From locked-in sensation to acute basilar artery occlusion: a rare case of posterior circulation stroke in a young patient

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Abstract

We report the case of a young male patient who presented to our emergency department complaining of a transient headache and a feeling of motor impairment in his right lower limb, both of which completely resolved spontaneously. After a few minutes of his arrival, he developed a locked-in sensation with anxiety and agitation, followed by a rapid onset of left facio-brachio-crural hemisyndrome. Contrast-enhanced CT of the brain and neck showed the occlusion of the proximal-intermediate basilar artery. Acute basilar artery occlusion (BAO) is a rare life-threatening stroke that requires prompt diagnosis and appropriate treatment to avoid rapid fatal complications (coma and death). The prognosis is poor, with long-lasting sequelae in the survivors. The diagnosis is often difficult since BAO accounts for only 1% of all strokes, and the prodromal symptoms are often very mild, transient, and non-specific, such as nausea, dizziness, headache, confusion, and vertigo. In the presence of convulsion-like symptoms or an unexplained altered level of consciousness, emergency physicians should always consider BAO in the differential diagnosis.

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Highlights

- *Basilar artery occlusion (BAO) is a very rare posterior circulation stroke and a true neuro-interventional emergency.*
- *The clinical presentation ranges from transient symptoms (nausea, dizziness, headache, confusion, and vertigo) to sudden and dramatic neurological impairment (hemiparesis/quadriparesis, aphasia, dysarthria, dysphagia, loss of consciousness, respiratory dysfunction, and coma).*
- *If not promptly recognized and early treated, BAO results in rapid deterioration with fatal consequences.*
- *The prognosis is poor: ~90% mortality depending on the location, and high morbidity in the survivors.*
- *Enhanced-contrast CT scan of the brain and neck is mandatory to diagnose BAO.*

Case Report

About four hours after returning from the gym, a 31-year-old male presented to the Emergency Department (ED) of Piacenza (Emilia-Romagna, Italy), complaining of a transient left frontal headache and a feeling of motor impairment in his right lower limb. At admission, he was completely asymptomatic with GCS of 15, blood pressure of 120/80 mmHg, heart rate of 60 bpm, oxygen saturation of 97% on room ambient, and a body temperature of 36°C. Venous blood gas analysis revealed normal pH, blood glucose level, serum electrolytes, and lactates. ECG and bedside ultrasound showed sinus rhythm and normal cardiac, pulmonary, and abdominal findings, respectively. Laboratory tests were all within the normal range, including negative urine toxicological analyses. He denied using anabolic-androgenic steroids as well as other substances and medications. His past medical history was unremark-

able. Familiar cases of sudden death, and neurological and/or cardiac diseases were not reported.

On neurological examination, he exhibits fluent speech, intact comprehension, autonomous ambulation with slight right leg entrainment, a negative Romberg test, cranial nerves uninjured, and a tendency to hollow the right hand (NIHSS score of 2). Continuous ECG monitoring was immediately started. After approximately five minutes, he complained of chest oppression with subjective dyspnea, peribuccal tingling, generalized tremors, and a locked-in sensation, followed by a rapid onset of left facio-brachio-crural hemisyndrome (NIHSS score of 8) with unchanged vital signs and normal sinus rhythm. Non-contrast brain CT scan showed a hyperdensity (12-13 mm) in the proximal-intermediate tract of basilar artery. The filling defect was confirmed on CT Angiography (CTA) and was consistent with acute thrombosis. Vertebral Arteries (VA) and P2 segments of the posterior cerebral arteries opacify normally. A diagnosis of Basilar Artery Occlusion (BAO) was done.

