Research Square

Preprints are preliminary reports that have not undergone peer review.
They should not be considered conclusive, used to inform clinical practice,
or referenced by the media as validated information.

Cerebral infarction caused by an ascending aortic thrombus associated with systemic capillary leak syndrome: a case report

Yuki Kuranari

yuki_kuranari@keio.jp

Hiratsuka City Hospital

Shogo Yoshida

Hiratsuka City Hospital

Yoshiaki Kuroshima

Hiratsuka City Hospital

Akiyoshi Nakamura

Hiratsuka City Hospital

Yoichi Furuya

Hiratsuka City Hospital

Case Report

Keywords: Systemic capillary leak syndrome, Cerebral infarction, Ascending aortic thrombus

Posted Date: August 20th, 2026

DOI: https://doi.org/10.21203/rs.3.rs-10566382/v1

License: This work is licensed under a Creative Commons Attribution 4.0 International License.
Read Full License

Additional Declarations: No competing interests reported.

Version of Record: A version of this preprint was published at BMC Neurology on October 8th, 2026. See the published version at https://doi.org/10.1186/s12883-026-05467-3.

Abstract

Background

Systemic capillary leak syndrome (SCLS) is a rare disorder characterised by recurrent episodes of hypovolemic shock, haemoconcentration, and hypoalbuminaemia, caused by transient increases in vascular permeability. Although venous thromboembolic complications have been reported in patients with SCLS, arterial thrombosis involving the ascending aorta is rare. Herein, we report a case of cerebral infarction caused by an ascending aortic thrombus associated with SCLS and discuss the possible pathophysiological mechanisms.

Case presentation

A 62-year-old man presented with generalised oedema, oliguria, severe hypotension, marked haemoconcentration (haematocrit, 70%), acute kidney injury, and hypoalbuminaemia. Based on the recurrent episodes of hypotension, haemoconcentration, hypoalbuminaemia, and the presence of serum IgG-kappa monoclonal protein, the patient was diagnosed with SCLS. During the acute leak phase, the patient developed compartment syndrome requiring a decompressive fasciotomy. Two days after admission, he suddenly developed aphasia and right hemiplegia while eating breakfast. Magnetic resonance imaging (MRI) of the brain revealed an acute cerebral infarction due to occlusion of the left internal carotid artery. Endovascular thrombectomy achieved successful reperfusion (Thrombolysis in Cerebral Infarction grade 2b). Contrast-enhanced computed tomography angiography revealed a mural thrombus in the ascending aorta without evidence of an aneurysm, dissection, or significant atherosclerosis. Anticoagulation therapy with heparin, followed by warfarin, resulted in complete resolution of the thrombus. Histopathological examination revealed a fibrin-rich thrombus with neutrophil and lymphocyte infiltration, compatible with an early stage thrombus.

Conclusions

This case suggests that severe haemoconcentration, abnormal blood flow, and endothelial dysfunction during the acute phase of SCLS may synergistically promote de novo ascending aortic thrombus formation, resulting in cerebral embolism. Comprehensive vascular imaging should be considered to identify potential arterial embolic sources in patients with SCLS who develop acute neurological deficits. Further accumulation of similar cases is required to clarify the mechanisms and optimal management of arterial thrombotic complications in SCLS.

Background

Systemic capillary leak syndrome (SCLS) is a rare and potentially life-threatening disorder characterised by recurrent episodes of hypovolemic shock, haemoconcentration, and hypoalbuminemia [1]. SCLS is

associated with a broad spectrum of systemic complications, including acute renal failure, pulmonary oedema, rhabdomyolysis, and compartment syndrome [1]. In addition, severe haemoconcentration and endothelial dysfunction may predispose patients to thrombotic complications. Cases of deep venous thrombosis in patients with SCLS have been reported [1, 2]; however, aortic thrombus formation in patients with SCLS is exceedingly rare. Ascending aortic thrombi can cause life-threatening systemic embolization. Herein, we report a case of cerebral infarction due to an ascending aortic thrombus in a patient with SCLS and discuss the possible pathophysiological mechanisms and clinical implications.

