/or left ventricle. Adequate echocardiographic examination can efficiently visualize the fistula however, in some cases it may be difficult and other diagnostic tools may be needed; at least to confirm the diagnosis (16).

Computed tomographic and magnetic resonance angiography can provide images of the anatomy of the arteriovenous communication, typically with early contrast filling in the vein during the arterial phase. Detailed anatomical views of the involved vessels and the site and size of the AVF is helpful when making decisions about the best treatment option. Cardiac CT scan has increased risk of ionizing radiation but it has very short study time, its spatial resolution is as little as 0.4 mm. Meanwhile, the cardiac magnetic resonance imaging (CMRI) can acquire cine image in any desired plane without the risk of ionizing radiation; available post-processing tools and real time images can be obtained without intravenous contrast. However, CMRI has long study time and its long axis resolution is just below 1 mm (17). Time-resolved non-contrast enhanced 4-Dimensional dynamic magnetic resonance angiography can provide detailed information on the location and size of the fistula and the dynamic flow of blood similar to what we can get by invasive angiograph. Angiograph during cardiac catheterization is not usually required for routine diagnosis but may be used at the time of therapeutic procedures or when other non-invasive imaging fails to identify this abnormality (15).

Closure of the fistula is recommended because of their tendency to become hemodynamically more significant with time, the risk of bacterial infection, pulmonary embolism and thrombosis and the possibility of developing aneurismal or degenerative changes or even rupture. If amenable, trans-catheter occlusion of the fistula can be done, however, subclavian fistula is difficult for trans-catheter occlusion because of the close proximity to the heart and the major vessels and we failed to do trans-catheter occlusion of the fistula in our case. Nevertheless, surgery is usually the recommended treatment for either symptomatic or non-symptomatic arteriovenous fistulas if the fistula is not

amenable to trans-catheter occlusion or after its failure as in our case. The choice of treatment is determined by the location and anatomy of the lesion (15, 18).

Conclusion:

Congenital subclavian AVF may present with congestive heart failure very early during the neonatal region or late during adulthood. There are different diagnostic modalities including Two-dimensional echocardiography with color Doppler imaging, computed tomographic and magnetic resonance angiography and conventional angiography. Early closure of the AVF is indicated to avoid many complications including heart failure, infection or aneurismal dilations.

COMMENTS

Case characteristics

Three weeks old male was referred to our hospital with the diagnosis of subclavian artery to subclavian vein fistula. Documented clinical presentation and progress, imaging findings and operative treatment were investigated.

Clinical diagnosis

The baby presented with respiratory distress immediately after birth and diagnosed and managed as neonatal respiratory distress syndrome. After, that signs of heart failure with bounding peripheral pulses in both upper and lower limbs, normal and a continuous machinery murmur, which were heard at the left upper sternal border.

Imaging diagnosis

Chest roentgenogram demonstrated cardiomegaly. Echocardiographic study showed a dilated right side of the heart, moderate tricuspid valve regurgitation and 2 mm arteriovenous fistula between the subclavian artery and vein with PSPG of 50 mmHg with dilated superior vena cava as well as subclavian vein and artery with continuous flow in the subclavian artery and diastolic run off in the descending aorta. Cardiac magnetic resonance angiography (CMRA), cardiac CT and cardiac catheterization was done and confirms the diagnosis.

Differential diagnosis

Subclavian artery to subclavian vein fistula should be differentiated from causes of hyperdynamic heart failure, other congenital arteriovenous fistulas and total anomalous pulmonary venous drainage.

Treatment

Trans-catheter occlusion if amenable, surgical closure will be determined by the location and anatomy of the lesion

Related reports

There are few reports about AVF involving the subclavian artery.

Experiences and lessons

Closure of the fistula is indicated to avoid many complications including heart failure, infection or aneurismal dilations.

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251 Figure 1: 2-D Echocardiogram showing the aortic arch with dilation of the left subclavian artery.

