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Case report

Misdiagnosis of Pulmonary embolism in a Fontan's patient: When standard protocol CT Pulmonary angiogram is inadequate☆,☆

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ABSTRACT

Patients who have undergone a Fontan's procedure have an abnormal circulation that presents a unique challenge when performing computed tomography pulmonary angiograms. In a standard imaging protocol, contrast is injected into the upper limb veins that feed into the superior vena cava. In Fontan's patients the Computed tomography pulmonary angiograms bypasses the heart and preferentially fills the right lung, with only a small amount of mixture of contrast and noncontrast blood in the pulmonary arteries.

In this article, we present the case of a 35-year-old female complaining of chest and abdominal pain with oxygen saturations of 85% on room air. Computed tomography pulmonary angiograms showed suboptimal imaging of the left lung and apparent filling defects in the right lung suggesting a radiological diagnosis of a pulmonary embolism. The abnormal flow and distribution of contrast in the pulmonary arteries can result in a false positive diagnosis of pulmonary embolism. To overcome this, experts advise using a dual-injection of contrast via upper and lower limb central veins to achieve optimal imaging.

Background

Fontan procedures are performed in children with congenital heart abnormalities that cannot support a biventricular circulation. The underlying abnormality often involves tricuspid atresia, whereby these patients have parallel pulmonary and systemic circuits, sharing a single ventricle.

The Fontan procedure was first performed in 1968 [1]. The procedure involves redirecting blood from the superior vena cava (SVC) and inferior vena cava directly into the pulmonary arteries, bypassing the heart. Oxygenated blood from the pul-

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monary veins enters the left atria and is pumped to the systemic circulation by the heart's single ventricle.

More recent variants in the procedure have included fenestrations which allow some flow of blood from the Inferior vena cava (IVC) directly to the right atria in order to reduce pulmonary pressures as in this case presented. Other recent variations utilize the right atria to pump blood from the systemic venous system into the pulmonary arteries.

Clinical presentation

A 35-year-old female presented to hospital with 2 days of non-specific chest and abdominal pain. She also noted that her lips appeared to have a more bluish color than normal, indicating worsening cyanosis. She was not known to any local consultants and had not attended this hospital before.

At birth she had tricuspid atresia with a univentricle and mal positioning of the great vessels. She had several operations as an infant and underwent a Fontan procedure at the age of 7. She is currently being considered for heart transplant. Her only comorbidities included liver cirrhosis secondary to increased venous pressure.

On admission she was found to be saturation at 85% on room air. She informed the clerking team that she had a previous Fontan's procedure and that her baseline oxygen saturations were 88%-92%. Her d-dimer was negative (less than 500). Ultrasound scan (USS) of liver was performed that noted no new changes to her preexisting liver cirrhosis. Over the course of her admission her oxygen saturations stabilized and her observations were all within normal limits. She was discharged a day later.

Four days later she was re admitted under the surgical team with sudden onset abdominal pain lasting 4 days with tenderness in right iliac fossa. It was thought that this was potentially caused by liver capsular pain and a computed tomography (CT) Chest abdomen pelvis was requested, with a CT pulmonary angiogram to rule out a pulmonary embolism (PE).

Imaging findings

The initial report states the following, "Opacified blood flows from the right upper limb peripheral veins, through the SVC into the right pulmonary artery, right pulmonary vein, and eventually into the dilated ascending aorta. The arch vessels and descending aorta appear grossly normal."

The report states "Within the limits of the unusual circulation, admixture of contrast and phasing of the scan, there appear to be segmental emboli in the right pulmonary arteries," as can be seen in Images 1A. There is also a lack of contrast filling of the right main pulmonary artery as seen in Image 1B. Images 1A and B also show the lack of any visible contrast in the left pulmonary arteries.

The Fontan circulation can be seen clearly in Images 2 and 3 demonstration contrast from the SVC flowing directly into the right pulmonary arteries (Image 2) and noncontrast blood from the IVC flowing directly into the left pulmonary arteries (Image 3).

The CT Chest abdomen pelvis also noted a lesion on the right kidney suggestive of a right sided pyelonephritis.

Treatment and outcome

Treatment dose dalteparin was commenced along with empirical antibiotics to cover for pyelonephritis.