Soon after the CT scan, he appeared slowed and soporific with dysarthria, left facio-brachio-crural hemiparesis, hypoesthesia of the left hemisoma, and a bilateral Babinski sign (NIHSS score of 19). In the absence of contraindications, the patient was immediately treated with Intravenous Thrombolysis (IVT) with alteplase 0.9 mg/kg IV (10% of the total dose as an initial IV bolus over 1 minute and the remainder infused over 60 minutes) and transferred to Parma hospital (a tertiary care center), where he underwent angiography and mechanical thrombectomy using a stent retriever, which allowed a complete recanalization of the BA and a complete motor recovery with a residual mild dysarthria (NIHSS score 1) (Figure 1). After the procedure, MRI Diffusion Weighted Imaging (DWI) showed a small oval area of diffusion restriction on follow-up imaging at the left pontine tegmentum consistent with ischemic lesion (Figure 1H). Although angiography raised the suspicion of arterial dissection, the etiology remains undetermined after extensive diagnostic workup.

Discussion

The Basilar Artery (BA) is formed over the surface of the pons by two vertebral arteries to supply the critical areas of the brain and brainstem. Anatomically, it can be subdivided into three arbitrary segments: proximal from the VA to Anterior Inferior Cerebellar Arteries (AICA), middle from AICA to the origin of Superior Cerebellar Arteries (SCA), and finally, the distal segment from SCA to the terminal Posterior Cerebral Arteries (PCA). Basilar Artery infarct or Occlusion (BAO) is caused by the obliteration of blood supply to the posterior circulation or vertebrobasilar system of arteries.[1] BAO is a true neuro-interventional emergency, and if not treated early, brainstem infarction results in rapid deterioration until loss of consciousness and death.

Epidemiology

Posterior circulation stroke, which includes BAO, accounts for approximately 20% of all ischemic strokes. BAO is extremely rare (~1% of all strokes), with an unknown exact incidence.[1] In a population study including 129 patients with acute Large Vessel Occlusions (LVOs), the estimated incidence of BAO was 4/100