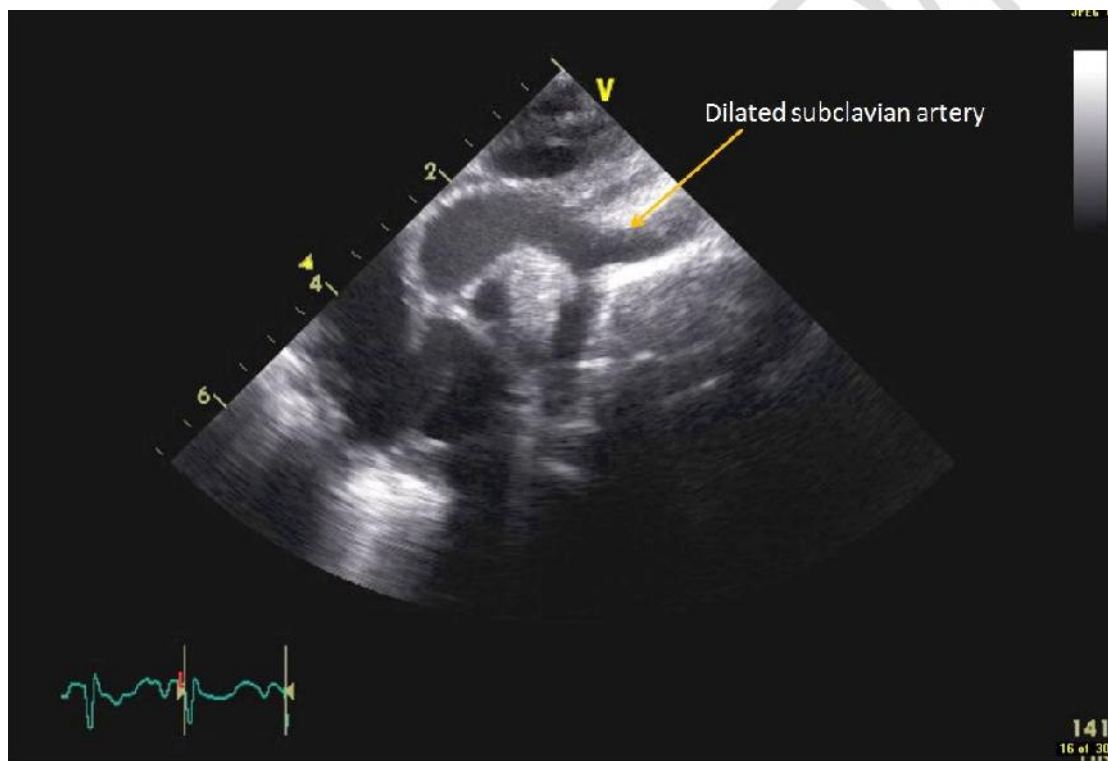


Figure 2: 2-D Echocardiogram with color flow Doppler showing dilated left subclavian artery with the fistula connecting the left subclavian artery and vein.

