

FATAL DISSEMINATED PARADOXICAL EMBOLISM IN INFERIOR SINUS VENOSUS ATRIAL SEPTAL DEFECT

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ABSTRACT

Inferior sinus venosus atrial septal defect (ASD) is a rare congenital cardiac anomaly. Similar with other types of ASD and patent foramen ovale, this defect results in arteriovenous shunting with the risk of developing paradoxical embolism (PDE) to the systemic circulation from venous emboli. We report a case of a 67-year-old lady who presented to emergency department with massive pulmonary embolism (PE) and recurrent acute limb ischaemia. Computed tomography pulmonary angiography also showed an incidental finding of inferior sinus venosus ASD which led to the diagnosis of PDE. Intravenous thrombolysis was administered followed by open mechanical thrombectomy. The patient developed massive lower gastrointestinal bleed post thrombolysis and passed away despite embolization treatment. This case report describes the catastrophic effect of PDE leading to disseminated multisystem thromboembolism. It also emphasizes the importance of early detection of a right-to-left shunt in patients who present with PE and recurrent acute limb ischaemia of indeterminate aetiology. Transthoracic echocardiography has lower sensitivity than transoesophageal approach in detecting inferior sinus venosus ASD.

Keywords: arteriovenous shunting, pulmonary embolism, limbs ischaemia

INTRODUCTION

Atrial septal defect (ASD) refers to a group of congenital cardiac anomalies that allows communication between the right and left atrium. It includes defects in the interatrial septum and at the cardiac terminations of the systemic and pulmonary veins i.e. sinus venosus defects.¹ ASD accounts for 10 to 15% of congenital heart disease, with a reported birth prevalence of 1 to 2 per 1000 live births.² Presence of ASD leads to an increased risk of paradoxical embolism (PDE) with reported incidence of up to 14%.³ PDE refers to the passage of venous thrombi into the systemic circulation via an arteriovenous shunt. It is a rare phenomenon and accounts for less than 2% of all arterial

emboli.⁴ The risk of PDE in patients with ASD increases with age due to chronic left ventricular hypertrophy which has decreased compliance and results in progressive enlargement of the ASD.⁵ Accurate diagnosis of this condition is challenging due to the variation in clinical manifestation and proven to be fatal without prompt treatment. We present a rare case of extensive thromboembolism due to PDE with an incidental finding of inferior sinus venosus ASD in an elderly.

CASE PRESENTATION

A 67-year-old female presented with a history of left upper limb numbness and coldness for five days associated with left upper limb weakness.

She denied any history of trauma, recent surgery, fever or seizures. No chest pain or shortness of breath. Physical examination of the left upper limb showed no skin discoloration but absence of left brachial, radial and ulnar arteries pulsation. No signal gained on arterial Doppler examination, which thus supported the diagnosis of acute left upper limb ischaemia. Emergency embolectomy was performed followed by heparin infusion. Upon discharge, she was well with good left upper limb perfusion. She was prescribed oral Cardiprin 100 mg daily for a month.

She presented again 25 days later with sudden onset bilateral upper limbs and right lower limb pain, worse on the right lower limb associated with weakness. Clinically, she had tachypnoea with tachycardia. Her right lower limb appeared dusky and cold on palpation. The distal arteries of her right lower limb were non-palpable. She was diagnosed as having recurrent acute limb ischaemia. Due to persistent tachycardia and a Wells score that indicated high probability of pulmonary embolism (PE), an urgent computed tomography pulmonary angiography (CTPA) and limb CT angiography (CTA) were performed. Massive PE was detected with saddle embolus in the pulmonary trunk bifurcation. Pulmonary artery was dilated but no evidence of pruning. However, there was an incidental finding of a defect at the inferior part of interatrial septum consistent with inferior sinus venosus ASD. The right heart chamber size appeared mildly prominent. The interventricular septum was intact but flattened and appeared to be in systolic phase suggestive of increased right heart pressure. There was no significant evidence of right ventricular wall hypertrophy (Figure 1 and 2). The pulmonary veins demonstrated normal drainage into the left atrium. Other cardiovascular structures were also unremarkable.

