

CASE REPORT

An atypical case of pulmonary embolism from a jugular vein

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Summary

Neck venous malformations and their potentially life-threatening complications are rarely reported in the available literature. Cases of aneurysmal or hypo-plastic jugular vein thrombosis associated with systemic embolization have not been frequently reported. We present the case of a 60-year-old male, without any known risk factors for thromboembolic disease, admitted for sudden onset dyspnea. The physical examination was remarkable for a right lateral cervical mass, expanding with Valsalva maneuver. Thoracic CT with contrast established the diagnosis of bilateral pulmonary embolism and raised the suspicion of superior vena cava and right atrial thrombosis. Bedside transthoracic echocardiography confirmed the presence of a large right atrial thrombus, with intermittent protrusion through the tricuspid valve. Systemic thrombolysis with Alteplase was initiated shortly after diagnosis, in parallel with unfractionated heparin, with complete resolution of the intracavitory thrombus documented by echocardiography. The patient showed significant improvement in symptoms and was later started on oral anticoagulation. Computed vascular tomography of the neck was performed before discharge, showing hypoplasia of the left internal jugular vein and aneurysmal dilation of the contralateral internal jugular vein, without thrombosis. There were no identifiable systemic causes for thrombosis. Surgical resection of the aneurysmal jugular vein was excluded, because of its potential to cause intracranial hypertension. The preferred therapeutic option in this case was long-term oral anticoagulation.

Key Words

- ▶ jugular vein aneurysm
- ▶ acute pulmonary embolism
- ▶ jugular vein hypoplasia
- ▶ three-dimensional computed angiography

Learning points:

- Internal jugular venous malformations, such as aneurisms or hypoplasia, could be associated with an increased risk of thrombosis and major embolic events.
- Systemic thrombolysis can be an efficient solution in cases of pulmonary embolism with right heart thrombosis.
- Multimodality imaging is greatly valuable in clarifying the diagnosis of atypical cases.

Background

Venous aneurisms of the neck are rare entities, typically asymptomatic, potentially complicated by thrombosis or rupture (1, 2). Venous malformations of the neck associated with thromboembolic events are rarely reported in the available literature (3, 4).

We present the case of a patient with congenital hypoplasia of the left internal jugular vein, with a compensatory contralateral jugular vein aneurysm, admitted for pulmonary embolism with massive right heart thrombosis.

Case presentation

A 60-year-old male presented to the emergency department with acute onset of dyspnea and presyncope. The patient did not have any known risk factors for thromboembolic disease or history of thromboembolic events. On examination, the blood pressure was 110/60 mmHg, the pulse 120 beats per minute, the respiratory rate 22 breaths per minute and the oxygen saturation 95% while breathing in ambient air. Facial asymmetry and a right lateral cervical mass (3 cm in diameter), expanding with Valsalva maneuver was noted, the remainder of the examination was normal. There were no signs of deep venous thrombosis of the lower extremities.

Investigation

The electrocardiogram showed sinus tachycardia (120 beats per minute) and an incomplete right bundle branch block. The laboratory tests were remarkable for increased levels of serum NT-proBNP (1766 ng/dL) and Troponin I (0.055 ng/mL). Conversely, the blood cell count, hepatic and renal functions were normal.

The patient had a high clinical suspicion of pulmonary embolism. Emergency thoracic CT with contrast was performed, revealing bilateral pulmonary embolism (Fig. 1), as well as superior vena cava and right atrial thrombosis (Fig. 2, panels A and B). Bedside transthoracic echocardiography confirmed the presence of a massive right atrial thrombus, extending from the superior vena cava (Fig. 3, panels A, B, and C). The thrombus was highly mobile, hyper segmented and partially protrusive through the tricuspid valve into the right ventricle (Video 1). Additionally, echocardiography showed a positive McConnell sign, decreased tricuspid annular

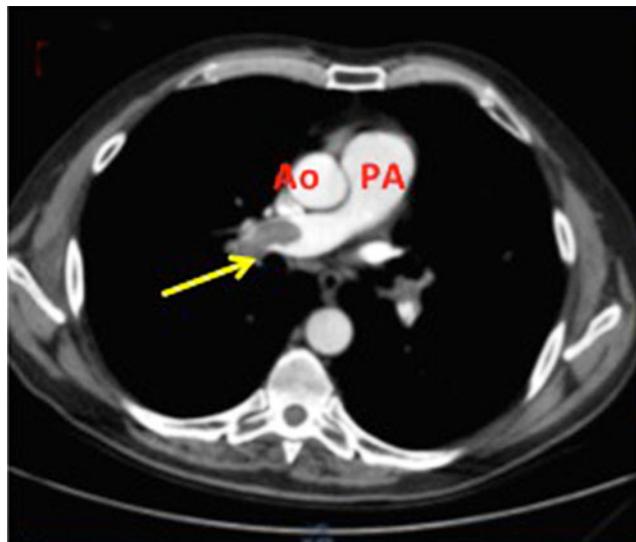


Figure 1

Thoracic computed tomography showing thrombi in the principal pulmonary arteries (yellow arrow). Ao, aorta; PA, pulmonary artery.

