



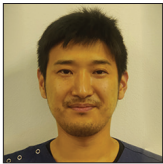
## Case Report

# The eagle jugular syndrome as the cause of delayed intracranial hemorrhage after microvascular decompression for hemifacial spasm: A case report

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Received : 12 August 2021

Accepted : 11 November 2021

Published : 30 November 2021

### DOI:

10.25259/SNI\_808\_2021

### Quick Response Code:



## ABSTRACT

**Background:** Eagle syndrome is a rare disorder whereby an elongated styloid process (ESP) causes not only some otolaryngological symptoms, but also cerebrovascular events caused by compression of the carotid artery. In recent years a syndrome, denominated as Eagle jugular syndrome, involving internal jugular vein (IJV) compression caused by an ESP has been proposed as a variation of Eagle syndrome. Clinical impact of the Eagle jugular syndrome on neurosurgical procedures has not been reported yet.

**Case Description:** We present a case of a 68-year-old woman who underwent microvascular decompression for hemifacial spasm of the left side and developed delayed intracranial hemorrhage on postoperative day 3. We also demonstrate that this patient developed ipsilateral IJV stenosis between an ESP and the muscle bundle of the rectus capitis lateralis with antero-flexion neck position, which would induce venous congestion in addition to surgical disruption of emissary vein.

**Conclusion:** This case is the first report demonstrating the association of an ESP with postoperative delayed intracranial hemorrhage. Our report elucidates the importance of the awareness among neurosurgeons of considering the ESP as an important bony anomaly, especially when planning for posterior fossa surgery.

**Keywords:** Delayed intracranial hemorrhage, Eagle jugular syndrome, Elongated styloid process, Hemifacial spasm, Internal jugular vein stenosis, Microvascular decompression

## BACKGROUND

The styloid process is a slender bony projection arising from the lower surface of the petrous portion of the temporal bone. In 1937, Eagle described two cases in which cervicofacial pain, dysphagia, and otalgia were caused by an elongated styloid process (ESP), which is well known as the Eagle syndrome.<sup>[7]</sup> This syndrome had been divided into two groups; “the classical styloid process syndrome” and “the styloid-carotid artery syndrome” according to the mode of presentation.<sup>[1,9,15]</sup> The former is characterized by spastic and nagging pain in the pharynx radiating into the mastoid region attributed to stretching of the sensory nerve endings of the fifth, seventh, ninth, and tenth cranial nerves by ESP, which is often produced

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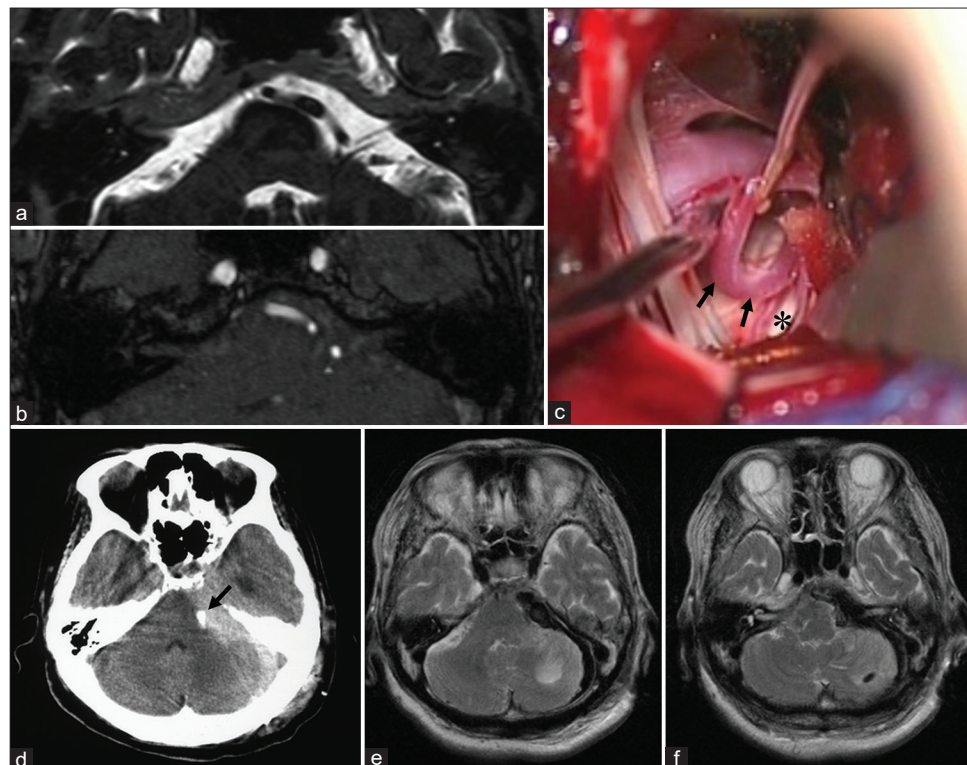
by post tonsillectomy fibrosis. The latter is attributed to impingement of the internal carotid artery (ICA) by ESP and is also characterized by cervicofacial pain along the distribution of the sympathetic nerve fibers around the ICA, which occasionally results in neurological symptoms, as a vascular type of Eagle syndrome, including transient ischemic attack, syncope, Horner's syndrome, and stroke due to ICA compression or dissection caused by an ESP.<sup>[3,8]</sup> Contrastively, clinical impact of the ESP on the internal jugular vein (IJV) has been rarely reported.

