CASE REPORT

Moving target: transient rotational stenosis precipitating jugular bow hunter's syndrome

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SUMMARY

A 60-year-old man presented with a 10-month history of stereotypical spells characterized by vertigo, tinnitus, blurred vision, left hemibody numbness, and occasional syncope, precipitated by turning his head leftwards. Cerebral angiography and CT angiography of the head and neck with provocative maneuvers did not demonstrate vertebral artery narrowing. However, there was narrowing of the left internal jugular vein due to extrinsic compression from the sternocleidomastoid with leftward head rotation in the setting of hypoplasia of the right internal jugular vein. The patient underwent a cervical venogram which confirmed the finding. Manometric evaluation demonstrated a gradient of 29 mm Hg across the stenosis with the head turned leftwards compared with 1 mm Hg in the neutral position. The patient was treated with myectomies of the left sternocleidomastoid, posterior belly of the digastric, stylohyoid and omohyoid and styloid process removal. Following surgery, the patient reported complete resolution of symptoms. Repeat venography demonstrated resolution of the stenosis and pressure gradient.

BACKGROUND

Bow hunter's syndrome is defined as posterior circulation ischemia due to vertebral artery compression occurring with head turning.¹ Given the close relationship of the carotid arteries and jugular veins to cervical muscular and osseous structures, it is conceivable that rotational compression of these vessels could occur, resulting in neurological symptoms.^{2,3} We present a case of rotational compression of the left internal jugular vein (IJV) which resulted in a bow hunter-like syndrome. This was successfully treated with sternocleidomastoid myectomy.

CASE PRESENTATION

A 60-year-old man presented with a 10-month history of stereotypical spells characterized by vertigo, tinnitus, blurred vision, left hemibody numbness, and occasional syncope, precipitated by turning his head leftwards and looking down. Neurological symptoms gradually developed 10–15 s after performing this maneuver and were associated with facial congestion and lip cyanosis. Past medical history was unremarkable.

INVESTIGATIONS

MR venography demonstrated a dominant left IJV with a hypoplastic right transverse sinus and right IJV. CT angiography (CTA) of the head and neck

showed a dominant left vertebral artery with a hypoplastic right vertebral artery which terminated in a posterior inferior cerebellar artery. CTA performed while turning his head leftwards and replicating his stereotypical spells demonstrated a widely patent left vertebral artery. There was extrinsic compression of the left IJV by the sternocleidomastoid with leftward head rotation, with a nearly 90% reduction in cross-sectional area (from 210 mm² to 21 mm²) (figure 1).

Angiography of the left vertebral artery was performed in both the neutral position and with the provocative maneuver. During the provocative maneuver the patient experienced his stereotypical spell and was unresponsive for 5 s. No compromise of the left vertebral artery lumen was present. However, we noticed delayed venous egress through the left IJV. We then performed a left IJV venogram in both the neutral position and with leftward rotation which confirmed a severe left IJV stenosis with leftward rotation. Transcatheter venous manometry demonstrated that venous pressures above and below the stenosis were 5 mm Hg and 4 mm Hg, respectively, while with leftward head rotation the venous pressures were 35 mm Hg and 6 mm Hg, respectively (figure 2).

TREATMENT

Based on the elevated venous pressures associated with head turning and the extrinsic mass effect on the left IJV in the setting of contralateral IJV hypoplasia, we proceeded to surgically release the compressive left cervical musculature.

The patient was taken to the operative room and positioned supine with the head neutrally

Figure 1 CT angiograms/venograms with and without provocative maneuvers. (A) CT venogram of the neck in the neutral position demonstrates a widely patent left internal jugular vein (J) positioned between the left common carotid artery (C) and sternocleidomastoid (SCM). (B) CT angiogram with head turned to the left demonstrates extrinsic compression of the left internal jugular vein (J) between the common carotid artery (C) and sternocleidomastoid (SCM).

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Figure 2 Angiographic evaluation. (A) Left vertebral artery cervical angiogram shows no evidence of stenosis or occlusion of the left vertebral artery. (B) With the head turned to the left and reproduction of the patient's symptoms, there is no vertebral artery narrowing. (C) Venous phase images from the left vertebral artery angiogram show a large and capacious left internal jugular vein (black arrowhead) and a hypoplastic right transverse sinus and right internal jugular vein (white arrowhead). (D) Left internal jugular vein venogram in neutral position shows a widely patent jugular vein. (E) With the head turned to the left and reproduction of the patient's symptoms, there is a severe stenosis of the left internal jugular vein.

positioned. Demonstration of mechanical compression was undertaken by the surgeon placing a hand between the IJV and the sternocleidomastoid and then rotating the head. The inferior half of the muscle demonstrated compressive force with passive motion, and thus the inferior half of the muscle was removed via myectomy. The accessory nerve was identified and preserved as it entered the muscle, as was the posterior branch to the trapezius. After this maneuver there were several other places of relative stenosis appreciated visually where the omohyoid, posterior belly of the digastric, and stylohyoid muscles came across the jugular sheath. Therefore, 1 cm myectomies were performed of the aforementioned muscles. We then explored underneath the patient's jaw where an elongated and potentially compressive styloid process was identified and also removed ([figure 3](#)).

