Vertebrobasilar Fusiform Aneurysms

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KEYWORDS

• Aneurysm • Vertebrobasilar • Vertebral • Fusiform • Dolichoectatic

KEY POINTS

- Vertebrobasilar aneurysms present most commonly with brainstem ischemic stroke or compressive symptoms of the brainstem, cerebellum, or cranial nerves.
- The natural history of patients with fusiform vertebrobasilar aneurysms presenting with ischemic stroke or compressive symptoms is for the presenting signs and symptoms to steadily progress.
- Flow reduction or flow reversal is the most time-tested management strategy for fusiform vertebrobasilar aneurysms presenting with compressive symptoms, but this treatment relies largely on the presence of adequate collateralization through the posterior communicating arteries.
- Mixed results have been reported with the use of flow diversion for fusiform vertebrobasilar aneurysms, and their use should be reserved for patients with compressive symptoms and poor collateralization through the posterior communicating arteries.
- Patients with vertebrobasilar aneurysms presenting with ischemic stroke and no compressive symptoms are best managed with anticoagulation.

INTRODUCTION

Vertebrobasilar fusiform aneurysms are among the most daunting lesions treated by cerebrovascular surgeons. Many names have been linked to these lesions, including giant serpentine aneurysm, giant fusiform aneurysm, S aneurysm, megadolichobasilar artery, dolichoectacic artery, fusiform aneurysm, and transitional aneurysm. Perhaps the most universal definition of these lesions is aneurysms with separate inflow and outflow ostia. The earliest descriptions of fusiform aneurysms of the vertebrobasilar arterial system were most consistent with a dolichoectatic basilar artery. There is no arteriography available for most cases, and most patients presented with cranial neuropathies.[1]

A fusiform aneurysm of the basilar artery was first described by Wells in 1922[2] on surgical exploration in a patient with paresis of cranial nerves 6 to 8 and obstructive hydrocephalus. Dr Walter Dandy[3] operated on a series of 10 patients with trigeminal neuralgia and described a so-called S aneurysm. Greitz and Lofstedt[4] in 1954 reported 5 cases of ectasia of the basilar artery. The patients of their series were more consistent with compressive or ischemic presentations that are associated with fusiform aneurysms. Reports since this time have drastically expanded our understanding of these lesions. In this article, fusiform vertebrobasilar aneurysms are reviewed, including incidence, presentation, natural history, pathophysiology, and treatment, with a suggested algorithm for treatment based on review of the literature. Dissecting aneurysms, which have a distinct behavior of presenting with subarachnoid

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hemorrhage (SAH) followed by frequent early rebleeding, are not discussed.[5–9]

INCIDENCE

The incidence of fusiform aneurysms of the vertebrobasilar system is low. Dolichoectasia of any intracranial artery in the general population is estimated at less than 0.05%.[10] In cases of vertebral angiography, the incidence ranges from 17 of 10,000 (0.17%) for all indications to 132 of 2265 (5.8%) in cases of stroke.[11,12] Another series of 387 patients undergoing computed tomography (CT) or magnetic resonance (MR) angiography (MRA) for stroke showed 10 patients (2.6%) with vertebrobasilar dolichoectasia.[13] Autopsy series also show a wide range, but remain low in incidence, ranging from 6 of 5762 (0.10%) in a series from 1914 to 1956 at Columbia University[14] to 5 of 7500 (0.07%) in a VA hospital series.[15] One clinical series of treated posterior circulation aneurysms[16] classified 4 of 528 (0.76%) aneurysms as fusiform vertebrobasilar aneurysms.

The discrepancy in incidence in these studies is likely related to a combination of the loose definitions of these lesions and subselection. In addition, the incidence may increase in high-risk populations. Yu and colleagues[11] found a strong correlation with hypertension (64%) and tobacco smoking (74%) with intracranial arterial ectasia. In addition, Mitsias and Levine[17] described a high incidence in Fabry disease, with symptomatic disease of a dilated vertebrobasilar system found in 60% of heterozygotes and 67% of homozygotes. Conversely, vertebrobasilar aneurysms have been reported in children but are rare.[18–20] In 3000 postmortem brain examinations in children, Housepian and Pool[14] found no case of any intracranial aneurysm.

PRESENTATION

Because of the low incidence of these lesions and the relative paucity of reported cases, initial reports focused primarily on the striking pathologic features and rarely provided adequate clinical details.[1] However, a review of published series and case reports shows some common features. Unlike saccular aneurysms, vertebrobasilar fusiform aneurysms show a significant male predominance, accounting for greater than 70% of the 408 reported cases (**Tables 1** and **2**). Although affected patients ranged from age 5 years to 87 years, the average reported age at diagnosis was ~60 years, significantly older than that typically presenting with saccular aneurysms.[21] Although not often reported, commonly encountered comorbidities include hypertension (31%–69%), diabetes mellitus (10%–15%), hyperlipidemia (40%), coronary artery disease (23%–28%), and smoking (50%).[21,28,31] Data regarding connective tissue disorders were almost universally absent, an oddity given the mechanisms underlying vessel ectasia and the known importance of family history in the risk of aneurysm formation.[39,40] This subject was addressed only in the well-studied Mayo Clinic cohort, in whom 4% were found to have a known connective tissue disorder, including Fabry disease and autosomal-dominant polycystic kidney disease.[21,41,42]

In addition to the small subset of patients in whom vertebrobasilar fusiform aneurysms were incidentally diagnosed, the presenting symptoms in most patients are related to 3 basic mechanisms: mass effect, ischemia, or aneurysmal rupture. Mass effect, which was observed in 43% of patients (see **Table 1**), occurred when the ectatic vessel compressed surrounding tissues, including the brainstem and cerebellum, resulting in numerous cranial nerve palsies or in noncommunicating hydrocephalus, and typically developed over years.[1,28] In our experience, even although the clinical course is characteristically slowly progressive, it is often punctuated by stuttering episodes of abrupt exacerbation, typically as an early manifestation but also as a late preterminal manifestation. These episodes often correspond to intramural hemorrhage or microdissection evident on brain MR imaging (MRI) studies (**Fig. 1**).

When discussing cranial neuropathies caused by vertebrobasilar fusiform aneurysms, the most common cranial nerves involved are V to VIII.[21] Nishizaki and colleagues,[25] for example, reported dysfunction affecting the facial nerve in 4 of 6 patients, whereas in Herpers and colleagues' study,[26] hemifacial spasm accounted for 22% of patients presenting with compressive cause. Similarly, trigeminal neuralgia and abducens nerve palsy were frequently implicated as clinical correlates of brainstem compression.[1,26–29] Defects involving other cranial nerves, including cranial nerves IX, X, and XII, were more rarely described.[25,33] Although varying by aneurysm location, obstructive hydrocephalus was consistently reported as a sequela of brainstem compression.[21,23,28] In many of these patients, headache was a principal presenting symptom.[28]

Perhaps the most common presentation of patients with vertebrobasilar fusiform aneurysms involves ischemic stroke symptoms. Accounting for ~44% of patients (see **Table 1**), this subgroup comprises clinical syndromes ranging from transient ischemic attacks (TIAs) to catastrophic pontine ischemia consistent with a locked-in