

A DAY IN THE LIFE OF A NEUROCRITICAL CARE TRAINEE



# Bilateral Bow Hunter's Syndrome Mimicking a Classic Seizure Semiology

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## Introduction

Bow Hunter's Syndrome, or rotational vertebrobasilar insufficiency, is a rare disorder characterized by posterior circulation ischemia elicited by neck rotation. The term was coined in 1978 when a 39-year-old man presented with lateral medullary syndrome after practicing with a bow in preparation for deer hunting [1]. His symptoms were hypothesized to be caused by the rotation of his neck while sighting an arrow resulting in dynamic, unilateral vertebral artery occlusion. Since then, the definition of rotational vertebrobasilar insufficiency has been refined to mechanical vertebral artery occlusion in the setting of poor collateral flow, with most cases occurring transiently [2]. The occlusion may have multiple etiologies including bone spurs, fibrous bands, chiropractic manipulation, cervical disk herniation, and rarely trauma [2, 3]. Symptoms occur for a median duration of 12 months before initial presentation and most commonly include vertigo, visual changes, pain, nausea, vomiting, and syncope or pre-syncope [2, 4]. Diagnosis may involve multiple imaging modalities including computed tomography (CT) angiography, magnetic resonance angiography (MRA), transcranial doppler, and digital subtraction angiography. Dynamic angiography in which the patient's head is rotated during imaging to demonstrate vertebral artery occlusion is considered the gold standard [5].

Here, we report a challenging case of rotational vertebrobasilar insufficiency that presented to the neurological and neurosurgical intensive care unit (NNICU). This case involved several diagnostic and management challenges—including an atypical history, a masquerading set of symptoms, and bilateral lesions—that required

complex decision-making and provided multiple learning points for neurocritical care trainees.