Limbs CTA showed extensive thrombus within bilateral upper limbs and right lower limb arteries. In the right upper limb, complete occlusion of the subclavian, axillary, radial and ulnar arteries was seen with partial occlusion of the right brachial artery. Long segment thrombus was also seen in the left subclavian, axillary, and brachial arteries (Figure 3). Complete occlusion of the right lower limb arteries was noted involving

the entire length of the right common and superficial femoral arteries (Figure 4).

Transthoracic echocardiography (TTE) examination revealed normal atrial and ventricular size. There was interventricular septal hypertrophy and the interatrial septum 'appeared' intact. No obvious ASD was identified.

Disseminated intravascular coagulopathy screening showed normal fibrinogen level at 2.4g/L (normal values: 2.32 – 4.44g/L), which made coagulopathy a less likely cause of the extensive thromboembolic disease.

She received alteplase infusion and underwent open mechanical thrombectomy of right lower limb. Unfortunately, she became markedly anaemic with melena during thrombolytic treatment. Mesenteric CTA confirmed the presence of active lower gastrointestinal bleed. The bleeding artery was successfully embolized through transcatheter approach. However, her condition deteriorated further during hospitalization due to multiorgan failure and worsening septicaemia. She succumbed to the condition after ten days of hospitalization.

DISCUSSION

PDE is a rare phenomenon especially in combination with inferior sinus venosus ASD. To the best of our knowledge, this is the first case of an inferior sinus venosus ASD causing extensive PDE affecting the extremity arteries and pulmonary artery. Inferior sinus venosus ASD is a defect below the atrial septum which leads to an overriding inferior vena cava (IVC) and interatrial connection. Previous case reports have described the association of PDE with patent foramen ovale (PFO). One of the cases was found in an adult patient with bilateral deep vein thrombosis associated with recent air travel that later developed massive PE and acute left upper and lower limbs ischaemia. The subsequent investigation with contrast-enhanced transoesophageal echocardiography (TOE) revealed right-to-left shunt through channel-like interatrial communication consistent with PFO.⁶ A case series described two patients with arteriovenous thromboembolism due to PDE. In the series, one patient had concurrent episodes of acute cerebral infarction and PE in the presence of

an ASD, complicated with atrial septal aneurysm and a left-to-right shunt. The other patient had PFO associated with upper extremity artery embolism, venous thromboembolism and PE.⁷

Concomitant limb ischaemia and PE with indeterminate aetiology should alert the treating physician the possibility of arteriovenous shunt and PDE as depicted in our case. Early diagnosis is crucial due to its high mortality risk if left undetected, particularly among older generations. Nonetheless, inferior sinus venosus ASD is extremely rare. Its posterior location also makes the diagnosis through routine TTE a great challenge, hence missed as proven in our case. In view of the proximity between the sinus venosus defects and the transducer, guidelines from the American Heart Association suggested TOE to diagnose this anomaly.⁸ However, due to the lack of early clinical suspicion of intracardiac shunt, presence of technical issues with the apparatus and deteriorating clinical condition, TOE was not performed in our patient.

The inferior sinus venosus defect was incidentally discovered in our CTPA examination, prompting the scrutiny of other possible associated cardiovascular anomalies particularly the anomalous right inferior pulmonary venous return to the right atrium. Despite the limited functional information in CTPA, careful evaluation of the cardiovascular structures still able to provide useful indirect information particularly related to complication of longstanding uncorrected shunt as we have described above. Although this type of defect can

cause volume overload and is less likely to result in Eisenmenger syndrome due to its low pressure, our patient had some features which suggested the presence of pulmonary arterial hypertension, likely related to the extensive PE. For this reason, TTE is able to compare the chamber size and function; measure the ventricular wall thickness; measure the flow velocity across the tricuspid and pulmonary valves; assess the morphological and dynamic changes of interventricular septum and IVC. The more invasive TOE technique allows better morphological and flow assessment of sinus venosus defect. Having the advantage of unrestricted field of view and non-invasive approach, magnetic resonance imaging is able to accurately demonstrate the location and size of the defect. Using the velocity encoding gradient, it also can accurately quantify and compare the flow volume of the right and left heart chambers, hence grade the severity of the shunt.

CONCLUSION

PDE secondary to intracardiac shunt should be considered in differential diagnosis for patients with concomitant limb ischaemia and PE of unknown aetiology since it can potentially lead to extensive thromboembolism. The high mortality rate can be secondary to disease severity or complications of treatment. CTA is the alternative imaging modality to detect sinus venosus ASD provided there is sufficient knowledge of congenital cardiovascular disease and its complication.

