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Case Report

Dolichoectasia of the vertebral basilar and internal carotid arteries: A case report and literature review

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Abstract

Background: Dolichoectasia is a rare disorder of the cerebral vasculature consisting of vascular elongation, widening, and tortuosity, usually involving the vertebral and basilar arteries. Its neurological symptoms and signs are highly variable.

Case Description: We present a case of dolichoectasia of the vertebrobasilar system in a patient with a long standing history of multiple falls. Repeat neuroimaging revealed an increase in size of the dolichoectatic segment. In addition, a new fusiform dilatation of the contralateral petrous segment of the internal carotid artery and isolated ventriculomegaly had developed.

Conclusion: Vertebrobasilar dolichoectasia can cause multiple clinical manifestations, with hydrocephalus being less common. In addition, having dolichoectasia of both posterior and anterior circulation is extremely rare.

Key Words: Carotid artery, hydrocephalus, vertebrobasilar dolichoectasia

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INTRODUCTION

Vertebrobasilar dolichoectasia (VBD) is an uncommon but well recognized vascular anomaly. It is asymptomatic in 90%.[37] When symptoms are present, they can be divided into: Ischemic, hemorrhagic, and mass effect. Reported manifestations include cerebellar dysfunction, ischemic stroke, trigeminal neuralgia, and brainstem compression syndrome.[24,47,48]

A review of the literature revealed only six other cases of VBD as a cause of hydrocephalus,[17] two of them with symptoms.[20,41]

CASE REPORT

A 67-year-old male with a known dolichoectasia of his vertebrobasilar artery, presented with a history of increasing falls over the past 3 months. A computed tomography (CT) scan of the head [Figure 1] and magnetic resonance imaging (MRI) of the brain [Figure 2] demonstrated a VBD with formation of a focal fusiform aneurysm in the proximal basilar artery. This caused mass effect on the left anterior aspect of the adjacent pons.

A magnetic resonance (MR) angiogram [Figure 3] revealed fusiform dilatation of the intradural distal right vertebral artery giving rise to the dysplastic dilated basilar artery, which had increased in size. The lumen was within a larger partially thrombosed vessel. This gave rise to a short segment of normal caliber basilar artery, which then again dilated just proximal to the basilar tip. There was also a new fusiform dilatation of the posterior cavernous left internal carotid artery consistent with a fusiform aneurysm. There was presence of ventriculomegaly of the lateral and 3rd ventricle, with the 4th ventricle being collapsed.

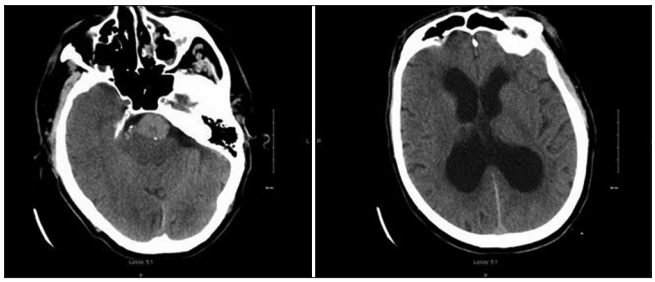

Figure 1: Axial CT brain scan showing dilatation of the vertebrobasilar system. Ventriculomegaly is appreciated as well

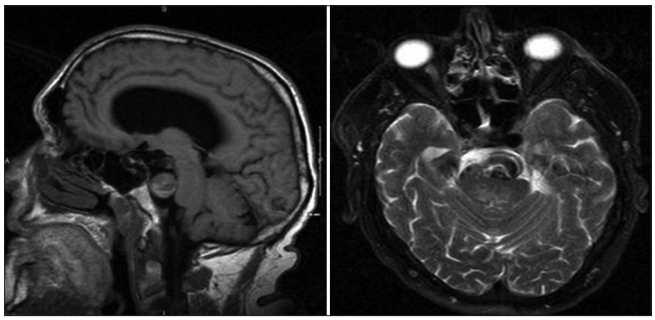

Figure 2: (Left to right) Sagittal T1-weighted, axial T2-weighted brain MRI showing right-sided vertebral artery dolichoectasia compressing the brainstem