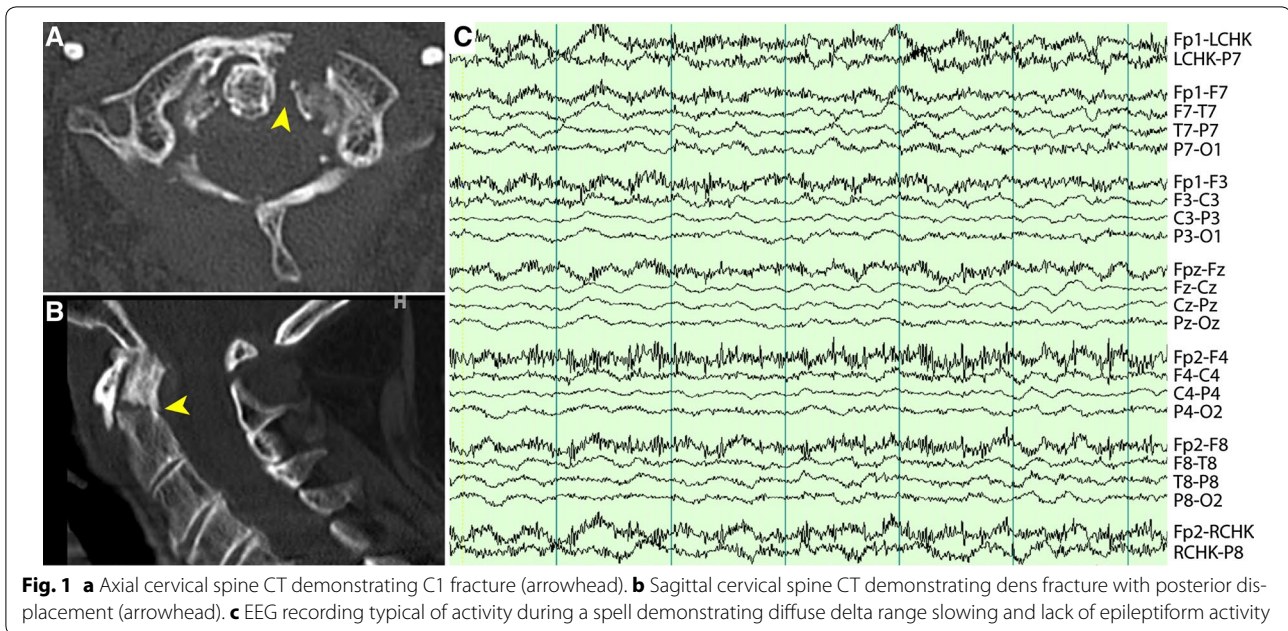
## Case Presentation

An 84-year-old woman with a history of rheumatoid arthritis, major depressive disorder, and childhood seizures presented to our hospital with several days of vertigo and postural instability. At baseline, the patient was wheelchair-dependent because of severe rheumatoid arthritis. Two weeks prior to presentation, she suffered an incidental fall out of bed and struck her head. She was taken to a community emergency room where X-ray imaging was unrevealing and she was discharged home. After discharge, she suffered near-constant neck pain.

Three days prior to presentation, she began experiencing spells of postural instability, vertigo, and nausea. These symptoms had not occurred prior to her fall out of bed. She presented to our emergency room where a head CT was unremarkable. A neck CT revealed a complex C1 vertebral fracture (Fig. 1a) as well as an odontoid fracture with posterior displacement (Fig. 1b) in the setting of severe degenerative disease. She was placed in a cervical collar. On examination, she was mildly somnolent, but arousable to voice and fully oriented. Her neurologic examination was otherwise unremarkable. Notably, there were no cranial nerve abnormalities, no areas of focal weakness or sensory loss, and no ataxic signs.

Immediately after admission, she exhibited recurring, stereotyped spells for which she was transferred to the NNICU. These spells lasted approximately 30 s and were characterized by sudden unresponsiveness to stimulation, sonorous breathing, and gaze deviation to either the right or the left. Most strikingly, she exhibited classic fencer posturing with tonic extension of the right or left arm and concurrent contralateral arm flexion. Spells were followed by an immediate return to her mental status baseline.

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Given the patient's history of childhood seizures, her recent head trauma, and fencer posturing, we had a high suspicion that these spells represented epileptic events. We therefore monitored the patient on continuous video electroencephalography (EEG) for several days while anticonvulsant medications (levetiracetam, lacosamide, and lorazepam) were trialed. These interventions did not alter the frequency of observed spells, and none of her events were associated with clear epileptiform activity. Instead, diffuse delta range slowing was the most prominent electroencephalographic finding during spells (Fig. 1c).