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Author Contributions Statement:

ZKAI contributed to writing the manuscript.

KAS contributed to case selection, proofreading the manuscript and providing expert opinion.

MSA supervised the overall manuscript preparation.

Consent to Participate:

Verbal consent was taken from the next of kin of the patient described in the case report since the patient died due to the illness. Ethical board approval for submission of this case report was obtained from Research Ethical Committee of Hospital Universiti Sains Malaysia

FIGURE LEGENDS

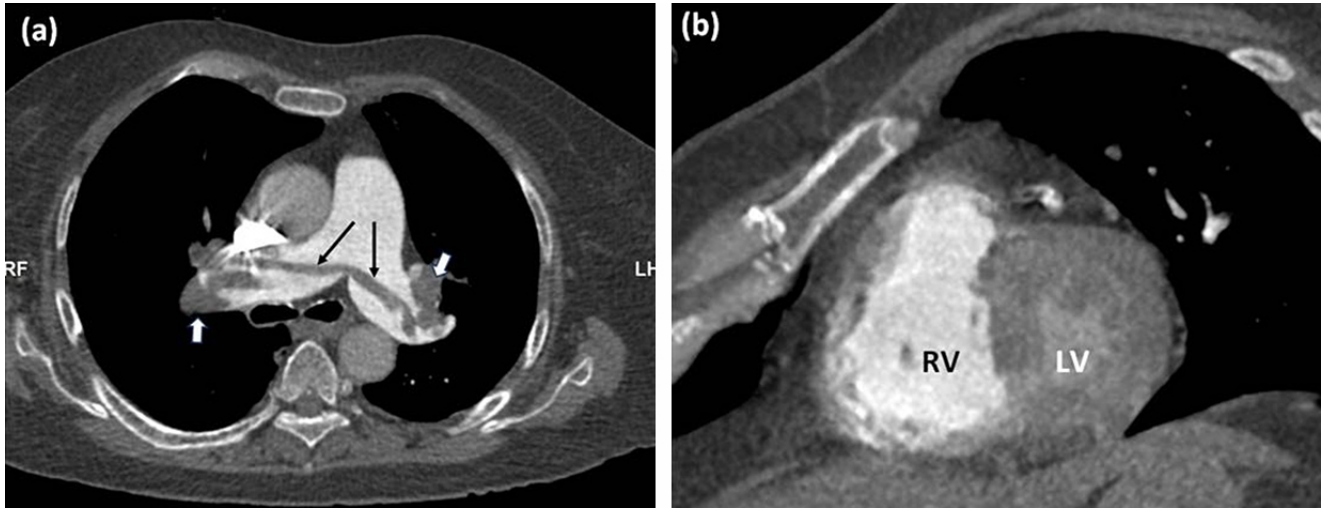


Figure 1: (a) Axial CT pulmonary angiography at the level of pulmonary artery bifurcation demonstrates dilated pulmonary trunk with presence of saddle embolus extending within both main pulmonary arteries (thin arrows). Large emboli are also seen within both segmental branches of pulmonary arteries (thick arrows). (b) There is straightening of interventricular septum indicating presence of pulmonary arterial hypertension.

