A Case of Twists and Turns: a report of Vertebrobasilar Dolichoectasia

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Introduction

• Dolichoectasia is diagnosed using Smoker’s criteria which requires both ectasia (diameter greater than 4.5 mm) and elongation of arteries
• Incidence is less than 0.05%
• Twisting of the arterial branches of the vertebrobasilar arteries due to dolichoectasia decreases blood flow and causes ischemic symptoms
• The most commonly affected cranial nerve is the facial nerve, followed by the trigeminal nerve
• Associated with congenital diseases such as Marfan’s Syndrome or autosomal recessive polycystic kidney disease, as well as infections such as syphilis or expression of abnormal matrix metalloproteinases
• Risk factors include male sex, age, history of smoking, hypertension, and previous strokes

Case Presentation

Subjective Presentation

• 77-year-old male with a past medical history of Parkinson’s Disease, essential hypertension, hyperlipidemia, atrial fibrillation, and a previous basilar cerebrovascular accident approximately 15 years prior
• Presented with five days of left-sided headache, blurred vision, and difficulty swallowing
• Inability to sleep the night before prompted presentation
• Pain was intermittent and stabbing for a few seconds followed by a persistent, residual dull pain, not worsened with chewing but with movement, particularly with standing
• 12 point review of systems was negative

Objective Presentation

• Vital signs normal
• Pleasant, appropriate-for-age male, in a moderate amount of distress. He was alert and oriented to person, place, time and situation. The heart, lung, and abdominal exams were normal. The patient’s pupils were symmetric and reactive, extraocular movements intact. He had a symmetric smile with globally intact sensation. The ear canals were clear, with translucent tympanic membranes and no air fluid level. The left external pinna was tender to manipulation
• Platelet count of 83 mCL, CRP <0.5 mg/DL, ESR 1 mm/hour. All other laboratory values normal
• MRI and MRA with contrast revealed vertebrobasilar dolichoectasia with mild compressive deformity of the left ventral aspect of brainstem resulting in mild compressive deformity

Treatment

• Pain control: ketorolac, tramadol, gabapentin and morphine with minimal success
• Neurology and Neurosurgery consultants recommended an attempt at medical management and noted no acute or emergent surgical intervention
• Limited trial of dexamethasone 10 mg once followed by two mg tab every eight hours for one week and carbamazepine 200 mg twice a day and tramadol 50 mg every six hours, as needed for pain
• Outpatient follow-up revealed no improvement in pain or neurologic symptoms. Patient has an upcoming appointment at an outside academic facility for a cranial decompressive surgical evaluation

Discussion

• There are currently no guidelines concerning the management of VBD, nor is there evidence of a definitive cure
• Medical management of symptoms is first line therapy, however larger sized vascular ectasia increases adverse outcomes, including risk of hemorrhage
• Even with optimal medical management, 43% of diagnosed VBD patients have disease progression including worsening symptoms such as headache, nerve compression symptoms, or stroke, which is the most common cause of death
• If a patient fails traditional medical therapies, surgical options are microvascular decompression and gamma knife radiosurgery
• Microvascular decompression has decreased long term post-operative pain compared to gamma knife
• Both surgeries have a low success rate
• The most common complication is neighboring cranial nerve compression, which restarts the treatment plan but with a new cranial nerve involved

Conclusion

• While we do not know what the long-term outcome was for this patient, the diagnosis of this disease was important
• He did not respond to traditional medical management and ultimately required surgical evaluation
• The five-year mortality for patients with VBD is 36.3%
• An understanding of risk factors, physiology and pathology and curative treatments is important and necessary to prolong life in these individuals

References


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