plane systolic excursion (11 mm) and a high velocity of the tricuspid regurgitant jet (4.2 m/s).

Video 1

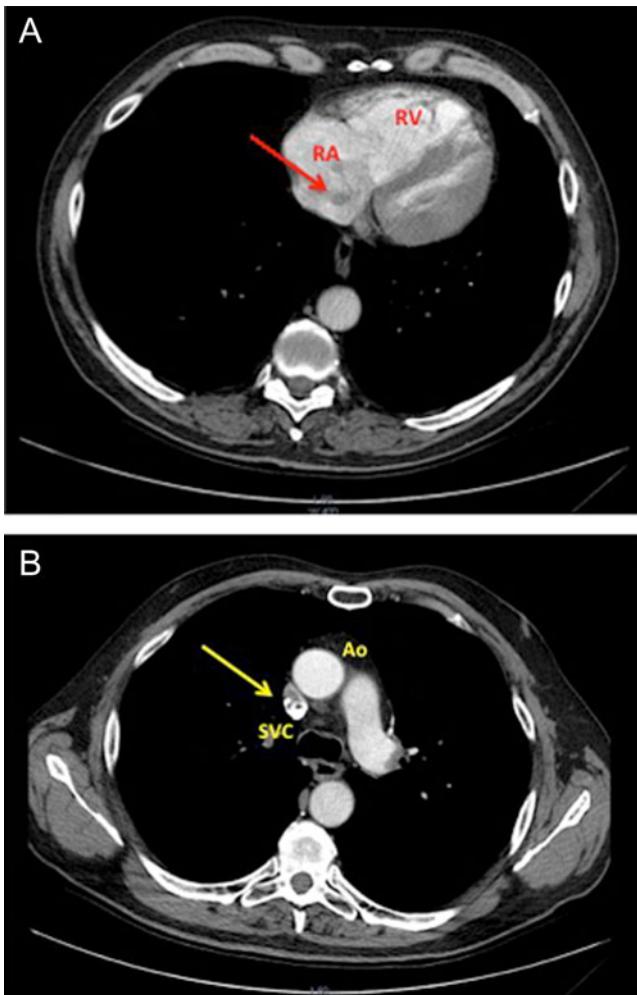
Thrombus of large dimensions, highly mobile, is crossing the tricuspid valve into the right ventricle. View Video 1 at <http://movie-usa.glencoesoftware.com/video/10.1530/ERP-18-0029/video-1>.

Extensive workup was negative for hematological disease associated with thrombosis, malignancy, systemic autoimmune disease or thrombophilia. Venous Duplex ultrasound of the lower limbs showed no signs of thrombosis. Considering the presence of the lateral cervical mass and the superior vena cava thrombosis, we further investigated the cervical region. Venous Doppler of the cervical veins, performed 3 days after the acute event, revealed aneurismal dilation of the right internal jugular vein, without signs of residual thrombosis. Furthermore, cervical CT angiography showed an unusual anatomy of the neck veins: hypoplasia of the left internal jugular vein and aneurismal saccular dilation of the right internal jugular vein, with no signs of thrombosis (Figs 4 and 5).

Treatment and outcome

Systemic thrombolytic therapy with alteplase was administered (100 mg in 2 h), in parallel with a continuous infusion of unfractionated heparin (adjusted according to