Case presentation

A 62-year-old man presented to the emergency department with generalized oedema and decreased urine output for three days. Upon hospital arrival, his blood pressure was 70/36 mmHg and pulse was 110 beats/min. Physical examination revealed non-pitting pedema of the upper and lower extremities. Initial blood tests showed a haematocrit of 70% (normal 40–50%), creatinine of 2.72 mg/dL (normal 0.36–1.06 mg/dL), and hypoalbuminemia with a serum albumin level of 2.8 g/dL (normal 3.8–5.3 g/dL). He had no fever, and there was a non-specific increase in C-reactive protein to 0.67 mg/dL (normal < 0.30 mg/dL). Cardiac markers, including creatine kinase-MB and troponin-T, were within the normal ranges. Transthoracic echocardiography revealed preserved left ventricular systolic function.

Two years previously, he had experienced similar symptoms after receiving the third dose of the coronavirus disease 2019 vaccine. Since then, similar episodes have recurred every three months. Five months before presentation, he presented to the emergency department with general fatigue. On admission, he had hypotension, and blood tests revealed severe haemoconcentration with a haematocrit of 69.9%, a creatinine of 3.45 mg/dL, and hypoalbuminaemia with a serum albumin level of 2.8 g/dL. He was diagnosed with prerenal acute kidney injury and was treated with intravenous fluids. The patient's symptoms rapidly improved after intravenous fluid therapy.

A diagnosis of SCLS was established based on recurrent episodes of hypotension, haemoconcentration, and hypoalbuminaemia after excluding other causes of shock, including septic and cardiogenic aetiologies, and was supported by the detection of serum immunoglobulin G-kappa monoclonal protein.

Sixteen hours after admission, the patient developed compartment syndrome in the right lower leg, for which decompressive fasciotomy was performed. During the first 24 hours, he was treated with 8 L of intravenous fluid and norepinephrine to maintain his blood pressure. Two days after admission, he suddenly developed aphasia and right hemiplegia while eating breakfast in the hospital ward. Urgent brain magnetic resonance imaging revealed acute cerebral infarction due to occlusion of the left internal carotid artery (Fig. 1A, B). Endovascular thrombectomy resulted in successful reperfusion (Thrombolysis in Cerebral Infarction, grade 2b). The thrombus obtained during endovascular thrombectomy is composed of fibrin, neutrophils, and lymphocytes. Contrast-enhanced computed tomography angiography (CTA) was performed to investigate the source of the embolism. CTA revealed a filling defect in the ascending aorta without evidence of an aortic dissection or aneurysm (Fig. 2A, B).

Anticoagulant therapy with heparin was initiated, followed by a transition to warfarin (controlled within a 2.0–3.0 international normalized ratio). Hypercoagulability screening revealed no specific findings. No recurrent embolic events occurred, and the filling defect disappeared on repeat CTA after two weeks (Fig. 3A, B). The patient was transferred to a rehabilitation hospital 30 days after admission.

Discussion

The present case illustrates a potentially life-threatening cause of cerebral infarction due to an ascending aortic thrombus in a patient with SCLS and highlights the importance of recognising the ascending aorta as a potential embolic source in this population. Venous thromboembolic complications have been reported in patients with SCLS. In the Mayo Clinic series, deep venous thrombosis occurred in approximately 8% of patients with SCLS [2]. Although Virchow's triad (abnormal blood flow, hypercoagulability, and endothelial injury) has traditionally been applied to venous thrombosis, it is also a useful conceptual framework for understanding arterial thrombus formation [3].

Accordingly, the development of an ascending aortic thrombus in this patient may be explained within the framework of Virchow's triad.