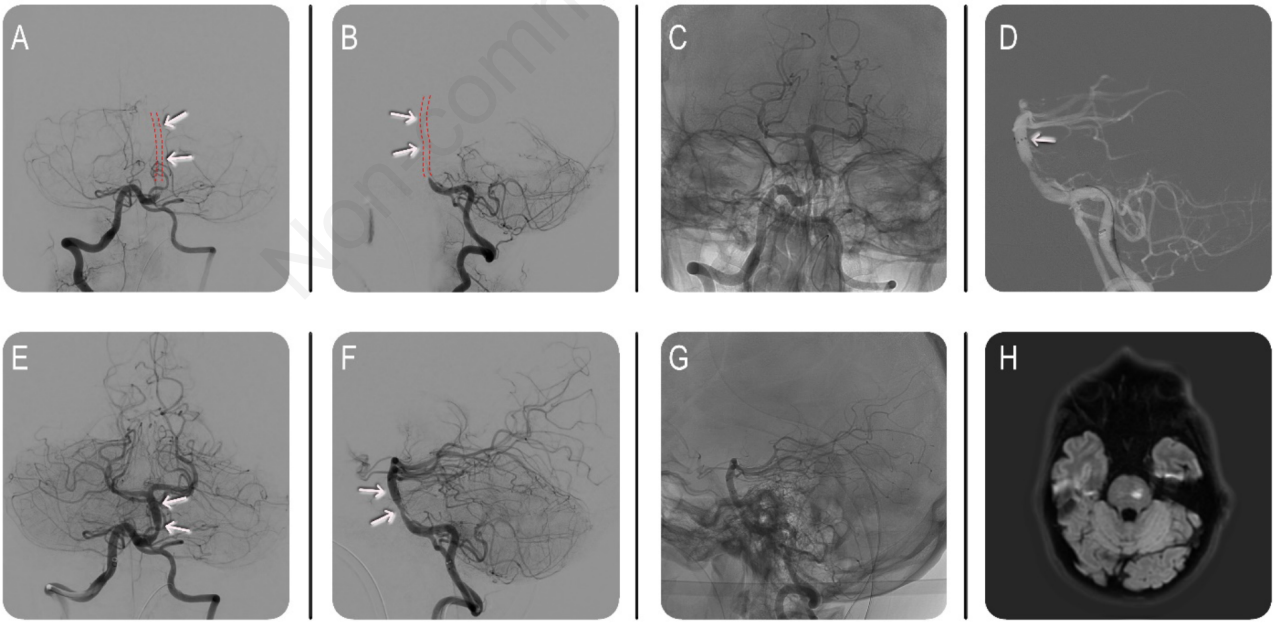

Figure 1. Digital subtraction angiography (DSA) demonstrating occlusion of the basilar artery (white arrows) before thrombectomy (red dotted lines describe the expected position of the basilar artery: **A**, frontal view; **B**, lateral view). After mechanical thrombectomy using a stent retriever, complete recanalization (white arrows) of the basilar artery (**E**, frontal view; **F** lateral view). In **C** and **G** the anatomical position of the recanalized basilar artery is shown superimposed on the skull image. The metal landmarks of the stent are visible, indicated with the white arrows in Figure **D**. MRI Diffusion Weighted Imaging (DWI) showed a small oval area of diffusion restriction on follow-up imaging at the left pontine tegmentum consistent with ischemic lesion (**H**).

000/year.[2] In the Basilar Artery International Cooperation Study (BASICS) registry, among 592 patients with BAO, the average age is 63 years, with females being 37%.[3]

Clinical presentation

BAO was first reported in 1828 by Scottish physician John Abercrombie, who described remarkable progressive bulbar symptoms, including dysphagia and speech difficulty.[4] Many authors in the 19th century described a waxing and waning clinical course for several days before profound coma and death. They also noticed signs and symptoms such as hemiplegia without loss of sensitivity and bulbar symptoms such as swallowing and speech impairment, vertigo, and altered consciousness.[4]

The clinical presentation of BAO ranges from very mild transient non-specific prodromal symptoms, such as nausea, dizziness, headache, confusion, and vertigo, to sudden and dramatic neurological impairment, due to devastating strokes with high mortality and morbidity. These symptoms include ipsilateral cranial nerve deficits, contralateral hemiparesis, sensory impairment, coordination deficits, quadriparesis, aphasia, dysarthria, dysphagia, loss of consciousness, coma, and cardiopulmonary compromise.[1,5]

The specific features will vary depending on the occlusion site, as follows: i) top of the basilar syndrome: visual and oculomotor deficits, behavioral abnormalities, confusion, somnolence,[6] hallucinations, dream-like behavior, and convulsions, motor dysfunction is often absent;[7] ii) proximal and mid portions of the BA (pons) can result in patients being "locked-in" with complete loss of movement (quadriparesis and lower cranial dysfunction) and respiratory muscle paralysis, but preserved consciousness and ocular movements (often only vertical gaze), as the oculomotor nerve is not affected.

Convulsion-like movement with rigidity and twitching can be an early symptom of BAO, which can easily be mistaken for epileptic seizures caused by compromised anterior circulation or cortical lesions.[7] In a single-center case series from 2015 to 2020, that investigated patients who underwent endovascular therapy for BAO, 21.9% (7/32 patients) presented with convulsive-like symptoms.[8]

Pathology

BAO can be due to either thromboembolism, atherosclerosis, or the propagation of intracranial dissection. Although these may occur anywhere, they have a predilection for different segments of the BA: i) vertebrobasilar junction: thromboembolism (e.g., cardioembolic), atherosclerosis with thrombosis, and propagation of VA dissection (rare); ii) midsegment: atherosclerosis with thrombosis; iii) distal third or basilar tip: thromboembolic (e.g., top of the basilar syndrome).