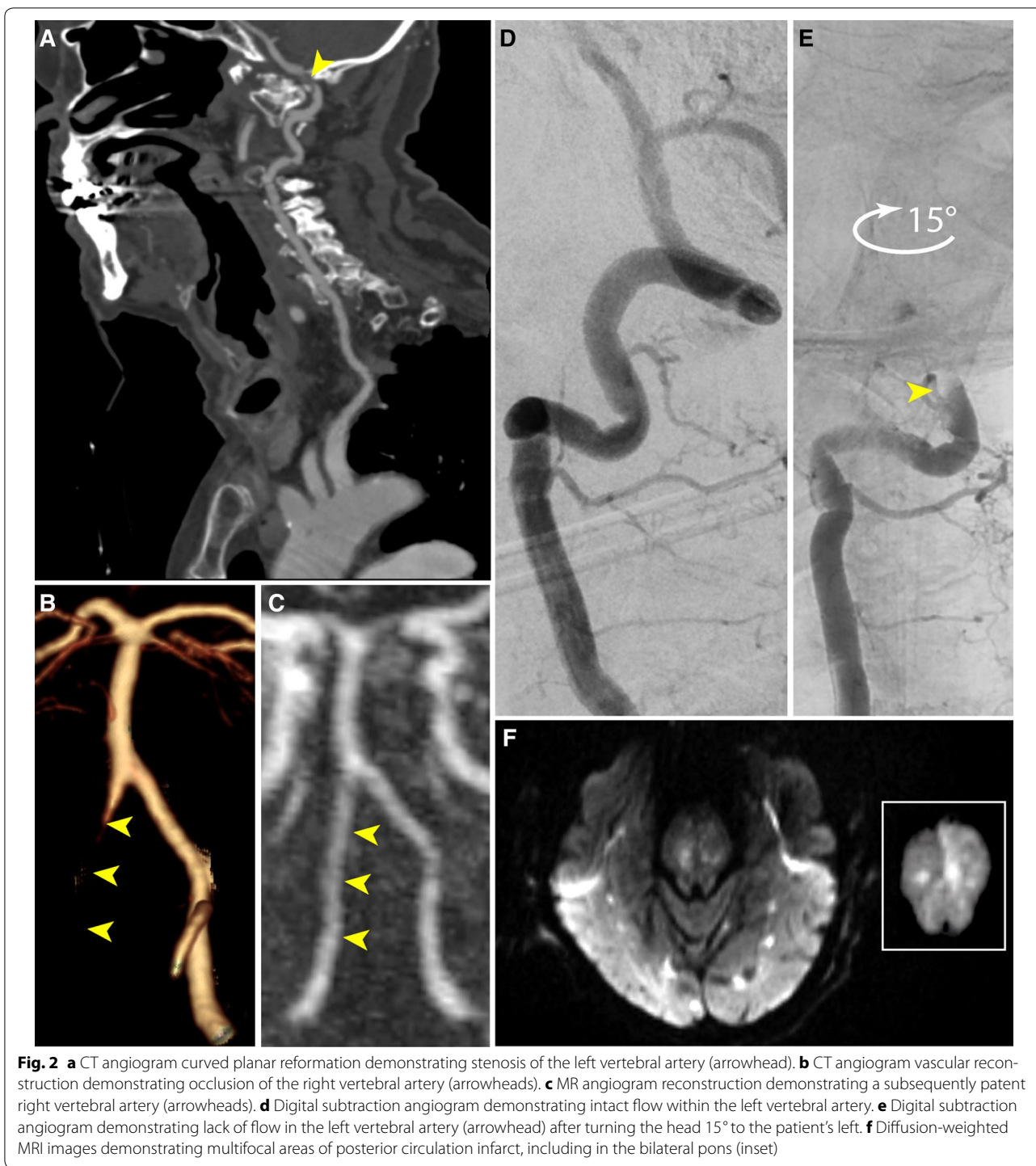
Coincident with her electrophysiological workup, vascular studies were performed. A CT angiogram (CTA) revealed a stenosis and likely dissection of the left vertebral artery (Fig. 2a), and occlusion of the right vertebral artery (Fig. 2b). Both the stenosis and occlusion of the vertebral arteries were near the C1 fracture. At this point, staff began noticing that spells seemed to be elicited with changes in position including bathing the patient or repositioning her in bed. An MRI with MR angiography demonstrated no evidence of stroke, but did demonstrate a patent right vertebral artery (previously occluded on CTA) as well as diminutive bilateral posterior communicating arteries (Fig. 2c). The initial occlusion of the right vertebral artery on CTA with subsequent resolution on MRA suggested intermittent cervical occlusion and raised the possibility of a dynamic vascular etiology for the patient's spells. She was briefly trialed on vasopressors

to a mean arterial pressure of 120 mmHg to optimize collateral perfusion in the setting of dynamic flow limitation. This also failed to mitigate her position-dependent spells.

Given the concern for a dynamic vascular etiology, catheter vertebral artery angiography was performed. Injection of the left vertebral artery initially demonstrated intact flow (Fig. 2d). Turning the patient's head 15° left during the procedure elicited a typical spell and demonstrated complete occlusion of the previously patent left vertebral artery (Fig. 2e; the right vertebral artery was not injected). This, along with an intermittently patent right vertebral artery, suggested a diagnosis of bilateral rotational vertebrobasilar insufficiency resulting in ischemic spells that mimicked a classic seizure semiology.

## Management

Because of her frequent and debilitating spells, the risk of brainstem stroke from dynamic vascular compression, and an unstable cervical spine, the patient underwent occipitocervical fusion. She was difficult to arouse following surgery, raising concern for intraoperative ischemia. A repeat brain MRI was performed which unfortunately demonstrated extensive posterior circulation strokes (Fig. 2f). She remained minimally arousable on mechanical ventilation in the ICU for the next several days. Given the patient's extensive brainstem injury and resultantly poor prognosis, in consultation with her family, care was redirected to prioritize comfort. She was subsequently discharged to hospice.