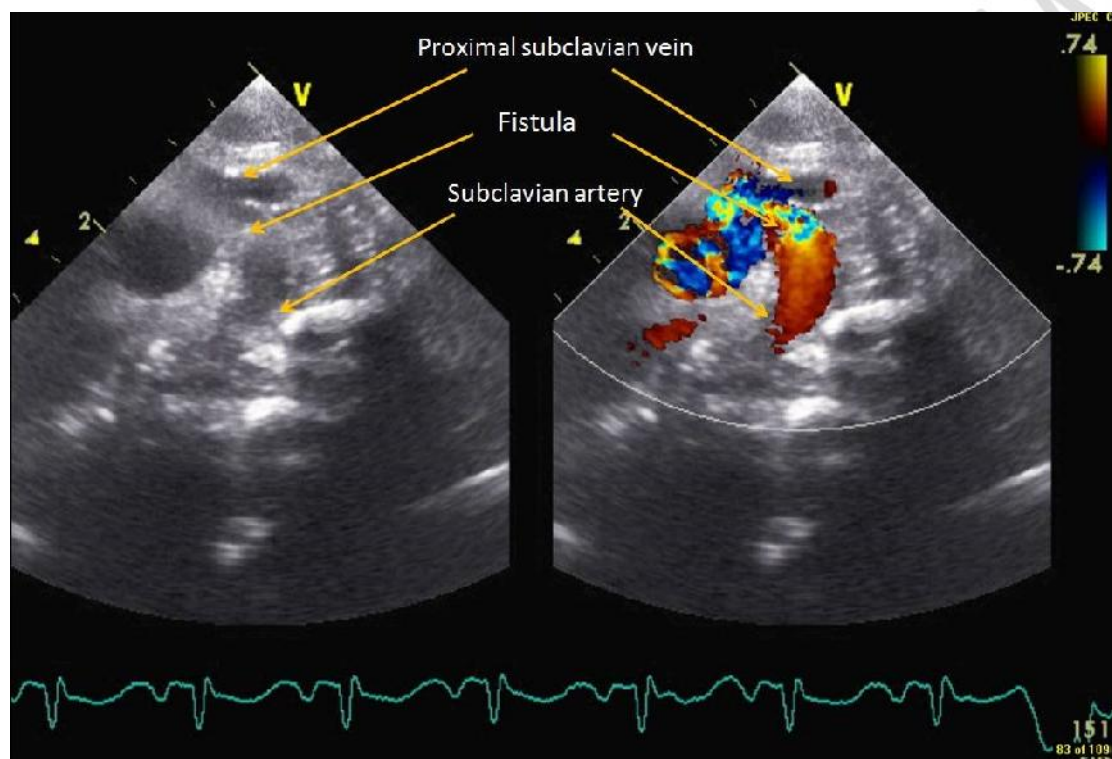
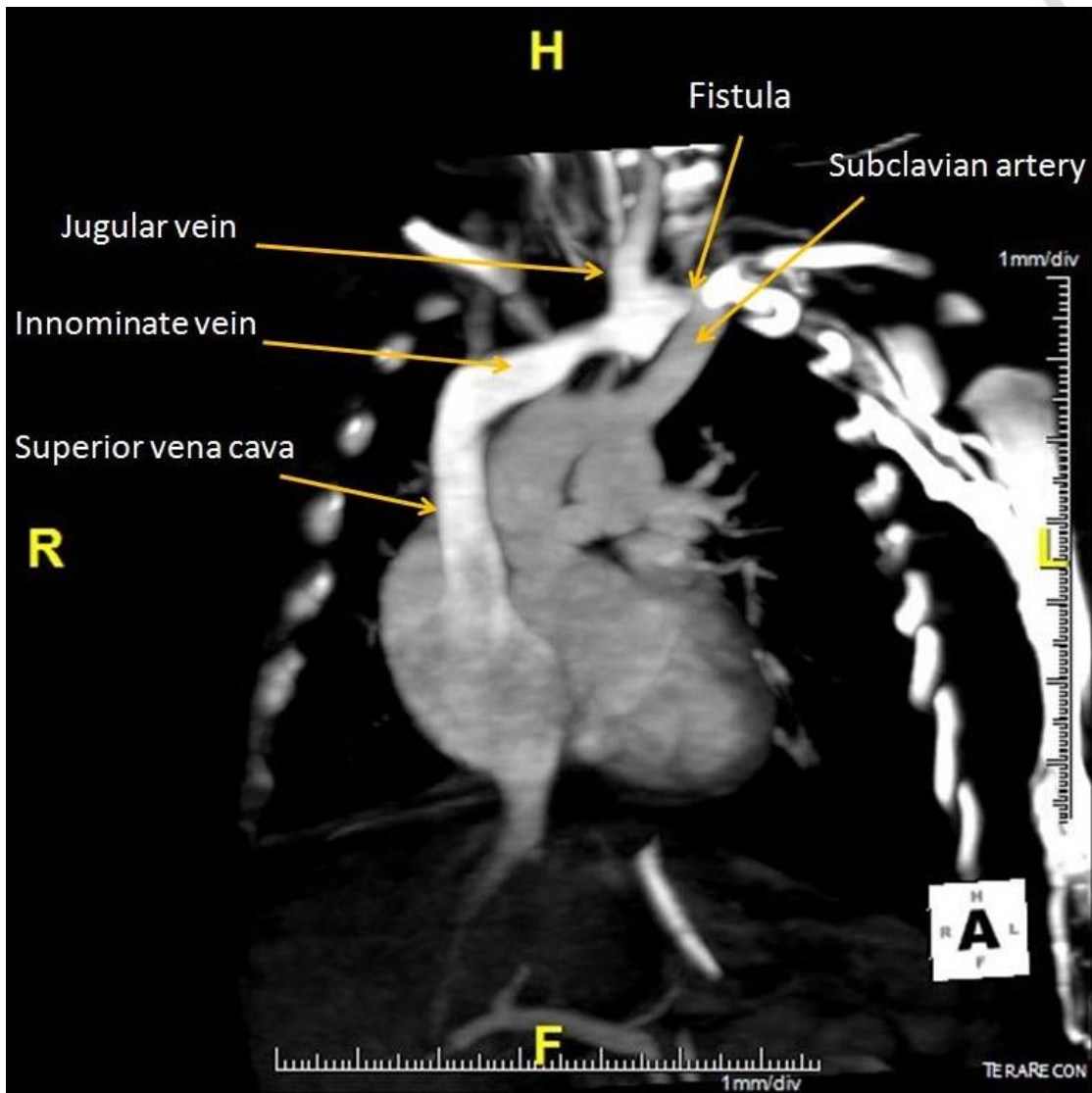