The most common causes are atherosclerosis (36%) and thromboembolism (35%) from LVO or the heart.[9] Dissection of BA is rare (5%). In the remaining 24% of the cases, the cause is undetermined, as reported in our case.[3]

Emboli usually arise from the heart or large arteries to cause occlusion of BA. In contrast, a thrombus may arise directly from the BA due to atherosclerosis to produce BAO or may propagate from a thrombus from a VA due to atherosclerosis or dissection. Both the proximal and middle segments often become occluded due to thrombi arising from bilateral VAs, and an embolus from VA can lodge directly into the distal section. The perfusion pressure drops due to the presence of an embolus in the proximal portion of the BA, causing a reversal of blood flow from bilateral PCAs, and this reflux can prevent an embolus from reaching the distal segment of the BA. The basilar syndrome is mainly caused by distal BAO and manifests as behavioral disturbance, confusion, oculomotor, and visual abnormalities, but often spares motor findings. The extent of thromboembolism can be a single-segment occlusion of BA or may involve all three segments of BA, and we can visualize this on vessel imaging, preferably CT angiography of the head and neck.

COVID-19 infection has been shown to worsen the prothrombotic state associated with pregnancy and produce BAO.[10,11] An "8-shaped" basilar artery fenestration malformation,[12] acute myelogenous leukemia,[13] and Crohn's disease[14] can also lead to BAO.

Diagnosis

When clinical findings suggest an acute brainstem disorder, BAO has to be confirmed or ruled out as a matter of urgency. If BAO is recognized early and confirmed with multimodal CT or MRI, intravenous thrombolysis or endovascular treatment can be undertaken. The goal of thrombolysis is to restore blood flow in the occluded artery and salvage brain tissue; however, the best treatment approach to improve clinical outcomes still needs to be ascertained.

On arrival at the ED, the emergency physicians should quickly gather information including blood pressure, blood glucose level, the time of Last Known Normal (LKN) or onset of stroke signs/symptoms, NIHSS score, medications like anticoagulants/antiplatelets, past medical and surgical history. The first imaging should be a CT head without contrast to rule out hemorrhagic stroke. If the CT head is negative for bleed and there is a concern for BAO, contrast-enhanced CT of the head and neck is mandatory to rule out an LVO. If not possible, brain MRI can be used to evaluate cerebral vessels.

Non-contrast CT shows hyperdense vessel is a sign of the BA, present in ~65%.[1] A high index of suspicion is needed in the correct clinical setting as the diagnosis can easily be missed (often only present on 1 or 2 slices); additionally, it is well recognized that acute clots are of lower attenuation than chronic clots.[15] Hypoattenuation delineates tissue with ischemic damage (beam-hardening artifacts limit visualization of the brainstem on CT). Contrast-enhanced CT fills defects within the vessel and distinguishes the ischemic penumbra area from an irreversibly damaged area (infarct core). Angiography remains the gold standard for the diagnosis of BAO, but it is used only after non-invasive imaging for therapeutic recanalization.[1] Images demonstrate a filling defect within the vessel. MRI shows loss of flow void within the BA on spin-echo and FLAIR images; restricted Diffusion Within Infarcted tissue (DWI); and hyperintense signal within infarcted tissue (T2/FLAIR). Transcranial Doppler is not routinely used in the emergency setting. It can show the absence of signal in the BA, and indirect signs, such as abnormal waveforms in the vertebral arteries and collateral flow.

Treatment and prognosis

BAO is a life-threatening event with a poor prognosis: ~90% mortality depending on the location, and high morbidity in the survivors.[16] The prognosis depends on the severity of the stroke, the time of LKN, the treatment offered, successful recanalization, and access to tertiary care hospitals and comprehensive stroke centers. Overall, mortality is very high: patients with successful recanalization after thrombectomy have a lower mortality rate of 33 to 50% versus 74 to 100% in patients without thrombectomy or failed reperfusion.[17] With early arrival and appropriate management, the outcome may be good, with minimum deficits. Late arrival and

failure of revascularization of BAO result in poor outcomes, including severe neurological deficits and very high mortality.