Due to the rarity of this case, the CT images were discussed with a local cardiologist, a local consultant radiologist, and her Grown-up-Congenital-Heart-Disease consultant. She was found to be saturating at baseline and a decision was jointly made that there was no need for a repeat Computed tomography pulmonary angiograms (CTPA) with a dual injection technique due to low clinical suspicion of a PE. Treatment dose dalteparin was stopped. An amendment was made to the original CTPA report after the discussion. The amendment recognized the preferential filling of the right lung via the SVC and that this can produce flow phenomenon that resemble filling defects.

The patient's clinical signs and symptoms were re-evaluated, and she was discharged on oral antibiotics to treat a right sided pyelonephritis.

Discussion

The altered anatomy after a Fontan's procedure results in an atypical flow of blood from the systemic venous system to the pulmonary arterial system, which presents a challenge when using standard contrast injection protocols. In a normal circulation contrast enhanced blood from the SVC is mixed with noncontrast blood from the IVC in the right ventricle leading to equal distribution through the right and left pulmonary artery. In a Fontan circuit, the lack of this sub-pulmonary ventricle results in an uneven distribution of contrast through the pulmonary arteries [2] and suboptimal timings of pulmonary artery enhancement [3]. This can mimic the appearance of a 1-sided PE in the lung that is preferentially supplied by the IVC. It can also mimic sub segmental emboli in the lung preferentially supplied by the SVC due to incomplete mixture of enhancing and nonenhancing blood.

Pulmonary artery filling defects can also be seen in non-thrombotic pulmonary emboli, primary pulmonary arterial neoplasm, Takayasu's arteritis, pulmonary arterial streak artefact [4]. These are all relatively uncommon however and were not likely to be the cause of these patients filling defects.

In this case a repeat CTPA using dual injection of contrast into the upper and lower limb could have been used to definitively rule out a PE, but based on clinical findings it was deemed unnecessary.

This abnormal flow phenomenon can result in unnecessarily treatment with anticoagulation of thrombolytics in these patients [5]. A study by Sandler et al 2016 reviewed CTPA imaging in patients that had previously undergone a

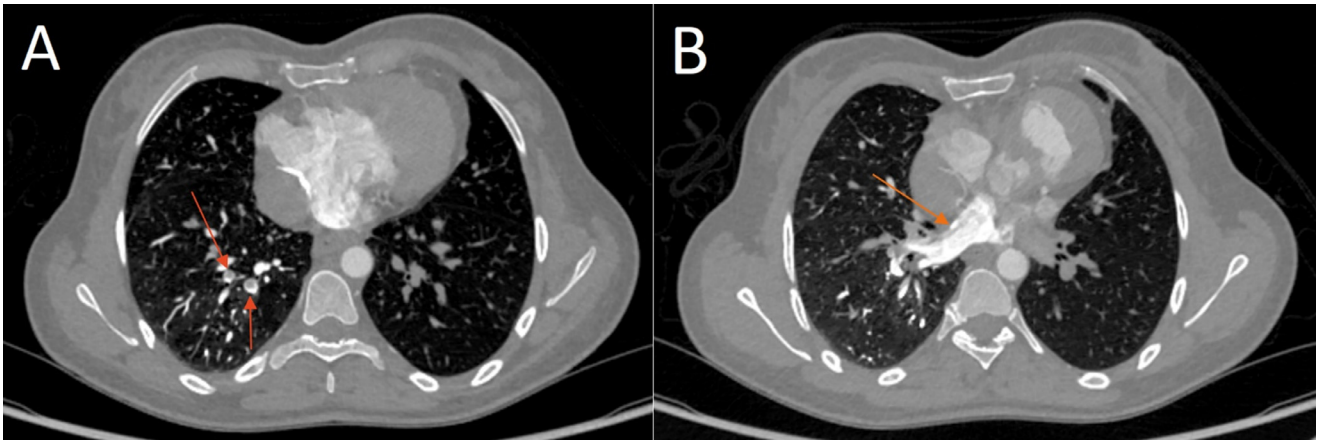

Image 1 – (A) and (B) Axial CTA images after contrast injection into the right cephalic vein. The right pulmonary arteries appear to have filling defects, similar to those typically seen in a PE. Filling defects can be seen in the pulmonary arteries as shown by the red arrows in figure A and the pulmonary trunk shown by the red arrow in figure B. The left pulmonary arteries have little to no contrast in them.