Figure 3: CTA image showing the site and size of the fistula with dilated innominate vein, left subclavian artery, jugular vein and superior vena cava.



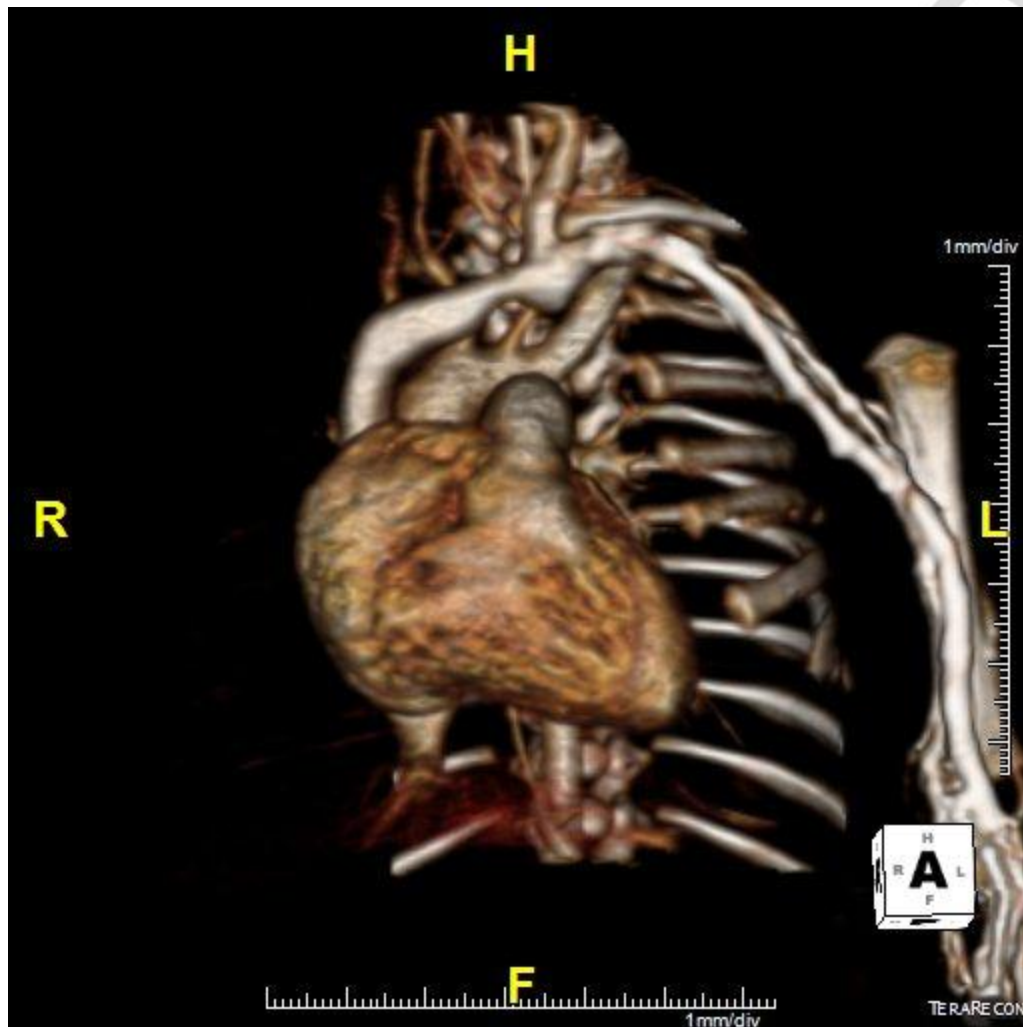
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286 Figure 4: 3-D CTA reconstruction showing dilated innominate vein, left subclavian artery,
287 jugular vein and superior vena cava and the relation to the clavicle. You can also see the dilation
288 of the venous system of the left arm.