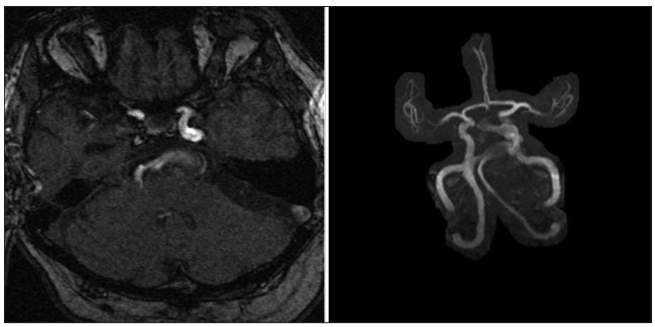

Figure 3: Axial Magnetic Resonance angiography of the brain showing dilatation of the vertebrobasilar artery and the internal carotid arteries

His past medical history included: Hypertension, dyslipidemia, long time smoker, chronic obstructive pulmonary disease, previous transient ischemic attack, depression, and prostate cancer treated with radiotherapy and NSTEMI treated with PCI (two bare metal stents).

Additional investigation

The presence of new ventriculomegaly on imaging raised the possibility of symptomatic hydrocephalus. A radionuclide cerebrospinal fluid (CSF) flow study was undertaken. It revealed normal migration of the radio tracer from the subarachnoid lumbar space to the basal cisterns and sylvian fissures, with no activity within the lateral ventricles in the 24 and 48 hour images. However, the patient also underwent a trial of lumbar drainage, during which his gait was assessed by the medical allied health team documenting an objective improvement in his gait. Consequently, the patient had a ventriculoperitoneal (VP) shunt inserted with no complications.

DISCUSSION

VBD is a rare dilatative arteriopathy that is defined as elongation or widening of the intracranial vertebral and or basilar arteries. The prevalence ranges from 0.06% to 5.8%.[8,37,50] It affects preferentially the vertebral and basilar arteries.[14] Involvement of both anterior and posterior circulation is rare.[44] It is also known as megadolichobasilar anomaly, basilar ectasia, or even as tortuous vertebrobasilar system. The prevalence of dolichoectasia increases with age, with the age of onset greater than 40 years and a male predominance.[7,8,15,25,50]

Pathogenesis

The pathogenesis of intracranial arterial dolichoectasia is unclear. Multiple pathophysiological processes might contribute to the development of such arterial ectasia vessels such as systemic arterial hypertension associated with atherosclerosis.[28,43] However, histological studies support the hypothesis that degeneration of the internal elastic lamina and thinning of the media secondary to smooth muscle atrophy is at the basis of this pathology[12,13,15,40] as well as prolonged systemic hypertension.[40] As such, many authors maintain that this dysfunction seems to be independent of atherosclerosis.[29,47,48] This was supported by the fact that atherosclerosis mainly involves the intima and endothelia of larger and medium size vessels, while dilatative arteriopathy involved mainly the intima of intracranial arteries.[21]

Symptoms

Most cases are asymptomatic. However, the tortuous dolichoectatic vertebrobasilar artery can produce pulsatile compression on the brainstem that may produce the syndromes of obstructive hydrocephalus, cerebellar dysfunction, or even trigeminal neuralgia.[31] As such, VBD may presents with symptoms due to (1) direct compression on cranial nerves causing cranial neuropathy, (2) direct compression on brainstem, (3) acute ischemia in the vertebrobasilar arterial territory[6,9,24,25,29,37,42,50] and (4) fatal outcomes due to vascular rupture.[20] Hydrocephalus is a rare complication and can cause compression either at the level the foramen of Monroe, 3rd ventricle, or cerebral aqueduct directly.[1,5,38,41,46]

Diagnosis

Some suggest the diagnosis of VBD as a clinical one of posterior circulation dysfunction.[48] Although VBD was first recognized on catheter angiography,[21] which still remains the gold standard for imaging of the cerebral

vasculature, other noninvasive modalities have emerged as adequate for many clinical situations. While the MRI sensitivity and specificity for VBD is unknown,[27] such noninvasive imaging provides invaluable information without the associated risks of a more invasive procedure.[31] It has now even become the method of choice in diagnosing VBD,[19,45,49] since it may also assist in demonstrating other potential causes of clinical symptoms.