First, profound hypovolemic shock likely contributes to systemic hypoperfusion and relative circulatory stasis. The patient presented with severe hypotension requiring norepinephrine support, which may have resulted in reduced systemic blood flow and relative circulatory stasis, predisposing him to in situ thrombus formation. Second, marked haemoconcentration was evident with a haematocrit of 70% on admission. Such a marked haemoconcentration is expected to increase blood viscosity, thereby promoting a hypercoagulable state. Third, endothelial dysfunction is considered central to the pathogenesis of SCLS. Increased vascular permeability and endothelial activation during acute episodes may disrupt the endothelial integrity and create a prothrombotic environment. Collectively, these factors may have acted synergistically to promote thrombus formation in the ascending aorta during the acute leak phase. Although thrombus formation likely occurred during the acute leak phase, cerebral embolization developed two days after admission, indicating that thrombus formation and embolization were temporally distinct events. Overall, these findings support the hypothesis that pathophysiological changes associated with acute SCLS contribute to de novo ascending aortic thrombus formation, which subsequently results in cerebral embolization.

Ascending aortic thrombi in the absence of structural abnormalities such as aneurysms or dissections are uncommon [4, 5]. When present, it is often associated with procoagulant abnormalities, malignancy, or steroid therapy [6]. In our patient, contrast-enhanced computed tomography angiography revealed a filling defect in the ascending aorta without evidence of atherosclerotic plaques, aneurysms, or dissection, suggesting de novo thrombus formation. Hypercoagulability screening revealed no inherited or acquired thrombophilia. Furthermore, cardiac evaluation showed preserved left ventricular systolic function, and no overt cardiac source of embolism was identified. The temporal association between acute SCLS episodes, extreme haemoconcentration, and subsequent cerebral infarction supports the

hypothesis that SCLS-related pathophysiological changes contribute to thrombus formation and embolization.

The retrieved thrombus consisted predominantly of fibrin with infiltration of neutrophils and lymphocytes, findings compatible with an early stage thrombus, as previously described in histopathological studies on thrombus maturation [7]. Although detailed immunohistochemical analyses were not performed, these histopathological features further supported the plausibility of de novo thrombus formation during the acute leak phase.

From a clinical perspective, this case underscores the importance of a comprehensive embolic source evaluation in patients with SCLS who develop acute focal deficits. Although a cardiac source of embolism is commonly suspected in such patients, atypical arterial sources, including the ascending aorta, should not be overlooked. Early vascular imaging such as computed tomography angiography may facilitate the prompt identification of treatable embolic lesions. In our patient, timely endovascular thrombectomy resulted in recanalization of the occluded internal carotid artery, and subsequent anticoagulation therapy was associated with complete resolution of the aortic thrombus on follow-up imaging. Although the optimal antithrombotic strategy for SCLS remains undefined, this case suggests that anticoagulation may represent a reasonable therapeutic option after an arterial thrombus is identified, provided that the bleeding risk is carefully assessed.

The present report has several limitations. First, as this was a single case, a definitive causal relationship between SCLS and ascending aortic thrombus formation could not be established. Secondly, although major alternative embolic sources were not identified, occult cardioembolic sources and other undetected prothrombotic conditions could not be completely excluded. Third, we did not directly measure the blood viscosity or endothelial biomarkers, which may have provided additional mechanistic insights. Therefore, the proposed mechanism should be regarded as hypothesis generating rather than conclusive.

Conclusions

This case report suggests that severe haemoconcentration and endothelial dysfunction during an acute episode of SCLS may contribute to de novo ascending aortic thrombus formation, leading to cerebral embolism. Clinicians should be aware of this potential complication and consider comprehensive vascular imaging in patients with SCLS presenting with acute neurological symptoms. Further accumulation of similar cases is necessary to clarify the true incidence, mechanisms, and optimal management of arterial thrombotic complications in patients with SCLS.

Abbreviations

SCLS Systemic capillary leak syndrome

CTA Computed tomography angiography

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent for publication of this case report and any accompanying images was obtained from the patient's wife, as the patient lacked the capacity to provide consent owing to persistent neurological deficits.

Availability of data and materials

The data supporting the findings of this study are available from the corresponding author upon request.

Competing interests

The authors declare no competing interests.

Funding

Not applicable.

Authors' contributions

YKuranari drafted the manuscript. YS, YKuroshima, and AN critically revised the manuscript. YF conceived the study, supervised the project, and critically revised the manuscript. All authors read and approved the final manuscript.

Acknowledgements

The authors thank the patient and his family for providing consent for the publication of this case report.