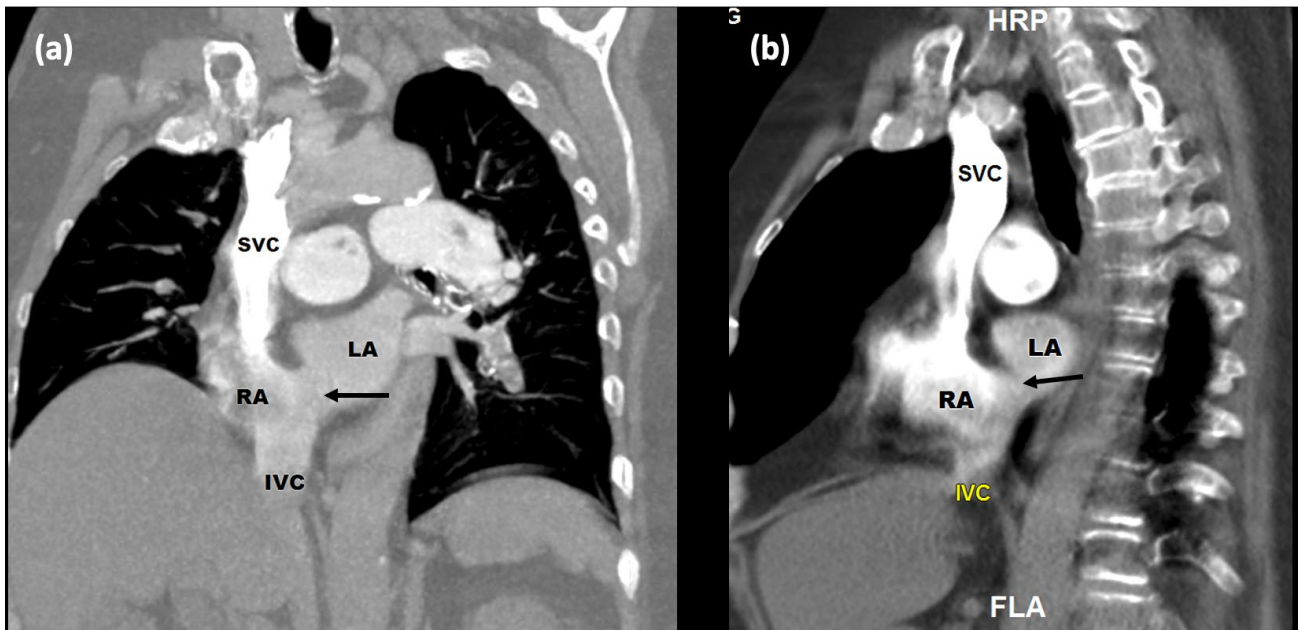


Figure 2: CT pulmonary angiography images in (a) oblique coronal and (b) oblique sagittal planes demonstrate the inferior sinus venosus defect (black arrow). Note the opacification of the left heart chambers during pulmonary arterial phase indicating interatrial right-to-left shunt. The IVC nearly overrides the defect, a characteristic feature of this anomaly, best illustrated in (a).