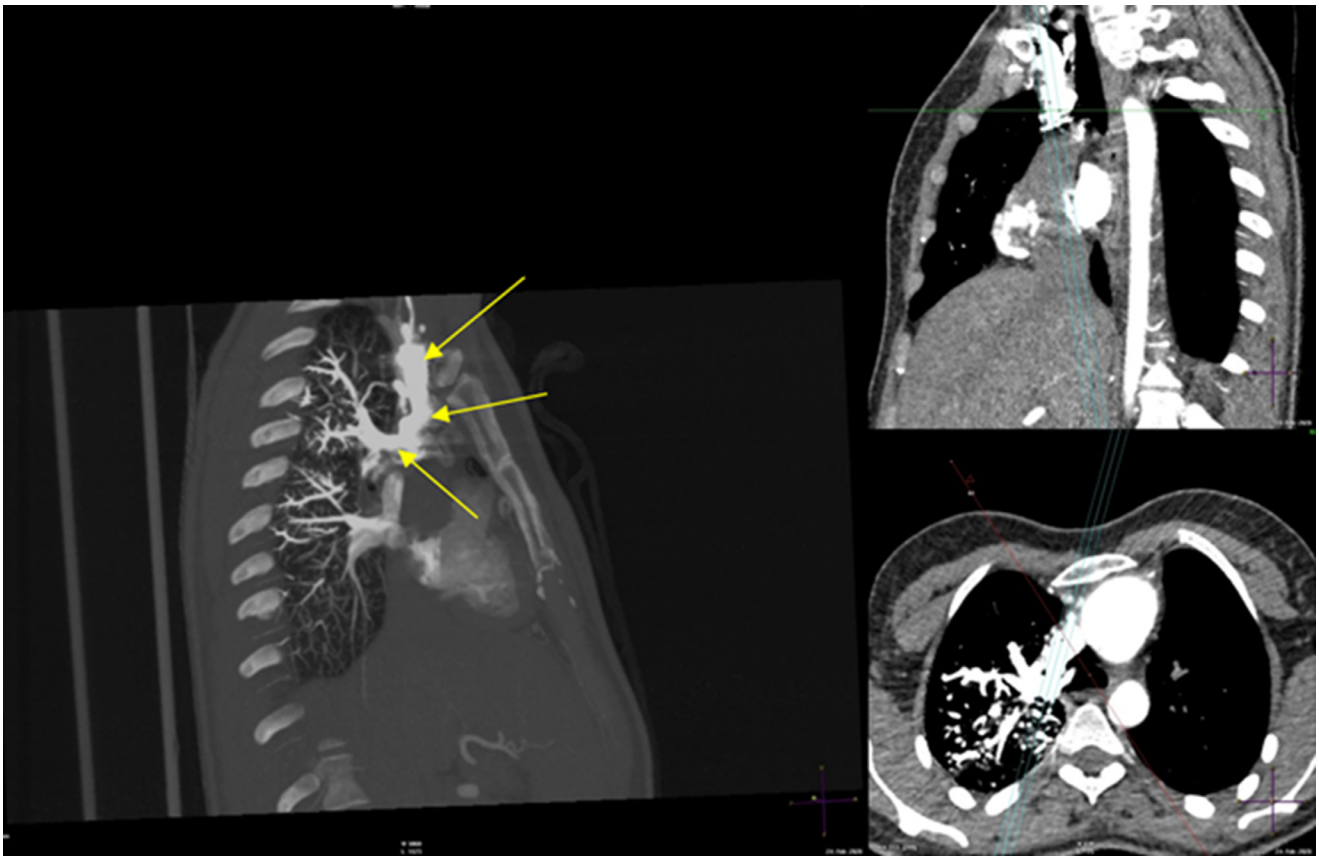

Image 2 – Sagittal oblique CTA image. The yellow arrows show the course of contrast flowing from the Superior vena cava directly into the right pulmonary artery.

Fontan procedure. 17 out of 19 had suboptimal imaging and 7 received anticoagulation therapy due to misdiagnosis of pulmonary emboli.

Thromboembolic disease is one of the leading causes of mortality in patients that have undergone Fontan surgery [6] as their altered anatomy predisposes them to an increased risk of intracardial thrombus formation and subsequent pulmonary emboli [7]. It is therefore critical that optimized imaging protocol is known and utilized to avoid suboptimal imaging or misdiagnosis.

