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Cortical Hand Knob Paradoxical Thromboembolic Stroke in an Adolescent with Secundum Atrial Septal Defect and Paget-Schroetter Syndrome

Maksim Parfyonov, MD[1], Danielle Porritt, BSc[2], Dakota Peacock, MD[3,4], Ryan Dragoman, MD[5] and James Lee, MD[3,4]

Abstract

Venous thoracic outlet syndrome (vTOS) is an increasingly recognized diagnosis in young patients in which the subclavian vein is compressed within the costoclavicular space. With repetitive compression, thrombosis can develop and has been referred to as "effort thrombosis" or the Paget-Schroetter syndrome. Here, we present a 16-year-old boy with vTOS who presented with acute ischemic stroke (AIS) in the hand knob region of precentral gyrus due to paradoxical embolus in the setting of atrial septal defect.

Keywords

Paget-Schroetter syndrome, venous thoracic outlet syndrome, TOS, stroke

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Background

Thoracic outlet syndrome (TOS) refers to compression, in order of frequency, of the brachial plexus (neurogenic TOS), subclavian vein (venous TOS), or subclavian artery (arterial TOS), as they exit the thoracic cavity.[1] Acute ischemic stroke (AIS) has been previously described in patients with arterial TOS.[2,3] Here, we report a 16-year-old male presenting with a paradoxical acute ischemic stroke in the setting of combined *venous* TOS and atrial septal defect (ASD).

Case Study

A previously healthy, right-handed 16-year-old male presented to our emergency department with left facial droop and left hand weakness which resolved after 3 h. Earlier in the day, he experienced paresthesias in his left leg, spreading to his left arm and hand, which then progressed to distal left hand weakness over the course of hours. Recent history revealed recurrent episodes of sensory disturbance along the medial aspect of his left forearm and hand lasting for hours at a time, precipitated by shoulder exercise. This was occurring 1-2 weeks prior to presentation. There was no associated pain, swelling or discoloration of his arms or legs.

On admission, his examination was notable for mild flattening of the left nasolabial fold and distal hand weakness (NIHSS score of 2, 1 for facial palsy and 1 for left arm drift). An urgent non-contrast computed tomography and computed tomography angiograph (CTA) showed no evidence of vascular stenosis or early ischemic changes. However, subsequent brain magnetic resonance imaging (MRI) on day 3 of admission identified restricted diffusion in the hand knob of the right pre- and post-central gyri on diffusion-weighted imaging (DWI) and reduced apparent diffusion coefficient (ADC) sequences (Figure 1). His symptoms gradually resolved by 48 h post onset of symptoms; he was symptomatic at the time of imaging.

[1] Neurological Institute, Cleveland Clinic Foundation, Cleveland, OH, USA
[2] Department of Biology, University of British Columbia, Vancouver, Canada
[3] Department of Pediatrics, Division of Neurology, BC Children's Hospital, Vancouver, Canada
[4] BC Children's Hospital, Vancouver, Canada
[5] Department of Radiology, Vancouver General Hospital, Vancouver, Canada

Corresponding Author:
James Lee, Division of Neurology, Department of Pediatrics, Faculty of Medicine, University of British Columbia, Vancouver, Canada.
Email: James.lee@cw.bc.ca

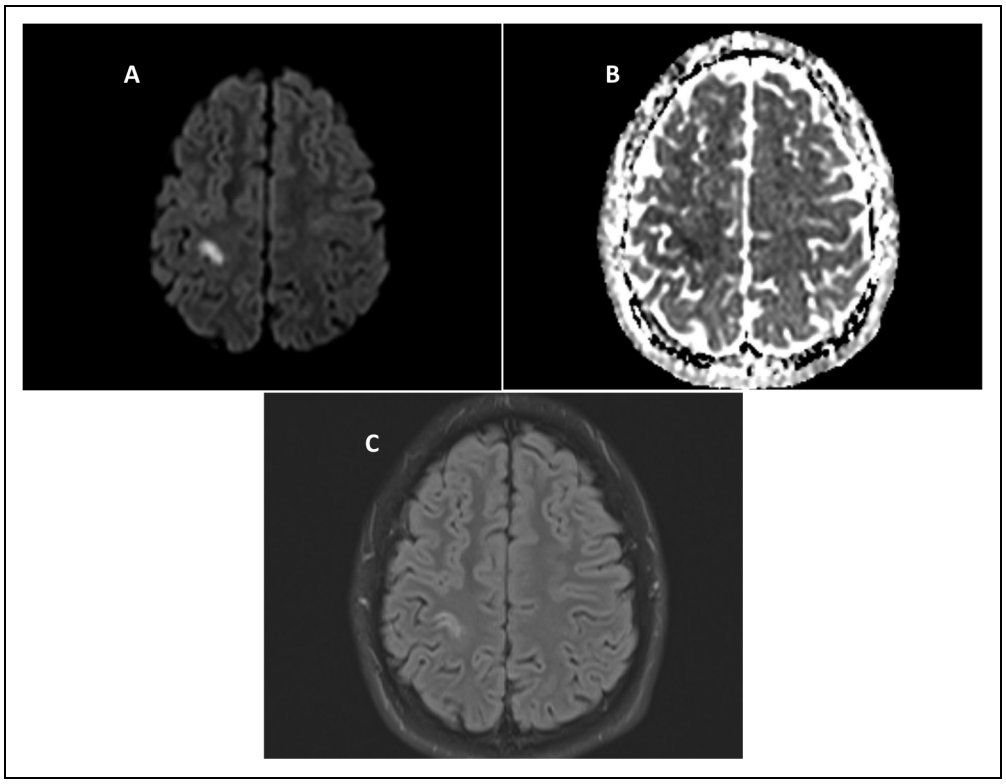

Figure 1. Brain magnetic resonance imaging 3 days post onset of neurological symptoms. (A) Diffusion-weighted imaging; (B) Apparent diffusion coefficient; and (C) Fluid attenuated inversion recovery.