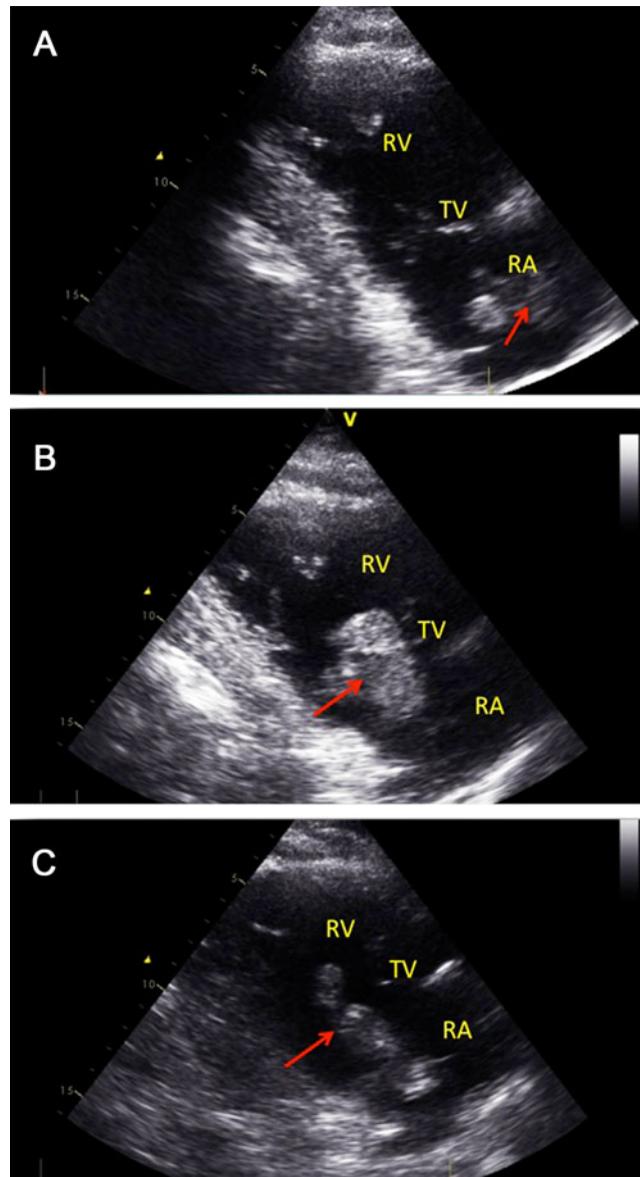
**Figure 2**

Two-dimensional chest tomography with contrast. Thrombus inside the right atrium (panel A, red arrow) and at the level of the superior vena cava (panel B, yellow arrow) can be visualized.

the activated partial thromboplastin time). The patient was later switched to low-molecular-weight heparin and oral anticoagulation with a vitamin K antagonist was initiated. There was a significant improvement in symptoms and no bleeding complications were noted. Echocardiography revealed complete resolution of the right atrial thrombus 3 h post thrombolysis, as well as an improvement in the systolic function of the right ventricle and a decrease in the pulmonary artery pressure. Three-dimensional echocardiography performed 24 h later showed dilated right heart chambers, but without signs of thrombosis (Fig. 6, panels A, B and Video 2)

Video 2

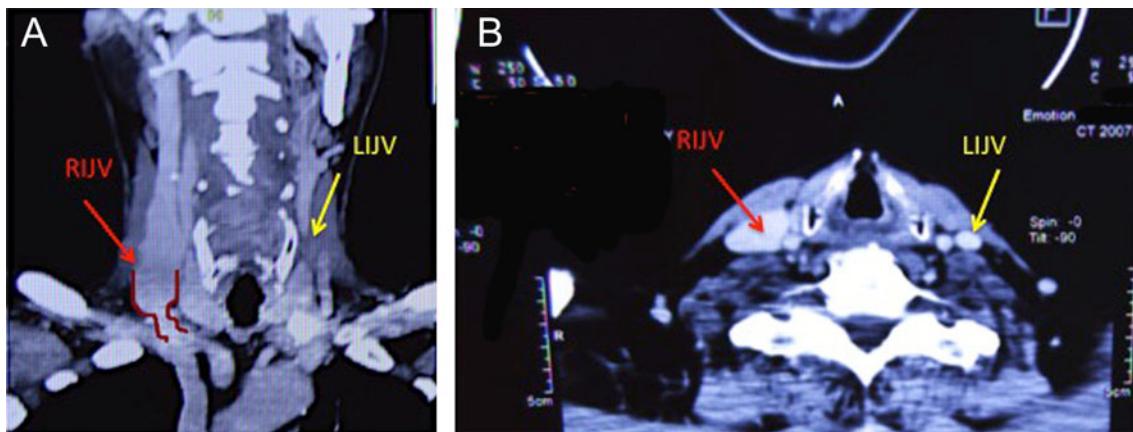
Three-dimensional echocardiography of the right heart chambers, 24 h post-thrombolysis. The video reveals

**Figure 3**

Two-dimensional echocardiography showing the parasternal long axis view of the right ventricle, tricuspid valve and right atrium. Large, hyper segmented right atrial thrombus (red arrow), protruding through the tricuspid valve (panel A). The thrombus is seen in the right atrium during the cardiac systole, when the tricuspid valve is closed. (Panel B) Opening of the tricuspid valve, showing the protrusion of the thrombus into the right ventricle. (Panel C) The fully extended thrombus into the right heart chambers. RA, right atrium; RV, right ventricle; TV, tricuspid valve.

dilated right atria and right ventricle, the opening and closing of a normal tricuspid valve and the inferior and superior vena cava. There are no signs of intra-cardiac thrombus 24 h post thrombolysis. View Video 2 at <http://movie-usa.glencoesoftware.com/video/10.1530/ERP-18-0029/video-2>.



