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Letter to the Editor: New Observation

Vertebrobasilar Dolichoectasia: Case Report and Management Review in an Underappreciated Cause of Bulbar Palsy, Weakness, and Ataxia

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A 70-year-old right-handed woman presented with one year of imbalance and right-sided weakness. She had new difficulty opening lids, clumsiness and was dropping objects from her right hand. These changes were accompanied by progressive gait and balance difficulties. These changes were superimposed on a four-year history of progressive dysarthria, slurred speech, and vocal hoarseness. She endorsed coughing frequently with solid foods. There was no associated weight loss, fasciculations, muscle cramps, ptosis, diplopia, nor did she experience any fluctuation or fatiguability. There was no family history of myopathy, motor neuron disease, or inherited ataxias. Her exam revealed a spastic dysarthria, hoarseness, right upper and lower extremity pyramidal weakness (4+/5), and hyperreflexia, along with a right extensor plantar response. She had tandem gait difficulties and left-sided dysmetria with dysdiadochokinesia.

An MRI of the brain showed compressive vertebrobasilar dolichoectasia (VBD) at the medulla and left inferior cerebellar peduncle (Figure 1). Neurosurgical consultation was arranged, and diffusion tensor imaging (DTI) (Figure 2) was obtained which demonstrated displacement of the medullary pyramidal tracts. Neurosurgical decompression was offered. The patient decided to be followed and managed conservatively. Now, four years later, she remains clinically and radiographically stable.

VBD has been reported as a rare cause of progressive bulbar palsy and ataxia and could misleadingly present clinically like many other neurological disorders.[1,2] VBD, defined as a basilar artery diameter >4.5 mm, is a dilatative arteriopathy characterized by progressive arterial dilation from underlying rarefaction of connective tissue within the tunica media and fragmentation of the internal elastic lamina.[2] There are many causes of dolichoectasia including vascular risk factors, aberrant matrix metalloproteinases, connective tissue disorders, and, rarely, lysosomal storage disorders.[2] VBD portends an increased risk of ischemic and hemorrhagic stroke, dissection, and arterial rupture.[2] A variety of clinical presentations can arise from compressive VBD including cranial neuralgias, hydrocephalus, bulbar paresis, ataxia, and paresis.[2]

An increased mortality risk with poor short-term survival has been reported with VBD.[2] A review including 375 patients reported the following 5-year risks: ischemic stroke 17.6%, brainstem compression 10.3%, hemorrhagic stroke 4.7%, hydrocephalus 3.3%, and subarachnoid hemorrhage 2.6%.[3] Another study reported a 5-year case fatality rate of 36.2% (95% CI: 30.6–41.85).[4] A better prognosis is more likely in those without symptoms at the time of diagnosis.[3] Over an average follow-up period of 11.7 years, those with any compressive symptoms at diagnosis had 6.7 times greater odds of any clinical event occurring and 2.4 times greater odds of stroke compared to those without any symptoms at diagnosis. On neuroimaging, increased basilar artery diameter, height of the basilar artery bifurcation, and degree of lateral displacement were all independently associated with ectasia progression.[4] Patients with progressive ectasia have an increased odds of both stroke and death.[4] The proportion of VBD survivors without clinical events was 54.1% at 5 years, 39.5% at 10 years, and 23.5% at 15 years.[4]

Unfortunately, there is weak evidence to guide medical management of this patient population. The most recent Canadian Stroke Best Practice Recommendations do not provide specific guidance for VBD management.[5] Given that patients with VBD are at risk of ischemic and hemorrhagic stroke as well as subarachnoid hemorrhage, it is difficult to know if antiplatelet therapies confer benefit or harm. Some cohort studies describe patients on antiplatelet agents and anticoagulation,[4] but none were designed to provide an unbiased estimate of the potential magnitude of ischemic stroke risk reduction compared to an increase in hemorrhage risk.[3] In one observational cohort study, 79 patients were treated with either an antiplatelet or anticoagulant while 77 remained untreated.[4] On follow-up, 42% of treated patients had ischemic stroke while 34% of untreated patients had ischemic stroke (a nonstatistically significant difference).[4] This was not a

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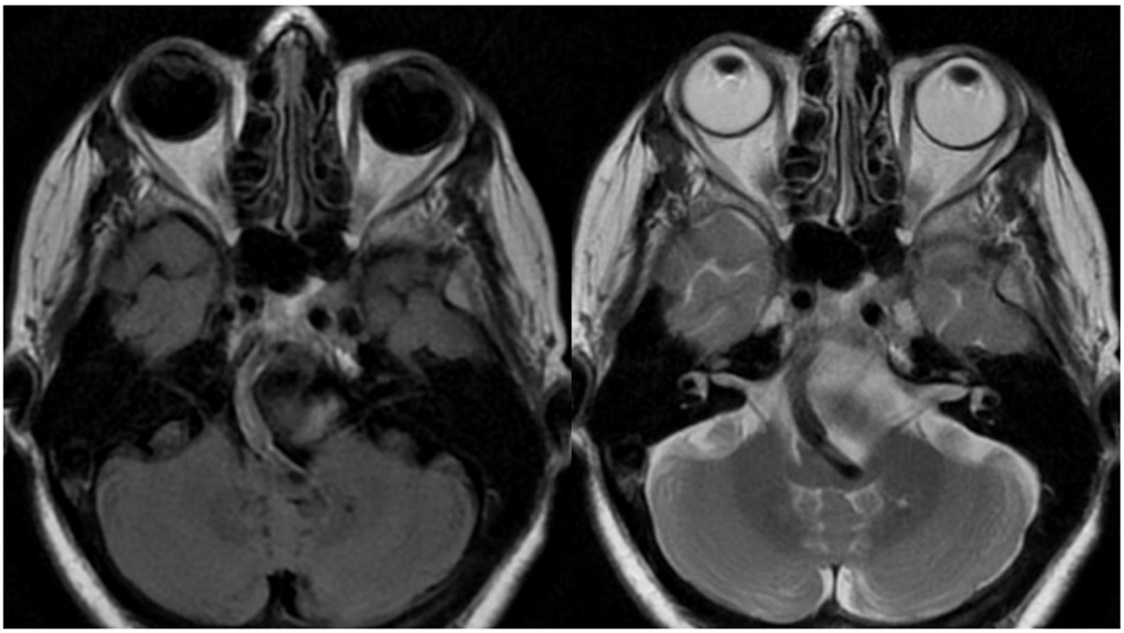

