

Introduction

Styloidogenic jugular venous compression syndrome (SJVCS) is a rare cause of idiopathic intracranial hypertension (IIH). This diagnosis is often reached after extensive diagnostic workup for a chronic headache.

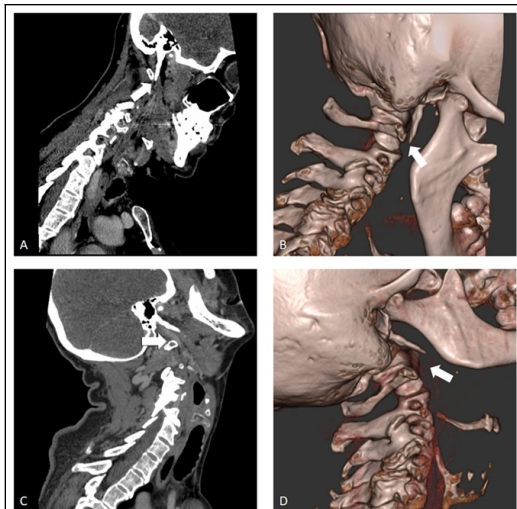
Methods

We conducted a retrospective review of all consecutive patients with a diagnosis of SJVCS who underwent microsurgical decompression between April 2009 and October 2017. We reviewed medical records and clinical images to abstract diagnostic and clinical features. We also compared this series with a control group diagnosed as IIH who had normal results on venography and manometry.

Results

10 patients with SJVCS presented with headaches; 7 of the 10 patients had headaches that were exacerbated by neck flexion. A total of 11 controls with IIH who underwent venography but were negative for SJVCS were identified and studied.

The styloid process was significantly longer in the SJVCS group (mean 31.0±10.6 mm vs. 19.0±14.1 mm; P=0.03), whereas the distance between the styloid process and the C1 lateral tubercle was shorter in the SJVCS group (mean 2.9±1.0 mm vs. 9.9±2.8 mm; P<0.01). Venography and manometry revealed significantly higher global pressure and higher pressure gradient across the stenosis site in the SJVCS patients than in control patients (mean 2.86±2.61 cm H2O vs. 0.13±1.09 cm H2O, p=0.03). Venous pressure elevation during contralateral neck turning was identified in all 10 SJVCS patients (mean 4.28±2.50 cm H2O). All SJVCS patients were treated with transcervical microsurgical decompression. 9 of the 10 patients experienced postoperative improvement or resolution of their symptoms. One patient had transient postoperative dysphagia, and another patient reported facial droop and jaw numbness.



Computed tomography angiogram (CT) showing the right side of the neck during neck flexion (A) and three-dimensional reconstruction of the right side (B) illustrating a much narrower space (arrows) between the styloid process and the C1 lateral tubercle than during neck extension (C-D). Used with permission from Barrow Neurological Institute, Phoenix, Arizona.

Conclusions

SJVCS is a novel clinical entity similar to IIH, and patients should be evaluated with venography with manometry. Jugular venous stenosis is caused by osseous compression from the C1 lateral tubercle and the styloid process. Surgical decompression is an effective treatment option for selected patients.

Learning Objectives

By the conclusion of this session, participants should be able to: 1) Identify a novel reason for refractory pseudotumor cerebri; 2) Obtain the clinical diagnosis and identify the treatment for this novel condition.

References

Dashti SR, Nakaji P, Hu YC, et al.: Styloidogenic jugular venous compression syndrome: diagnosis and treatment: case report. Neurosurgery 70:E795-E799, 2012.