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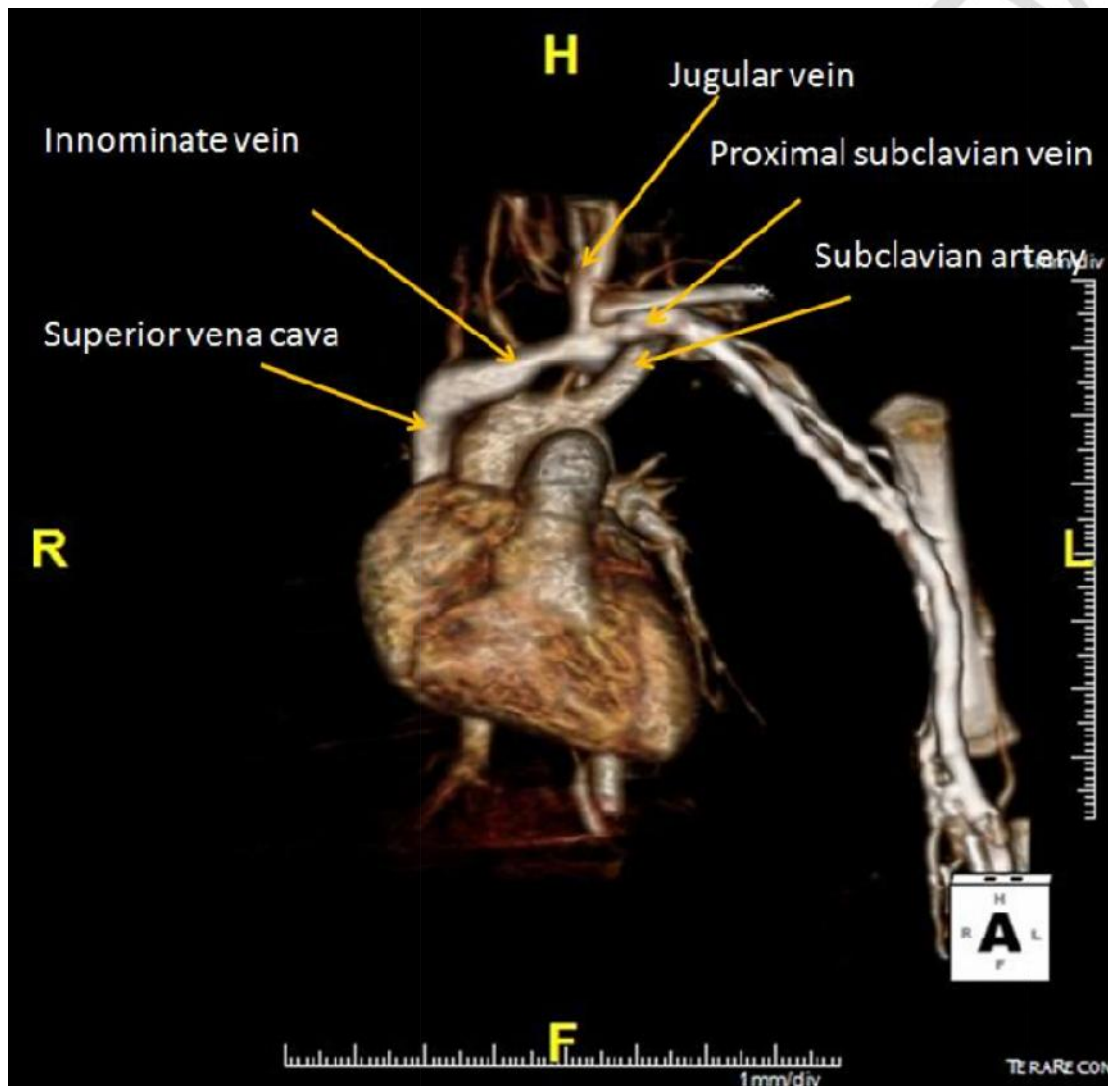
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298 Figure 5: 3-D CTA reconstruction showing dilated innominate vein, left subclavian artery,
299 jugular vein and superior vena cava and the relation to the clavicle. You can also see the dilation
300 of the venous system of the left arm.