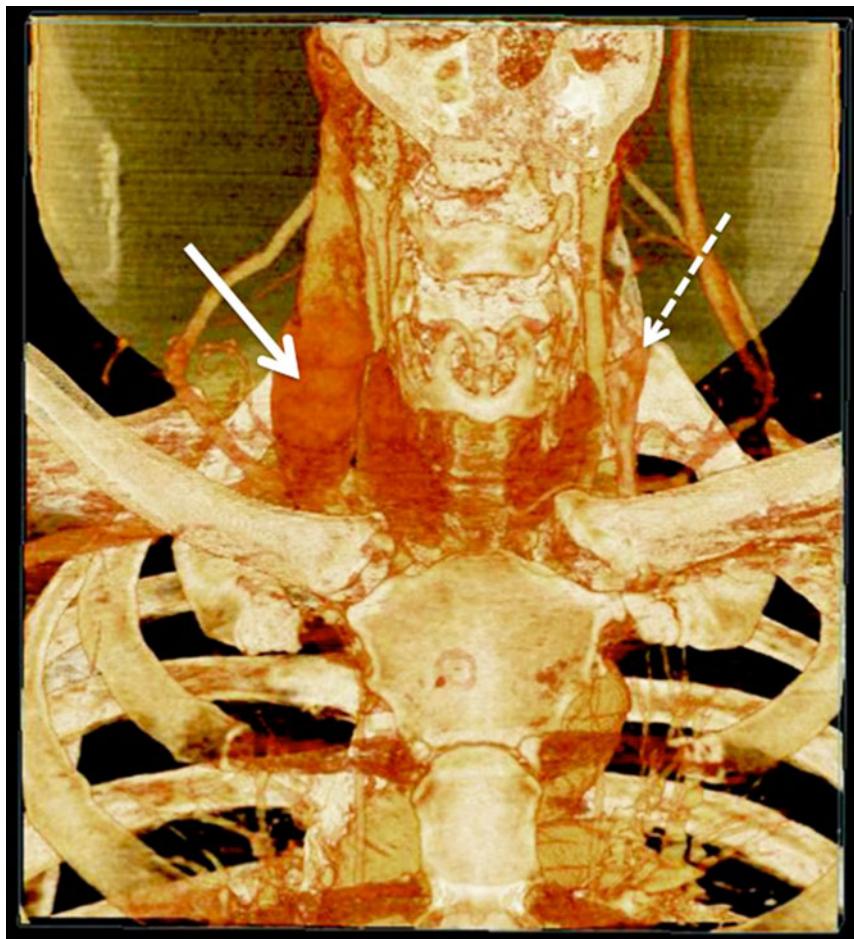
**Figure 4**

Two-dimensional tomographic images of the neck, in longitudinal (A) and transversal sections (B). The right internal jugular vein aneurysm (red arrows) and left internal jugular vein hypoplasia (yellow arrows) can be seen on both sections, using venous contrast. LIJV, left internal jugular vein hypoplasia; RIJV, right internal jugular vein aneurysm.