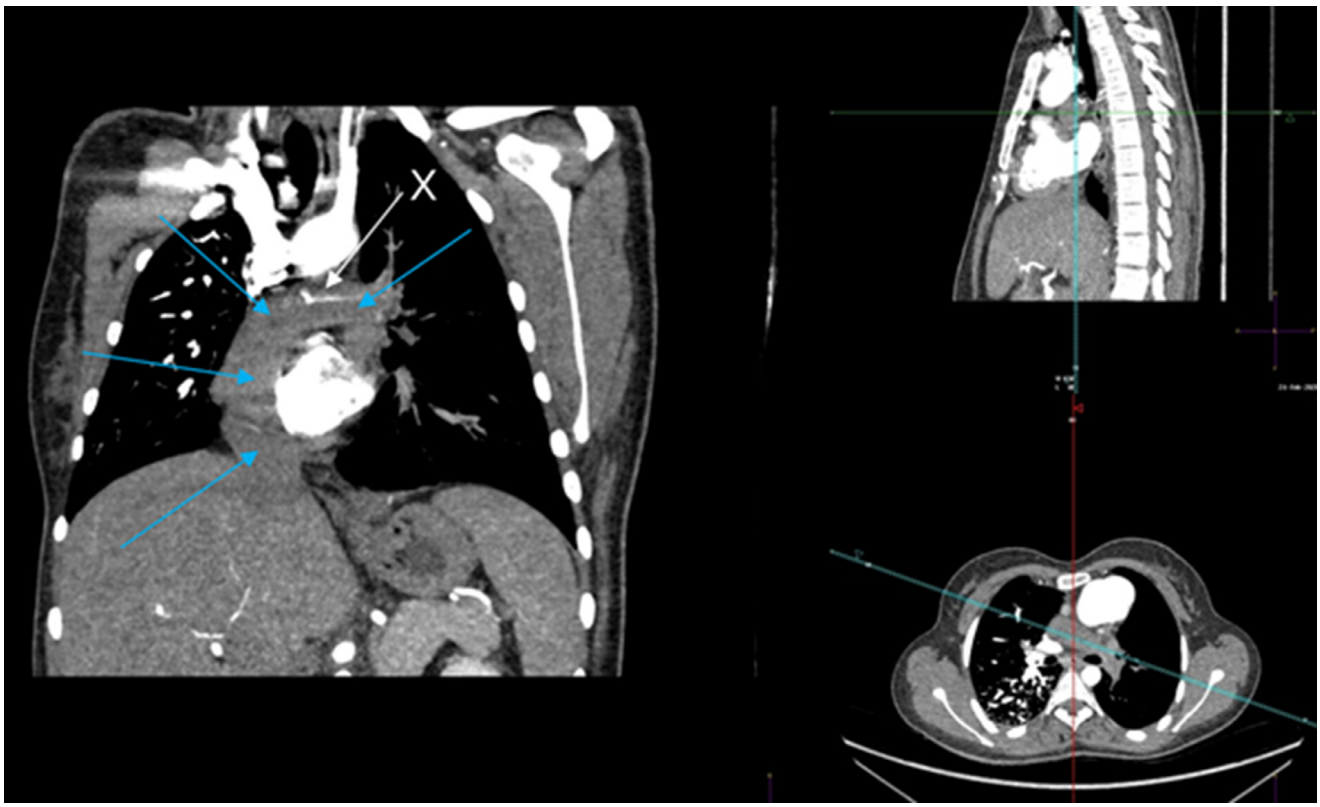

Image 3 – Coronal oblique CTA image. The blue arrows track the course of the Inferior vena cava with noncontrast blood, flowing directly into the left pulmonary arteries. A small amount of contrast can be seen at arrow X where the IVC becomes the left pulmonary artery demonstrating some mixing of contrast and noncontrast blood.

Studies show that a simultaneous injection of contrast into upper and lower limb veins leads to a more uniform distribution of contrast in the pulmonary arteries [3].

Learning points

1) Standard single contrast injection in patients that have undergone a Fontan procedure will result in suboptimal imagine and can lead to a misdiagnosis of pulmonary embolism.
2) Dual contrast injection via the upper and lower limb veins is the best practice to assess pulmonary blood flow and to rule out a PE.

Consent

Written informed consent was obtained from the patient for publication of this case report, including accompanying images.

Acknowledgment

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