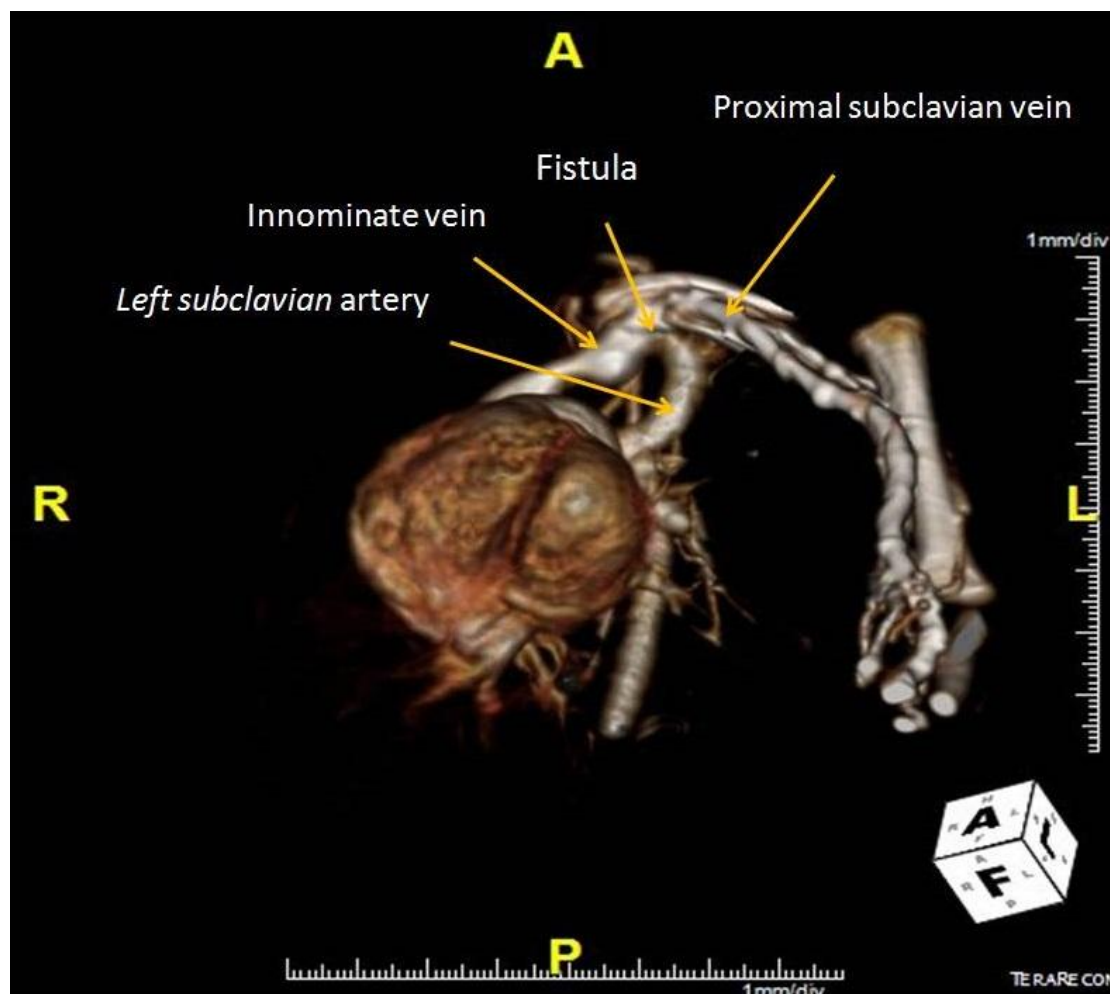
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Figure 6: 3-D CTA reconstruction showing the site of the fistula.

