Case report

New Eagle’s syndrome variant complicating management of intracranial pressure after traumatic brain injury

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1. Introduction

Eagle’s syndrome is a rare condition consisting of throat pain, pharyngeal foreign body sensation, otalgia, and neck pain. It is due to abnormally long styloid processes or by progressive mineralisation of Reichert’s cartilage in the stylohyoid ligamentous complex. The styloid process may also compress the internal or external carotid arteries and the perivascular sympathetic fibers causing neck and face pain or syncope with head rotation. We report a case of factitiously increased intracranial pressure after traumatic brain injury due to unilateral traumatic sigmoid sinus occlusion in combination with contralateral jugular venous compression by an elongated and posteriorly positioned styloid process.

2. Case report

This 47-year-old male was found down unresponsive in his home. At presentation, the patient’s Glasgow Coma Scale score was 7. A CT scan revealed bilateral temporal contusions, left greater than right, a large left frontal contusion, right sided temporal bone fracture, and 4 mm of midline shift. The basal cisterns were visible. A fiberoptic intracranial pressure (ICP) monitor was placed, with an opening pressure of 35 mmHg.

Over the next 24 h, the intracranial pressure became increasingly resistant to sedation, paralytics, mannitol, and hypertonic saline. Despite these maneuvers, he continued to have unprovoked ICP elevations into the 50s. Repeat imaging demonstrated significant radiographic improvement. His pupils remained small and reactive. These elevations in ICP were managed with sedation, paralytics, mannitol, and hypertonic saline. Despite these maneuvers, he continued to have unprovoked ICP elevations into the 50s. Follow up CT scans continued to show open basilar cisterns and decreased cerebral oedema. The patient was pharmacologically paralysed, and his pupils did not dilate with the spikes of ICP.

During the postoperative course, a CT angiogram (CTA) was performed as part of a standardised institutional protocol to screen all patients with basilar skull fractures for blunt carotid and vertebral arterial injuries. This demonstrated the presence of right sigmoid thrombosis underlying the mastoid fracture, which was thought to be contributing to his ongoing elevated ICP.

Five days later, a subsequent review of the CTA noted the presence of the elongated styloid processes (4 cm), with associated compression of the residual patent jugular vein between the styloid and the lateral mass of C1. A brain tissue oxygen monitor (Licox, Integra Neuroscience, Plainsboro, New Jersey) was placed to help guide treatment. The patient’s PbO2 was found to be normal, even when there were spikes in the ICP. We felt this provided reassurance that the ICP figures were factitious, and not requiring any further aggressive therapy. Concurrently, the elevated intracranial pressures also began to diminish, and he was weaned off the sedation and paralytics. Over the next few days, these monitors were removed, and he began following simple commands. Heparin was then instituted to further facilitate dissolution of the sigmoid thrombus. He was then transferred to an inpatient rehab center, awake and alert, following commands in all his extremities, and starting to speak simple phrases. During his inpatient rehabilitation stay, it was also discovered that the patient had sarcoidosis. Three months after discharge, the patient’s Glasgow Outcome Score was 4.
3. Discussion

The styloid process is an elongated protrusion of the temporal bone. It arises from the second brachial arch (Reichert’s cartilage). It is near the carotid artery, the internal jugular vein, and cranial nerves VII, IX, X, and XII. Eagle’s syndrome consists of symptoms of recurrent throat pain, pharyngeal foreign body sensation, dysphagia, referred otalgia, and neck pain. In 1937, Eagle defined stylalgia related to an elongated styloid process, defining an elongated styloid process as >25 mm. Eagle described 2 separate syndromes associated with an elongated styloid process: classic stylohyoid syndrome and stylocarotid syndrome. The classic syndrome occurs almost always after tonsillectomy, involving dull pain in the tonsillar fossa that radiates to the ear. It is also associated with facial-cervical pain. The stylocarotid syndrome is due to compression of the internal and external carotid arteries and the associated sympathetic nerve fibers. Symptoms are exacerbated by turning the head to the extreme lateral position. If the ECA is compressed, pain is in the infraorbital, temporal and mastoid regions, and if the ICA is compressed the pain is in the ophthalmic area.