Microvascular decompression (MVD) is a type of comparatively safe and effective functional surgery for hemifacial spasm (HFS) that has a cure rate of approximately 85–90% with < 10% operative morbidity.<sup>[17]</sup> However, fatal complications also rarely occur after MVD with mortality ranging from 0.2% to 2.0%, and postoperative intracranial hemorrhage is one of the serious surgical complications leading to mortality, which ranges from 0.1% to 0.55%.<sup>[10,11,18,20]</sup> We report a case of "Eagle jugular syndrome" in a patient who developed delayed intracranial hemorrhage after MVD for HFS, which was caused by ESP related IJV stenosis.

## CASE DESCRIPTION

A 68-year-old woman who presented with a 5-year history of typical left HFS was admitted to our hospital for neurosurgical treatment. Magnetic resonance (MR) imaging revealed the presence of neurovascular compression at the root exit zone (REZ) of the left facial nerve by the left anterior inferior cerebellar artery [Figure 1a and b]. The patient underwent MVD through retro-sigmoid approach under park-bench position. During surgery, mastoid emissary vein was disconnected and hemostasis was achieved at the mastoid canal using bone wax as usual. After a retro-mastoid keyhole craniotomy, the affected nerve was decompressed with mobilization and dislocation of the offending artery by setting a piece of Teflon proximal to the REZ [Figure 1c]. Operative time was 2 h 20 min and there were no complications during and immediately after the surgery. Remission of the HFS was observed immediately after MVD.

Postoperative computed tomography (CT) scan on the next morning after surgery revealed no abnormal findings. Except for complain of headache, the postoperative course appeared uneventful. However, on the 3<sup>rd</sup> postoperative day,



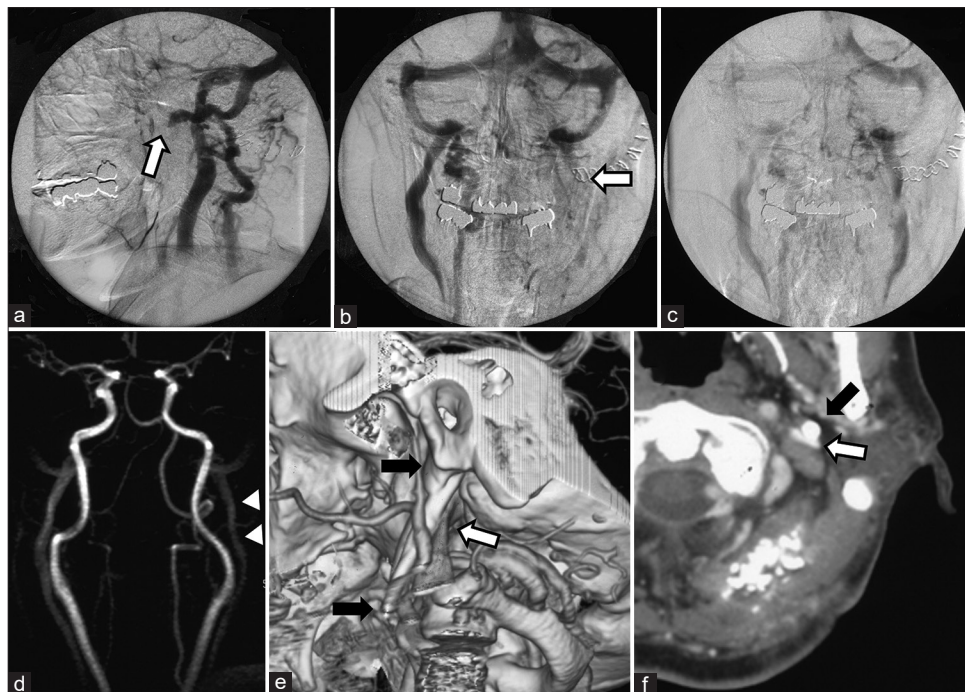
**Figure 1:** Preoperative images on heavily T2-weighted magnetic resonance (MR) image (a) and source image of MR angiography (b) showing neurovascular compression at the root exit zone (REZ) of the left facial nerve by the anterior inferior cerebellar artery. Intraoperative photographs showing (c) the REZ of the affected nerve (Asterisk) was readily relieved by the offending artery (Black arrow) with minimum manipulation. Postoperative day 3, (d) head computed tomography image showing a subdural hematoma in the left cerebellopontine angle (CPA). Outstanding high-density lesion at the REZ is surgical prosthesis (Black arrow). T2-weighted MR images (e, f) showing a subdural hematoma in the left CPA and a cerebellar hemorrhage with perifocal edema that could indicate venous infarction.