OUTCOME AND FOLLOW-UP

On postoperative day 1 the patient repeated the provocative maneuvers and reported complete symptom resolution. Repeat

left IJV venography in the neutral position and with the provocative maneuvers demonstrated complete resolution of the stenosis. In the neutral position the venous pressures above and below the stenosis were 6 mm Hg and 5 mm Hg respectively, while during the provocative maneuver the pressures were 7 mm Hg and 6 mm Hg ([figure 4](#)).

DISCUSSION

We present the first reported case of rotational compression of the IJV resulting in neurological symptoms from venous congestion and a large venous pressure gradient. This case is unique in that surgical release of the cervical musculature responsible for the compression was performed, resulting in symptom resolution and angiographic resolution of the IJV stenosis and pressure gradient.

While extrinsic compression of the IJV is common, a few studies have demonstrated that, in some patients, compression or occlusion of the IJVs results in venous hypertension and

Figure 3 Intraoperative photographs following left neck dissection, sternocleidomastoid myectomy, omohyoid myectomy, posterior digastric myectomy, stylohyoid myectomy, and styloid process resection. (A) With the head neutrally positioned, a wide margin between the external jugular vein (arrow) and internal jugular vein (arrowhead) is readily apparent, with no compression from the sternocleidomastoid remnant (triangle). (B) As the neck is rotated left, the internal and external jugular veins are brought in contact, with no evidence of stenosis or compression from the released sternocleidomastoid and omohyoid or other surrounding structures. (C) Superiorly, the styloid process has been removed from the skull base, visualized deep to the released posterior digastric and stylohyoid.

Figure 4 Postoperative angiographic evaluation. (A) Postoperative left internal jugular vein venogram in the neutral position shows no jugular vein narrowing. (B) With the head turned to the left, there is also no jugular vein narrowing.

elevated intracranial pressures.^{4–7} In general, those with severe intracranial pressure elevations following IJV occlusion have hypoplasia or occlusion of the contralateral IJV.^{4,5}

Acute symptomatic elevation of intracranial pressure from extrinsic IJV occlusion has been reported. In 1984, Gooding and Stimac reported two premature infants who had episodes of increased intracranial pressure with head turning following jugular catheter placement.³ In response to this, the authors performed a cadaveric study examining the effect of head turning on IJV caliber in an infant cadaver and noted occlusion of the ipsilateral IJV with forcible head turning. Based on this finding, the authors surmised that the combination of the jugular venous catheter and head turning in these infants resulted in obstruction of venous outflow and resultant intracranial pressure increase.³

Surgical treatment of extrinsic IJV compression has been reported in previous case reports and series. Dashti *et al*⁴ reported two patients who presented with chronic venous hypertension from extrinsic compression of a dominant IJV between the styloid process and lateral mass of C1 in patients with a hypoplastic contralateral IJV. Both patients were successfully treated with resection of the styloid process. In a series of five patients Ho *et al* reported five cases of extrinsic IJV compression due to juxtaposition of the styloid process and lateral mass of C1, which were also successfully treated with styloidectomy.⁸ These cases differ from our case in that symptoms were not associated with neck turning and were primarily due to extrinsic mass effect from osseous structures in the upper cervical spine rather than muscular structures in the lower cervical soft tissues.

CONCLUSION

Jugular bow hunter's syndrome is likely an under-recognized phenomenon which is caused by compression of a dominant IJV with ipsilateral head rotation. This diagnosis should be suspected in cases where patients have reproducible neurological symptoms with head rotation, but no evidence of vertebral artery compression. In the appropriate clinical setting, decompression of the jugular vein by release or resection of the offending muscular or osseous structures may be a safe and effective therapeutic strategy.

Learning points

- ▶ Rotational compression of the jugular vein can result in neurological symptoms similar to those seen in bow hunter's syndrome due to acute venous congestion.
- ▶ It is important to document the presence of a hypoplastic or absent contralateral jugular vein when diagnosing a patient with jugular bow hunter's syndrome.
- ▶ Close inspection of venous drainage patterns on angiography should be performed in cases where bow hunter's syndrome is suspected but there is no evidence of vertebral artery compromise.
- ▶ In cases where a structural cause of jugular venous stenosis is identified, surgical treatment can be safely performed and may be curative.

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Competing interests None declared.

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