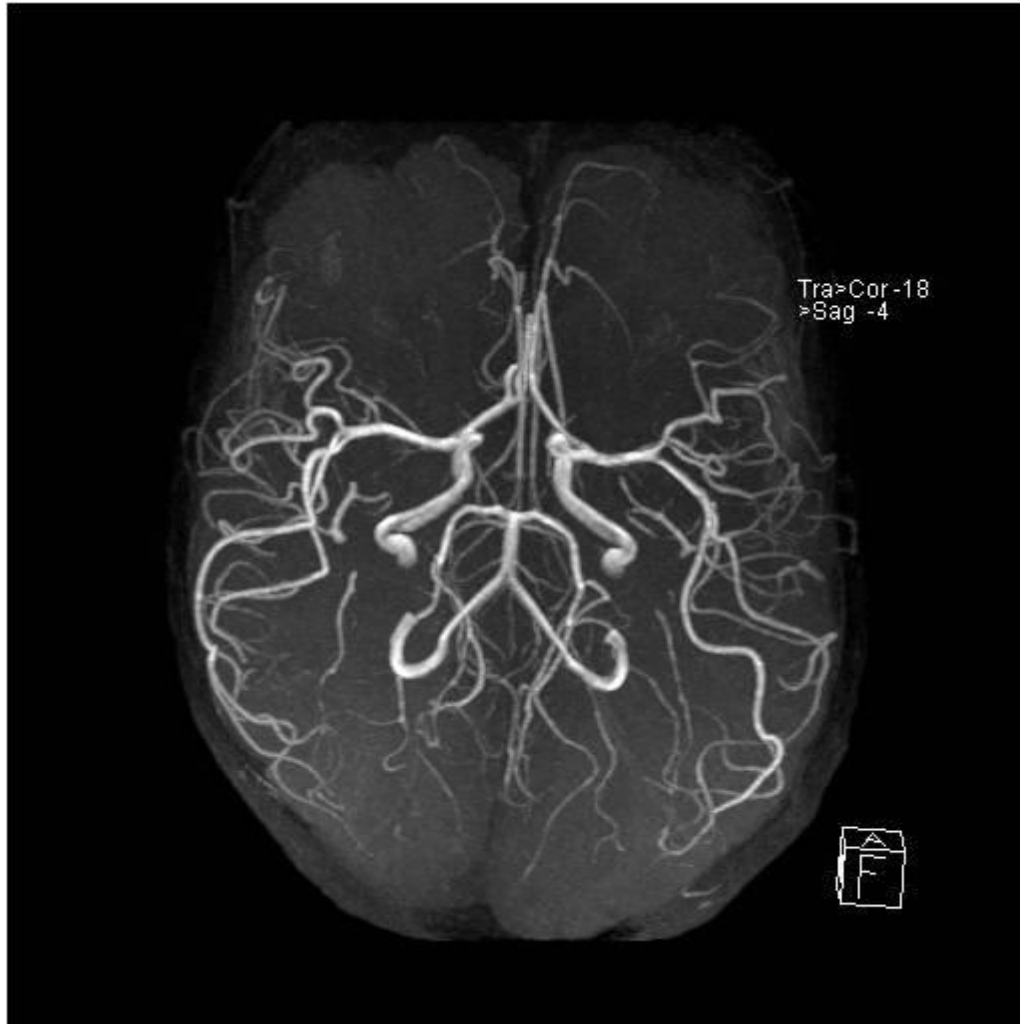


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319 Figure 7: MRA neck vessels showing dilated left subclavian artery, internal jugular vein with
320 dilated intracranial veno-venous collaterals (the innominate vein did not fill possibly due to
321 thrombosis following central line placement).