References

1. Druey KM. Narrative Review: Clarkson Disease-Systemic Capillary Leak Syndrome. Published online 2011.
2. Kapoor P, Greipp PT, Schaefer EW, Mandrekar SJ, Kamal AH, Gonzalez-Paz NC, et al. Idiopathic systemic capillary leak syndrome (Clarkson's disease): The Mayo clinic experience. Mayo Clin Proc. 2010;85:905-12.
3. Wolberg AS, Aleman MM, Leiderman K, Machlus KR. Procoagulant activity in hemostasis and thrombosis: Virchow's triad revisited. Anesth Analg. 2012;114:275-85.

4. Machleder HI, Takiff H, Lois JF, Holburt E. Aortic mural thrombus: An occult source of arterial thromboembolism. J Vasc Surg. 1986;4:473-8.
5. Laperche T, Laurian C, Roudaut R, Steg PG. Mobile thromboses of the aortic arch without aortic debris. A transesophageal echocardiographic finding associated with unexplained arterial embolism. The Filiale Echocardiographie de la Société Française de Cardiologie. Circulation. 1997;96:288-94.
6. Weiss S, Bühlmann R, von Allmen RS, Makaloski V, Carrel TP, Schmidli J, et al. Management of floating thrombus in the aortic arch. J Thorac Cardiovasc Surg. 2016;152:810-7.
7. Bacigaluppi M, Semerano A, Gullotta GS, Strambo D. Insights from thrombi retrieved in stroke due to large vessel occlusion. J Cereb Blood Flow Metab. 2019;39:1433-51.