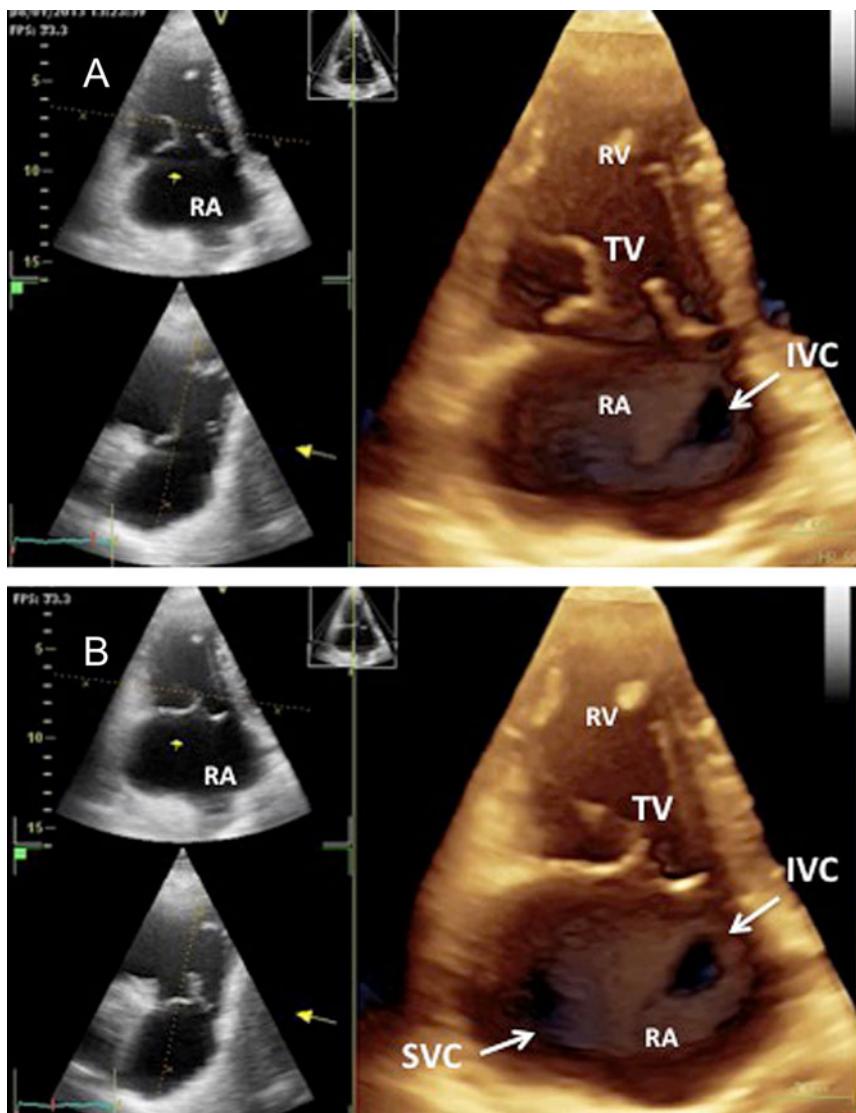
Discussion

Neck venous malformations are rare entities, which usually remain asymptomatic (1). Isolated internal jugular vein aneurysms are the most frequently encountered

anomalies of the venous system of the neck, and internal jugular vein agenesis and hypoplasia, associated with compensatory aneurysmal dilation of the contralateral venous axis have been also reported (1). Incidental thrombosis of a hypoplastic (5) or of an aneurysmal jugular

**Figure 5**

Three-dimensional computed angiography of the neck veins and arteries. Right internal jugular vein aneurysm (arrow) and left internal jugular vein hypoplasia (dotted line).

**Figure 6**

Transthoracic three-dimensional echocardiography of the right heart chambers after thrombolysis. No evidence of thrombus to the right atrium (RA) or the right ventricle (RV). IVC, inferior vena cava; SVC, superior vena cava; TV, tricuspid valve.

vein (6) have been previously described in patients with risk factors for thrombosis, generally without embolic complications. Usually, thromboembolic events are caused by aneurysms in the lower limb veins (popliteal, femoral), and a single case of a cephalic vein with secondary pulmonary embolism was previously reported (7).

Our case illustrates the potential of life-threatening thromboembolism associated with complex neck venous malformations, in a patient with no known risk factors for thrombosis. To the best of our knowledge, thrombosis and embolism from a congenital venous aneurism has not been previously reported.

The classic approach to venous aneurisms of the neck is surgical resection, especially if symptomatic (8). In our patient, the saccular dilation of the right internal jugular vein was interpreted as compensatory for the left venous axis hypoplasia. Therefore, such an approach was not

feasible, as it would have potentially caused intracranial hypertension. Data are scarce about the interventional or surgical treatment of symptomatic venous aneurisms occurring secondary to contralateral hypoplasia or agenesis. Therefore, long-term oral anticoagulation was the preferred treatment option for the prevention of further thromboembolic events in this patient.

Internal jugular venous malformations, such as aneurisms or hypoplasia, might be associated with increased risk of thrombosis and major embolic events, making their recognition critical for the institution of appropriate therapy.

Our case is the first to report a severe thromboembolic event from a jugular vein aneurysm, which was not spontaneous, but caused by the hypoplasia of the contralateral vein. A specific treatment plan was necessary in our patient, relatively different from the ones reported by the literature.



Declaration of interest

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this case report.

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Patient consent

Written informed consent has been obtained from the patient.

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