her conscious level deteriorated while asleep. Whereas she was resting sideways due to the pain of the wound for a while after the operation, her head position had been raised with a pillow at that time. CT scan revealed a subdural hematoma in the left cerebellopontine angle (CPA) with hydrocephalus [Figure 1d]. The patient was treated conservatively with close monitoring for further examination of the cause of this delayed hemorrhage. MR images also indicated an intracerebellar hemorrhage with perifocal edema [Figure 1e and f]. Angiography did not reveal any underlying vascular malformation; however, we noticed that the left IJV was obstructed at the level of C1 by some structure [Figure 2a-c], ipsilateral IJV stenosis was not evident on preoperative MR images [Figure 2d]. The three-dimensional CT (3D-CT) venogram reconstructed images revealed that this offending structure was an ESP that was 48 mm in length as measured [Figure 2e]. The former axial images of the 3D-CT venogram also indicated that the IJV passed transversely across between the ESP and the rectus capitis lateralis muscle and was pinched between them [Figure 2f]. The ESP did not require any surgical treatment because the hematoma in the CPA and the hydrocephalus were resolved gradually under management with appropriate neck positioning. MR venography performed before discharge also revealed

reduction of signal flow on ipsilateral transverse-sigmoid sinus by cervical antero-flexion compared with posterior-extension [Figure 3a and b]. We concluded that this is an interesting case developing delayed intracranial hemorrhage after MVD for HFS, which was caused by ESP related IJV stenosis. Fortunately, the patient was discharged with no neurological deficits and the HFS was resolved over a 2-year follow-up examination period.

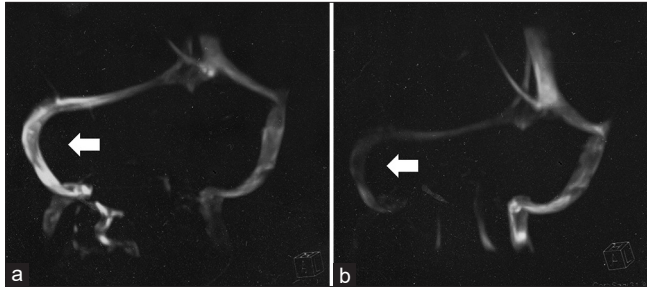
## DISCUSSION

Anatomically, the styloid process projects inferiorly and anteriorly into the parapharyngeal space where it is in close proximity to numerous structures. The retrostyloid compartment contains the IJV, the ICA, the sympathetic chain, and the glossopharyngeal, vagus, accessory, and hypoglossal nerves. The prestyloid compartment contains the internal maxillary artery, the lingual and auriculotemporal nerves, and is related to the tonsillar fossa inferiorly.<sup>[15]</sup> The normal length of the styloid process is reported to be between 21 and 29.5 mm, and the styloid process is defined as an ESP when greater than 30 mm in length.<sup>[9]</sup> Although the incidence of ESP is reported between a range of 3.7–43.89%,<sup>[4]</sup> only 4–10.3% of patients with ESP are symptomatic.<sup>[15]</sup>



**Figure 2:** Investigations for the causality of delayed intracranial hemorrhage revealed (a, b) interruption of the ipsilateral internal jugular vein (IJV) at the level of the C1 (White arrow) on angiography. (c) The venous outflow was congested but not occluded, which revealed on further delayed phase. (d) Preoperative MR angiogram showed no stenosis at this portion of the IJV (Arrow head). (e) The three-dimensional computed tomography (3D-CT) venogram showing an elongated styloid process (ESP) that was 48 mm in length (Black arrow) and compressed the IJV at the level of the third segment of the vertebral artery (White arrow). (f) The former axial images of the 3D-CT venogram also showed that the IJV (White arrow) was pinched between the ESP (Black arrow) and the rectus capitis lateralis. It is notable that the transverse process of C1 vertebra did not contribute to the pinching directly in this case.