Figures

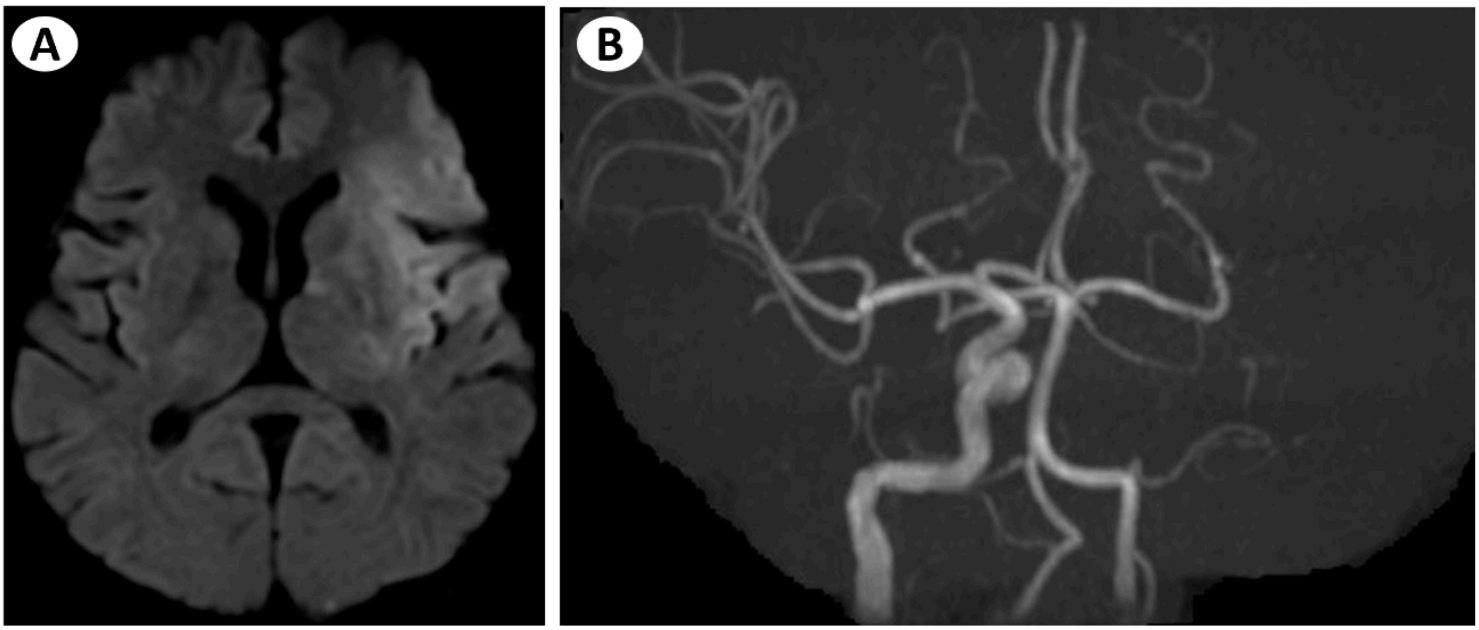


Figure 1

Brain magnetic resonance imaging obtained after the onset of neurological symptoms.

(A) Diffusion-weighted imaging demonstrates acute ischemic infarction.

(B) Magnetic resonance angiography demonstrates occlusion of the left internal carotid artery.

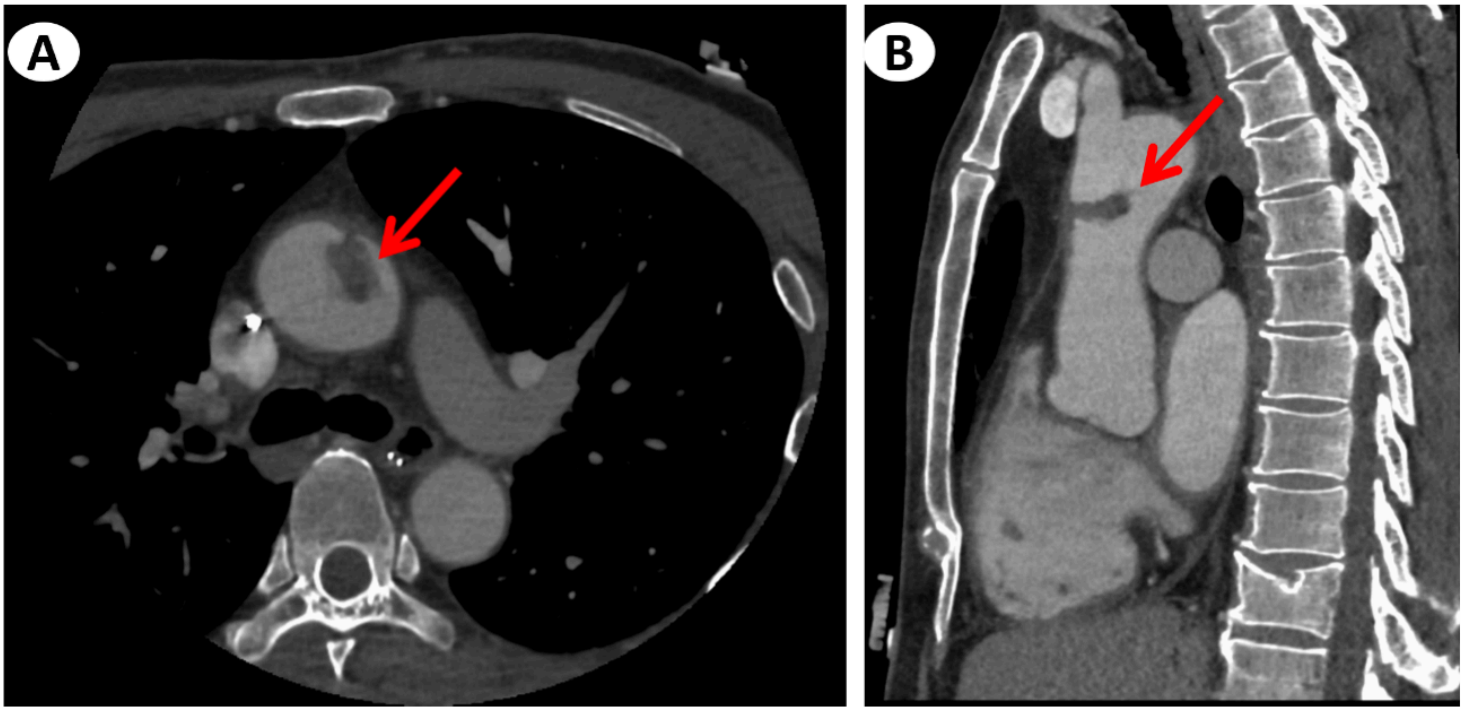


Figure 2

Contrast-enhanced computed tomography angiography (CTA) demonstrates a filling defect in the left anterior wall of ascending aorta (red arrows). (A) Axial image, (B) Sagittal image.