## Discussion

This patient's case presented an unusual diagnostic challenge. The differential diagnosis of transient neurological syndromes generally includes, at a first pass, seizure, ischemia, and migraine phenomena. Many other common conditions must also be considered including toxic

or metabolic abnormalities, alterations of intracranial pressure, cardiac dysrhythmias, vasovagal syndromes, orthostasis, and psychogenic syndromes. The stereotyped nature of this patient's spells, her sudden loss of responsiveness, and her gaze deviation were highly concerning for seizure. Additionally, fence positioning is classically



associated with frontal lobe seizures involving the supplementary sensorimotor area [6]. Her history of recent head trauma and childhood seizures further elevated this diagnosis on her differential. There were, however, also several indicators of a vascular etiology: First, the association of spells with a change in position suggested a dynamic, structural lesion. Second, the lack of a significant postictal period is inconsistent with most seizures. Ultimately, the absence of electrographic seizures on EEG during spells made a vascular etiology much more likely. Though preceding studies (CT and MR angiography) were concerning for a dynamic flow limitation, a conventional angiogram including head rotation was necessary to definitively diagnose Bow Hunter's Syndrome. When considering this case, it is interesting to note that both seizures [7, 8] and status epilepticus have been reported as presenting symptoms of basilar occlusion [9].

In addition to its diagnostic teaching points, this case is also novel and instructive in its etiology. Presentation following trauma is unusual, and this patient's symptoms were both more acute and more severe than is typically reported for Bow Hunter's Syndrome [2]. This may be due to the bilateral nature of this patient's transient vertebral artery occlusion and the minimal anterior-to-posterior collateral flow due to diminutive posterior communicating arteries, resulting in profound brainstem ischemia.

The patient's clinical course furthermore implicates fencer posturing as an important sign of brainstem ischemia. Fencer posturing, also known as the asymmetric tonic neck reflex, is a well described primitive reflex in infants as well as adults with neurodegenerative diseases [10]. Classic work in decerebrate cats demonstrated it to be a vestibular reflex [11]. Fencer posturing is also frequently observed following mild traumatic brain injury [12] and is hypothesized to be caused by transient neurovascular disruption of the vestibular nucleus in the ventral pons. It is tempting to hypothesize that similar transient disruptions repeatedly occurred in this patient following obstruction of blood flow to the ventral pons after positional vertebral artery occlusion. Abnormal limb movements [13, 14] as well as convulsions [15] have previously been reported following posterior circulation ischemia.

This case, to our knowledge, is the first report of Bow Hunter's Syndrome presenting with fencer posturing mimicking a seizure semiology. Diagnosis in neurology is susceptible to cognitive errors in the use of heuristics [16], and recognition of such errors is an important skill for neurocritical care trainees to gain. This case specifically provides an opportunity to consider framing error, anchoring error, and availability error (the relative frequency of seizures in the NNICU can make alternate diagnoses appear less likely than clinical data suggest).

Although this patient's diagnosis ultimately became apparent through testing, the case unfortunately serves to highlight the high risk of devastating stroke with lesions impacting the posterior circulation.

### Key Points

1. A unique case of bilateral rotational vertebrobasilar insufficiency (Bow Hunter's Syndrome) is presented.
2. The case had several elements which presented a significant diagnostic challenge:
  - The acute and severe nature of the patient's presentation after trauma
  - The likely involvement of bilateral vertebral arteries
  - Fencer posturing with loss of consciousness as a rotation-induced symptom
3. While MR and CT angiography aided in the diagnosis and helped rule out alternative diagnoses, dynamic catheter angiography was ultimately required to make a definitive diagnosis.

### Author's Contributions

Both authors contributed equally to the design, writing, and publication of this manuscript. Additionally, both authors directly cared for the patient described within the manuscript.

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### Compliance with Ethical Standards

### Conflict of interest

The authors declare that they have no conflict of interest.

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