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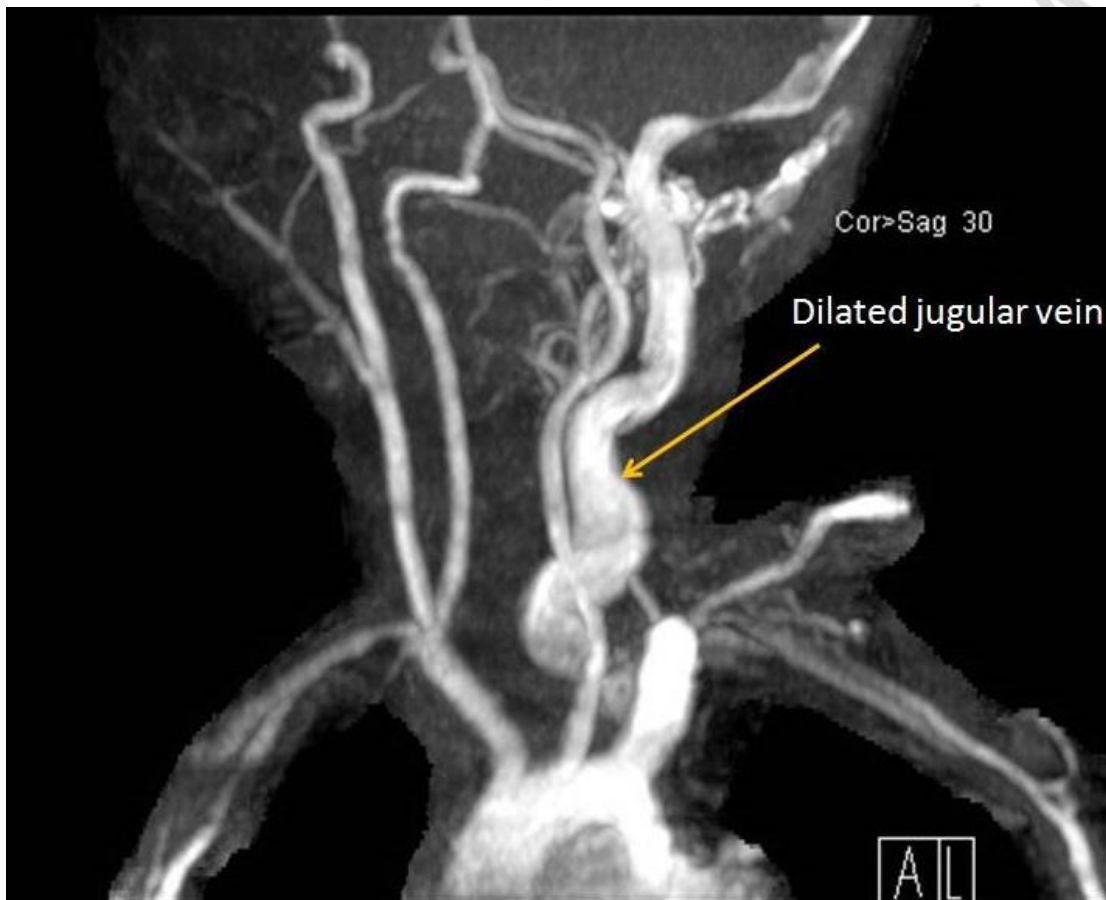
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330 Figure 8: CTA brain showing no other abnormal arterio-venous connections. In this arterial
331 phase you do not appreciate the venous system dilation or plexus of network.

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336 Video 1: 2-D Echocardiogram with color flow Doppler of Suprasternal view showed dilated left
337 subclavian artery with the fistula connecting the left subclavian artery and vein.

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340 Video 2 A & B: Left ventricular angiogram demonstrating fistula between left subclavian artery
341 and vein. Note rapid filling of superior vena cava and right atrium. The lateral view showed
342 clearly the fistulous tract between left subclavian artery and vein.

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Video 1 Fistula.avi

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Video 2 A.avi

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Video 2 B.avi

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