**Figure 3:** Magnetic resonance (MR) venography showing alteration of the signal flow on the bilateral transverse-sigmoid sinus. (a) MR venography with posterior-extension of the neck position showing high-signal flow on the left transverse-sigmoid sinus. (b) MR venography with antero-flexion of the neck position showing significant reduction of the signal flow on the affected side (a and b; White arrow).

Eagle syndrome presents various symptoms due to the mechanical compression of the surrounding anatomical structures by the ESP, which is occasionally triggered by neck mobility including flexion/extension and contralateral rotation.<sup>[9]</sup> ESP may disrupt intracranial venous outflow by inducing IJV stenosis and result in increasing of intracranial pressure. Clinical manifestations of IJV stenosis by the ESP have been also reported as insomnia, tinnitus, head noises, hearing loss, headache, and visual defects,<sup>[2]</sup> which may lead to intracranial hemorrhage as the result of intracranial venous hypertension. Recently, several cases with pseudotumor cerebri or perimesencephalic subarachnoid hemorrhage have been reported whose causes were attributed to intermittent IJV compression by an ESP, which were denominated as the “Eagle jugular syndrome,” a venous variant of the Eagle syndrome.<sup>[2,6,13,19]</sup> This is a valuable case in which delayed intracranial hemorrhage occurred after MVD, clearly implying a relationship between the ESP and the IJV.

Delayed intracranial hemorrhage after MVD has been rarely reported (with an incidence of 0–0.4%) as one of postoperative fatal complications.<sup>[10,11,14,18,20]</sup> Li *et al.* advocated that one of four possible mechanisms of such fatal complication is venous hemorrhagic infarction due to obstruction of the IJV triggered by intraoperative excessive rotation and flexion of the neck for surgical position.<sup>[14]</sup> Actually, venous walls are thinner than those of the arteries and lack smooth muscle and elastic fibers, thereby making veins more vulnerable to deformity under extrinsic compression.<sup>[15]</sup> However, several collateral venous channels such as the vertebral venous plexus, emissary veins, and pterygoid venous plexus usually compensate for the anterograde venous return during IJV stenosis.<sup>[12]</sup>

While this complication was unpredictable during the intraoperative procedure and, unfortunately, preoperative routine examinations, we speculate that the causes of

delayed hemorrhage may be both maintaining a specific neck position and the disconnection of these compensable collateral venous channels such as the mastoid emissary vein, whose disruption is often unavoidable during lateral suboccipital craniotomy.<sup>[16]</sup> As the IJV stenosis is maintained at antero-flexion of the neck position, this speculation of the mechanism developing delayed intracranial hemorrhage could be explained by the fact that it occurred during sleep while using a pillow but not during wakefulness.

In addition, the structure opposite of the ESP to pinch the IJV is the transverse process of the C1 vertebra in most of the studies;<sup>[2,6,13,19]</sup> however, the muscle bundle of the rectus capitis lateralis may also be a favored candidate as in this case, which is another new variation of the Eagle jugular syndrome with respect to the mechanism.<sup>[5]</sup>

## CONCLUSION

Postoperative delayed intracranial hemorrhage after MVD is an unexpected and fatal complication. Whereas Eagle jugular syndrome is a venous variant of the Eagle syndrome, neurosurgeons should regard the ESP as an important bony structure, which has the potential to induce venous congestion by compressing the IJV, especially when planning for posterior fossa surgery.

## Acknowledgments

We thank the nursing and anesthesiology teams for the perioperative management of the patient who underwent neurological surgery. We would like to thank Editage ([www.editage.com](http://www.editage.com)) for English language editing.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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**How to cite this article:** Nonaka T, Sakata K, Abe T, Hattori G, Orito K, Miyagi N, *et al.* The eagle jugular syndrome as the cause of delayed intracranial hemorrhage after microvascular decompression for hemifacial spasm: A case report. *Surg Neurol Int* 2021;12:584.