Venous outflow obstruction in the form of dural sinus thrombosis should be investigated in cases in which intracranial pressures appear out of proportion to the radiographic findings. In general, dural sinus thrombosis has rarely been associated with closed head injury. However, there are several series that document its occurrence. Satch et al. in 1993 presented two cases where the presenting sign was increased intracranial pressure. The mechanism of dural sinus thrombosis after closed injury is not known. It may be caused by bone fragments, sinus dissection, or sinus distortion that may initiate thrombus formation. Treatment may be complicated, as it was in our patient, by haemorrhagic injuries to the brain. Thus, the risks and benefits of anticoagulation must be carefully balanced.

In our patient, it appears that the compression of his internal jugular veins complicated the measurement and management of his intracranial pressure. Despite the improved appearance of the postoperative CT scans, his measured ICP continued to be significantly elevated and resistant to medical management. We believe that this was due to the obstruction of venous outflow from the combination of the sigmoid sinus occlusion and the bilateral jugular vein compression by elongated styloid processes. It is our opinion that this patient likely lived in a compensated state prior to his injury as a result of venous collateral drainage. Loss of one jugular vein as a result of the sigmoid thrombosis likely threw the patient into an uncompensated state, leading to the factitiously elevated ICP. Moreover, we feel that aggressive treatment of elevated ICP in the presence of venous compression is likely unwarranted, absent other causes that would normally require either medical or surgical intervention, or absent obvious symptoms. In this case, such a cause (contusion with shift) was treated, and ICP would likely have been normal had there not been venous outflow restriction.

It is interesting to note that the etiology of our patient’s fall is unknown. He was not alone in the house at the time he was found, yet was found unconscious and lying in a pool of emesis. There is no history suggestive of any known trauma prior to this. We have hypothesised that his styloid processes may have compressed one of his internal carotid arteries when he turned his head. This may...
Fig. 2. (a) Axial CTA with right sigmoid sinus occlusion (black arrow) and dye present in the left sigmoid sinus (white arrow). (b) CT venogram confirming the left internal jugular vein compressed between the styloid process (black arrow) and lateral mass of C1 (white arrow). (c) 3D CTA demonstrating the left internal jugular vein compressed between the styloid process (black arrow) and lateral mass of C1 (white arrow). (d) Left internal jugular vein, both compressed between the styloid and C1 lateral mass. (e) Sagittal view of subsequent CTA demonstrating compression of the right internal jugular vein, but some early recanalisation.
have lead to a brief episode of ischaemia, and then syncope. Fig. 2c demonstrates the proximity of the styloid to the carotid artery, in addition to the jugular compression. In questioning the patient’s family, he had at least one prior episode of unexplained loss of consciousness. Thus, this variant of Eagle’s syndrome may explain not only his unusual course, but also may explain the cause of the fall, as well. We doubt that his diagnosis of sarcoidosis had any influence on his clinical course at this time, as there was no evidence of neurosarcoidosis present during his hospitalisation.

4. Conclusions

Eagle’s syndrome is a rare condition that usually presents with pain syndromes or vascular insufficiency by elongated styloid processes. In our patient, this anatomical feature complicated measurement and management of his intracranial pressure, by presumably contributing to his elevated venous pressure. We believe that this demonstration of venous compression constitutes a new variant of Eagle’s syndrome. We conclude that, when managing patients with unexplained ICP elevations, one should attempt to correlate the CT findings with the clinical picture and vigorously search for various causes of venous outflow disturbance in cases in which the measured ICP appears out of proportion to the imaging findings. This variant of Eagle’s syndrome may represent a rare cause of such venous insufficiency that should be in the differential diagnosis of unexplained or disproportionately elevated ICP.

References