Multidisciplinary consensus for individualized management is difficult to achieve in a time-critical condition. Over time, improvements in prognostic scoring systems and neuroimaging have improved clinical decision-making. Recent randomized trials comparing Endovascular Therapy (EVT) and medical management for patients with BAO include BEST (Basilar Artery Occlusion Endovascular Intervention vs. Standard Medical Treatment), BASICS (Basilar Artery International Cooperation Study), BAOCHE (Basilar Artery Occlusion CHinese Endovascular Trial), and ATTENTION (Endovascular Treatment for Acute Basilar Artery Occlusion), compared EVT and medical management for patients with BAO. These trials yielded mixed results. ATTENTION and BAOCHE showed that EVT is beneficial for BAO within 24 hours of onset, with a therapeutic impact comparable to that of anterior circulation LVO ischemic stroke.[18,19]

If the patient has arrived at the ED within the alteplase (tPA) window (less than 3 to 4.5 hours since LKN) and has no contraindication to tPA, they should immediately receive tPA. If eligible, the patient should be transferred to the angiography suite for Mechanical Thrombectomy (MT) as soon as possible. Based on the BASICS trial, MT is effective if performed within 6 hours of the onset of symptoms.[3] MT with a clot retrieval device has been used in selected cases. Endovascular Reperfusion Therapy (EVT) improves recanalization rates in patients with emergent LVO.[19] Predictors of outcome after MT are the following.

Age and gender

Analysis of the BASICS randomized control trial reports no significant differences between age groups observed for recanalization rate and incidence of symptomatic intracranial hemorrhage. Patients aged 75 years or older with BAO have an increased risk of poor outcome compared with younger patients (18-54 years), but a substantial group of patients ≥ 75 years (22%) survive with a good functional outcome.[20] No significant gender differences in outcome and recanalization were observed, regardless of treatment modality (IVT, or combined IVT-intra-arterial thrombolysis (IAT), or IAT).[21]

Collateral flow

Several studies, including a series of 21 patients[19] and another of 104 patients carried out between 2010 and 2016,[22] have found that the presence of bilateral posterior communicating arteries on pretreatment CTA was associated with more favorable outcomes in BAO treated with EVT.

Vertebral artery stenosis

From the BASICS study, in patients with acute BAO, unilateral Vertebral Artery (VA) occlusion or stenosis ≥ 50% is frequent (66/141 patients, 47%), but not associated with an increased risk of poor outcome or death. The risk of death did not depend on the presence of unilateral or bilateral VA occlusion or stenosis. Patients with BAO and bilateral VA occlusion had a slightly increased risk of poor outcomes.[23]

Vertebrobasilar artery calcification

In a cohort study of 64 patients, vertebrobasilar artery calcification was found to be an independent predictor of outcome and associated with reduced functional independence and increased mortality.[24]

Posterior circulation Acute Stroke Prognosis Early CT score (pc-ASPECTS). An analysis of BASICS suggested that a cerebral blood volume pc-ASPECTS <8 may indicate patients with high case fatality. However, further evidence is needed as CTA was available in only 27/592 (5%) of BASICS patients.[25]

Differential diagnosis

BAO enters into differential diagnosis with basilar meningitis; basilar migraine; cerebellar infarct with brainstem compression and edema; drug-intoxication; encephalopathy; hemorrhagic stroke (cerebellar hemorrhage with brainstem compression, pontine hemorrhage, focal deficits due to brainstem bleeding from ruptured cerebral cavernous malformations, arteriovenous malformation, and fistulas); multiple sclerosis; seizures; and tumors in the posterior fossa, leading to mass effect and brainstem compression.[9]

Conclusions

BAO is a true neuro-interventional emergency since it is a severe and life-threatening stroke with a poor prognosis and a high risk of long-lasting disability. The real challenge for the emergency physician is the prompt diagnosis. In most cases, patients report unspecific and very mild symptoms, like headache, nausea, vertigo, and dizziness, which can cause delays in diagnosis and fatal consequences (coma and death). Emergency physicians should consider BAO when managing first-time generalized convulsive-like symptoms or in the presence of an unexplained reduced level of consciousness. With early arrival and proper management, the outcome may be good, with minimum deficits, as described in our case report. The key to properly managing and treating patients with suspected BAO is the cooperation of emergency physician, neurologist, neuroradiologist, and intensive care specialist.

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