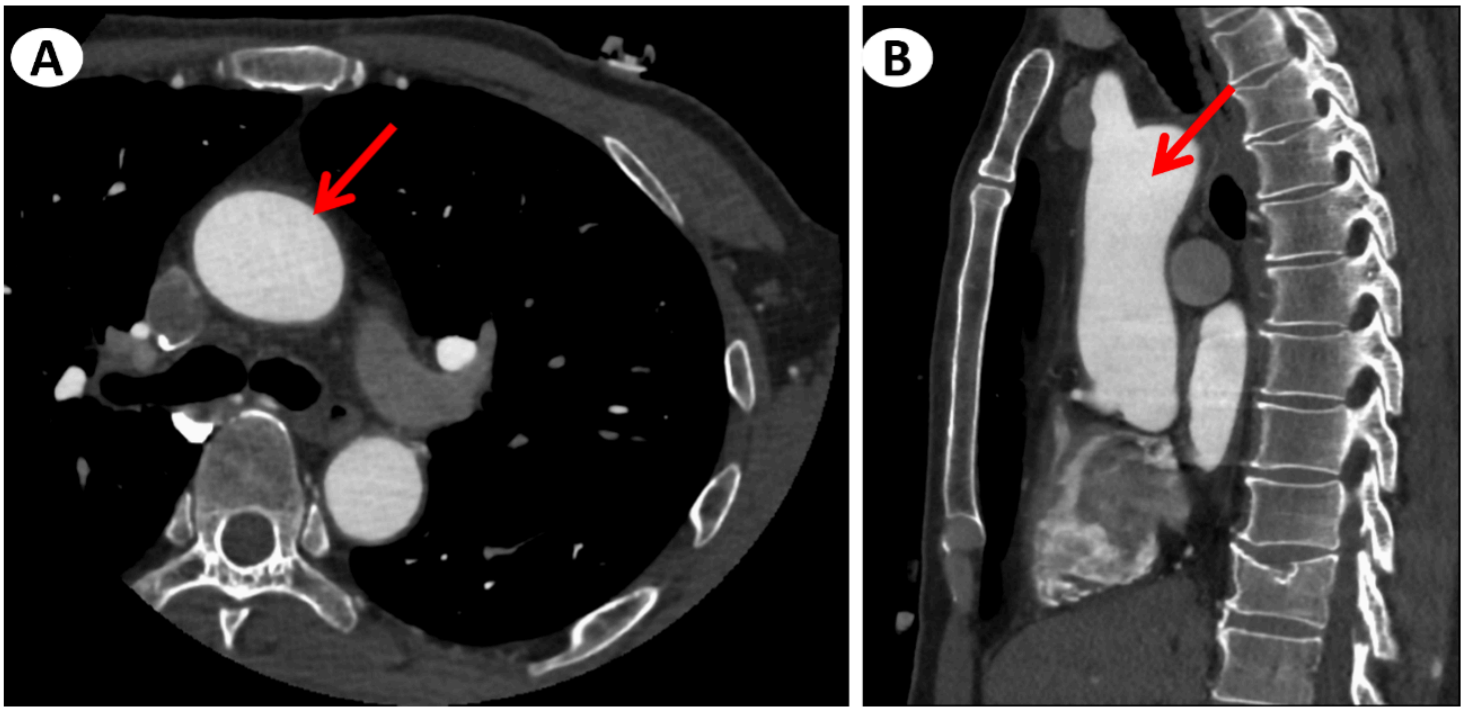


Figure 3

Follow-up contrast-enhanced computed tomography angiography (CTA) after 2 weeks of anticoagulant therapy shows that the previously identified filling defect have completely resolved (red arrows). (A) Axial plane, (B) Sagittal plane.