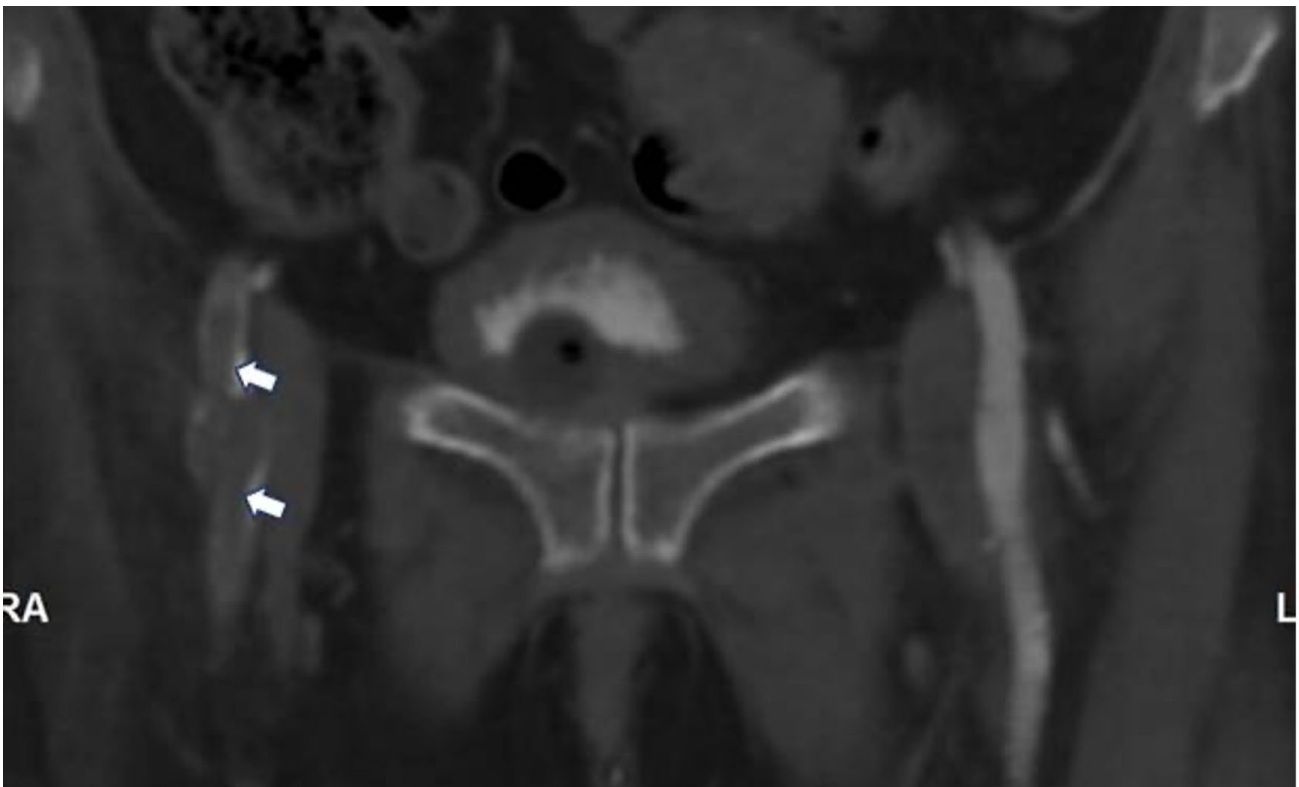
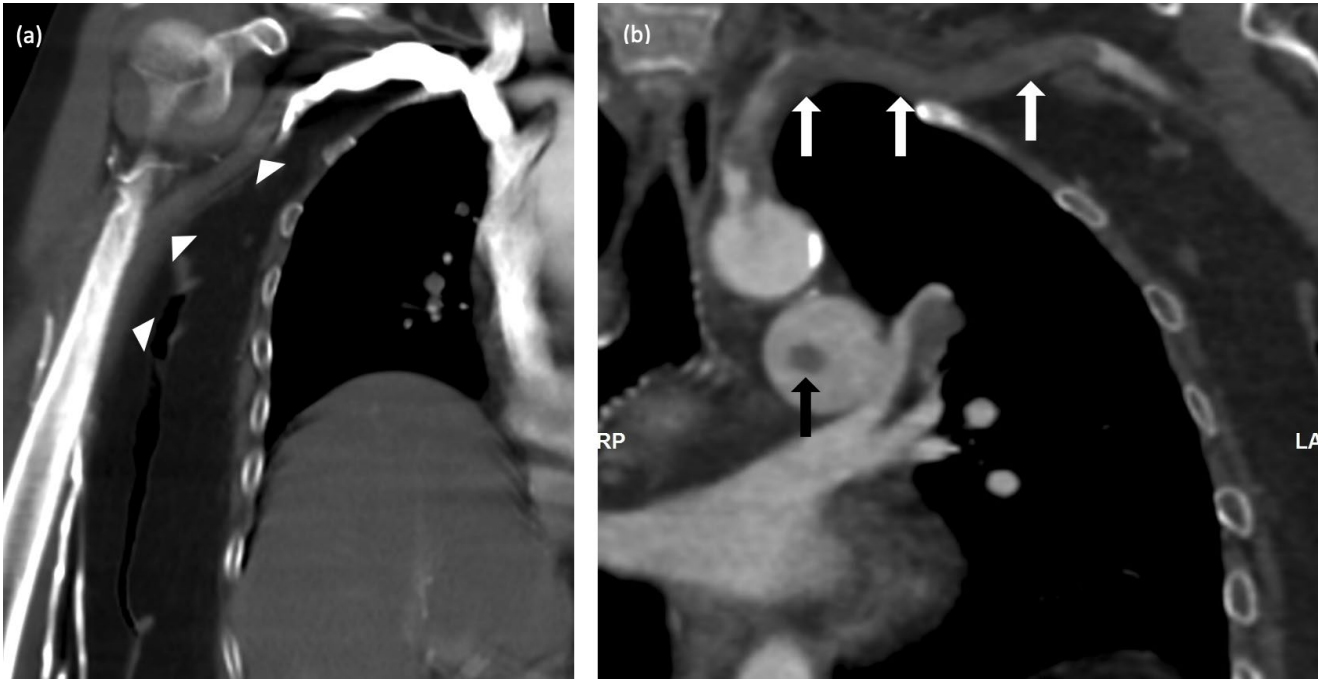


Figure 4: Coronal reformatted CT angiography image of the lower limbs demonstrates long segment filling defect within the right common and superficial femoral artery (arrows). The left femoral artery is well-opacified.

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