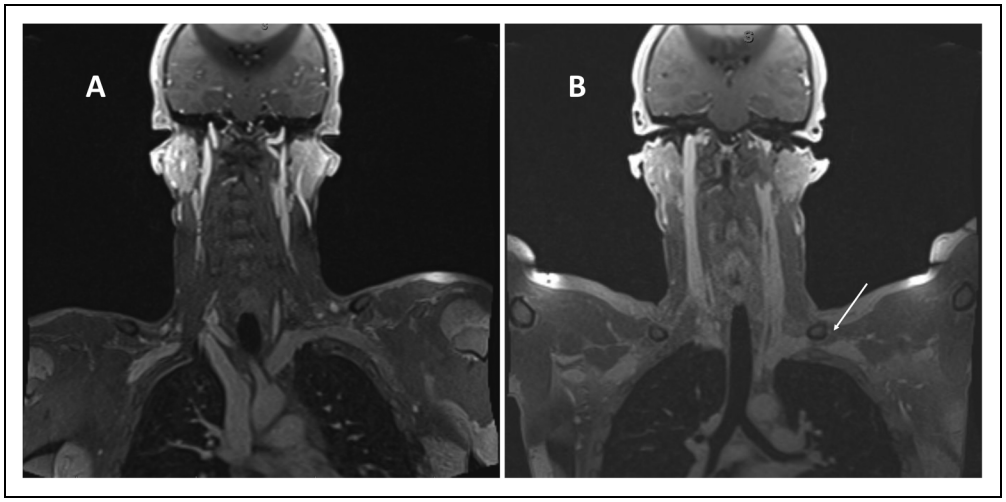

Figure 2. Magnetic resonance venogram (MRV) performed post-gadolinium enhancement demonstrating (A) patent left subclavian vein with arms down and (B) complete occlusion of the left subclavian vein with arms abducted.

Contrast-enhanced MRI of the neck revealed dynamic bilateral compression of the subclavian veins with arms raised (Figure 2), and a thrombus present in the left brachial vein. This finding, together with echocardiogram showing secundum atrial septal defect confirmed our suspicion for stroke mechanism. Echocardiogram demonstrated atrial septal defect measuring approximately 13 mm with left-to-right flow. A comprehensive workup for hypercoagulable factors was negative, and the patient was commenced on therapeutic anticoagulation. He subsequently had endovascular ASD closure and left transaxillary first rib resection. At one year follow-up, his neurologic function was intact.

Discussion

Cardioembolic stroke is a common cause of pediatric ischemic stroke, accounting for up to one third of cases[4] . Congenital

heart diseases including atrial septal defects significantly increase this risk.[5] The atrial defect creates a passage for venous emboli to be transmitted from the right to left atria, and evade the pulmonary circulation that would normally filter small venous thromboemboli. Often, the source of the original embolus is not found despite routine investigations, and other causes need to be considered.

Venous thoracic outlet syndrome (vTOS) is a rare diagnosis with incidence estimated between 1-2 in 100,00, most commonly seen in adolescent males.[6] Intermittent compression of the subclavian vein as it passes within the costoclavicular space can produce pain, edema and paresthesia. As in our patient, certain activities such as overhead movements can exacerbate the problem and cause repetitive injury, resulting in thrombosis which has been referred to as "effort thrombosis" or the Paget-Schroetter syndrome.[1]

While stroke has been reported as a complication of arterial TOS,[2,3,7] this is the first description of a paradoxical embolus associated with a *venous* TOS in the setting of atrial septal defect. Interestingly, an analogous condition has been described with co-occurrence of PFO and May-Thurner syndrome, in which chronic compression of the left common iliac vein by the overlying right common iliac artery results in iliofemoral deep vein thrombosis and has been associated with paradoxical stroke.[8,9]

Clinicians evaluating pediatric stroke patients with a suspected cardioembolic source should be aware of this rare association and consider imaging for vTOS in a patient reporting upper limb painful swelling or parethesias with repetitive shoulder exercises.

Author Contribution

MP contributed to conception and design contributed to acquisition, analysis, and interpretation drafted manuscript critically revised manuscript gave final approval agrees to be accountable for all aspects of work ensuring integrity and accuracy.

DP (Danielle Porritt) contributed to conception and design contributed to acquisition drafted manuscript critically revised manuscript gave final approval agrees to be accountable for all aspects of work ensuring integrity and accuracy.

DP (Dakota Peacock) contributed to conception and design contributed to analysis and interpretation drafted manuscript critically revised manuscript gave final approval agrees to be accountable for all aspects of work ensuring integrity and accuracy.

RD contributed to analysis and interpretation critically revised manuscript gave final approval agrees to be accountable for all aspects of work ensuring integrity and accuracy

JL contributed to conception and design contributed to analysis and interpretation drafted manuscript critically revised manuscript gave final approval agrees to be accountable for all aspects of work ensuring integrity and accuracy.

Declaration of Conflicting Interests

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Ethics and Patient Consent

Our institution does not require ethical approval for reporting individual cases. Verbal informed consent was obtained from the patient and their family for their anonymized information to be published in this article.

ORCID iD

Maksim Parfyonov https://orcid.org/0000-0003-0582-0852

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