Figure 1: 1.5 T MRI Brain. Fluid attenuated inversion recovery (left) and T2 weighted image (right) demonstrating vertebrobasilar dolichoectasia causing left medullary and inferior cerebellar peduncle compression.

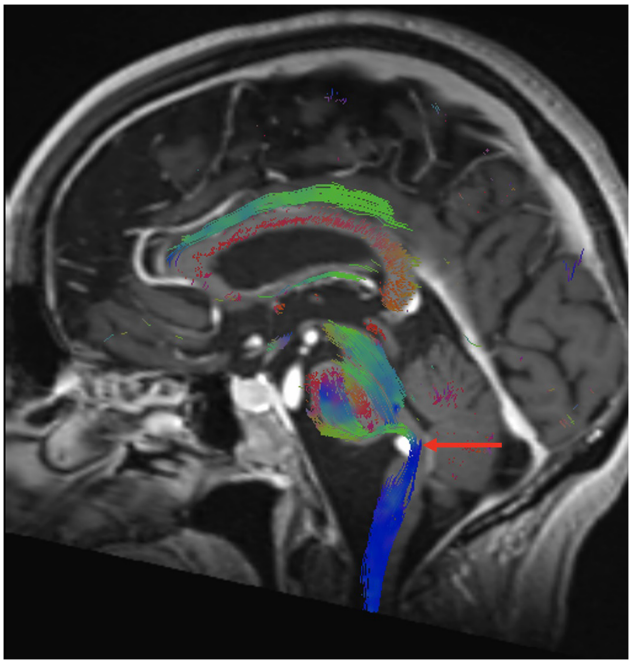

Figure 2: Diffusion Tensor Imaging demonstrating displacement of pyramidal tracts and other ascending/descending fibers at the level of the medulla indicated by the red arrow.

randomized trial and the majority of those treated with antiplatelet or anticoagulant had prior ischemic stroke, so the treatment group may have had higher risk to begin with. Another prospective cohort study in 156 consecutive patients with VBD were followed 9.35 years and 32 hemorrhagic strokes occurred over the study duration.[6] While basilar artery diameter, hypertension, and female sex increased the odds of hemorrhagic stroke 4–6 times, the use of anti-platelets or anticoagulants was associated with a three times greater odds of hemorrhagic stroke.[6] While there is clinical equipoise for the optimal medical management of VBD, stroke clinicians should always pursue the best optimization of vascular risk factors, especially hypertension and smoking.[5]

A reasonable treatment approach, based on expert consensus, has been proposed by Pico et al, advising a genetic and metabolic workup in those <50 years of age and optimization of vascular risk factors and screening for abdominal aortic aneurysm in those ≥50 years of age.[2] The authors also consider an annual brain MRI/MRA or sooner if new symptoms arise. Consideration of surgery or endovascular treatment is suggested for progressive enlargement ≥2 mm or a basilar artery diameter ≥10 mm (2) or in those who have saccular aneurysm or rupture.[7] While a repositioning technique to achieve decompression in those with compressive symptoms from VBD has been reported to be effective,[8] it is not without risk and stenting reconstruction for compressive symptoms has uncertain benefit.[7] There is no guiding endovascular or surgical randomized control trial.

In the absence of strong evidence to guide management, we obtained DTI to help characterize the degree of tract displacement in the brainstem (Figure 2) and to aid with potential surgical planning. Ultimately, we did offer surgery given her symptoms and the degree of displacement noted on the DTI. Comparing the baseline DTI to a post-operative DTI may have facilitated improved assessment of the extent of post operative decompression of the brainstem. However, the patient was not inclined to take on surgical risk and over the past four years with conservative management of vascular risk factors and monitoring she has done well. Monitoring and optimizing vascular risk factors may also, therefore, be a reasonable approach to management of VBD in some instances without high-risk features.

This case offers a novel contribution to the literature. First, clinical case presentations with progressive bulbar symptoms in this age demographic more commonly evoke a very different set

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of neurologic differential diagnoses (neoplastic, motor neuron, neuromuscular, and inherited disorders) and recognizing VBD as an uncommon cause of this presentation is important for those practicing in the clinical neurosciences. Second, the use of DTI was a novel way to objectively demonstrate the location and extent of brainstem tract displacement. Furthermore, our review of the literature supports that neurosurgical intervention can be offered to those with progressive symptoms given a higher risk of morbidity and mortality. Our patient declined surgery and she was followed for several years with demonstrated clinical stability. Given the degree of clinical equipoise in the literature to date, there is a need for clinical trials to rigorously evaluate the medical and surgical management of VBD in the future.

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