Radiographic criteria for VBD are vertebral or basilar artery (1) diameter > 4.5 mm in any location along its course, (2) lateral deviation > 10 mm perpendicular to a straight line joining its origin to its bifurcation, (3) origin at the level of the pontomedullary junction, (4) bifurcation above the suprasellar cistern, (5) lateral to the margin of the clivus or dorsum sellae, and (6) basilar length > 29.5 mm or intracranial vertebral artery length > 23.5mm.[48]

Prognosis and treatment

The long-term prognosis of VBD is mostly associated with the severity of the condition at diagnosis and on its evolution characteristics, which was associated with a higher mortality and morbidity.[1,29] This was reviewed by a prospective study where 48% patients developed a stroke, 20% developed new compressive symptoms and 1% had hydrocephalus.[36] There is still uncertainty in the optimal treatment of VBD, as there has been no systematic review or long-term results from the different surgical interventions such as neurovascular decompression technique.[31]

The first surgical treatment reported in the literature for VBD is that of occlusion of vertebral artery.[26] This was followed by using microvascular repositioning.[31] Direct surgery to this lesion is difficult, as such there are few reports of surgical treatment. The first surgical report describes a patient with an enlarged, tortuous basilar artery, relatively small right vertebral artery, and a turbulent flow in the dilated vertebrobasilar junction caused by blood supply from the right vertebral artery.[23] As such, the patient underwent a wide suboccipital craniectomy for posterior fossa decompression, and proximal ligation of the right vertebral artery resulted in reduction of turbulent flow. Postoperative, the patient had a reduction of his preoperative symptoms, although incomplete. Another series study by Anson *et al.*[2] looked at the clinical characteristics and surgical treatments of dolichoectatic and fusiform aneurysms. Various surgical procedures were performed, including direct clipping, trapping with bypass, proximal occlusion, resection with reanastomosis, transposition, aneurysmorrhaphy with thrombectomy, and wrapping. Overall, their outcome at late follow up was good based on the Glasgow Outcome Scale scores of 1-2 in 78% of patients. This demonstrated that there still remains no consensus as to the optimal surgical treatment, and the benefits of surgery are still controversial.

Recently treatment with the use of coil-assisted stent reconstruction has been explored to determine the feasibility and long-term effectiveness in preventing ischemic/infarction events. There have been numerous reports of trials with different kinds of stents and coils with good results.[3,10,16,51] As such, endovascular reconstruction by using coil-assisted stent placement techniques or stent placement alone has been shown to be a safe and effective treatment for VBD.

In addition, medical therapies used to treat this condition have not been systematically evaluated.[14] There are studies that have determined that arterial hypertension not only plays a role in the formation and enlargement of intracranial dolichoectasia, but also contributes to the increased incidence of both ischemic stroke and intracranial hemorrhage.[30,33] Nonetheless there are no guidelines as to the blood pressure management that should be applied.

Since the greatest mortality of VBDE is associated with ischemic events, it would be wise to consider anticoagulation as a primary treatment. However, there are a few dilemmas regarding anticoagulation that one must consider before. Cerebral infarction associated with VBDE is caused by luminal thrombi that obstruct arterial branches, and this is different from the pathophysiology associated with atherosclerosis and cerebral aneurysm. In addition, there is a clear difference in prevalence and prognosis when comparing intracranial dolichoectasia to atherosclerosis and aneurysm.[11,22] More importantly, the pathophysiology of VBDE is that of an arteriopathy rather than atherosclerosis, and as such the response to thrombolytic or anticoagulant therapy is not as efficient as atherosclerotic or embolic infarction[8,18,30] and it would increase the potential of VBD rupture.[11,35] Hence, despite extensive studies in the pathophysiology of VBDE, related vascular risk factors, and the relation to systemic arterial disease,[32-34] the use of conventional anticoagulation in the treatment of VBD is still unclear.

Dolichoectasia most frequently involves the vertebrobasilar arteries, and or basilar arteries.[50] While the anterior circulation may be affected as well, involvement of both the vertebrobasilar and carotid system is rare.[4,39] In our case, we report a rare case of dolichoectasia of both anterior and posterior circulation associated with noncommunicating hydrocephalus.

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