Fusiform and dolichoectatic aneurysms

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Introduction

This article includes discussion of fusiform and dolichoectatic aneurysms, nonsaccular aneurysms, dilatative arteriopathy, basilar artery ectasia, s-aneurysm, and tortuous basilar artery. The foregoing terms may include synonyms, similar disorders, variations in usage, and abbreviations.

Overview

In this updated article, the author reviews current knowledge about nonsaccular intracranial aneurysms, including fusiform and dolichoectatic intracranial aneurysms. Fusiform and dolichoectatic aneurysms occur in any of the intracranial arteries, but particularly in the vertebrobasilar and internal carotid arteries. They present in a wide variety of ways, ranging from hemorrhage to cranial nerve or brain parenchymal compression to embolic stroke. Treatment of these life-threatening aneurysms must be individualized and can be challenging. Surgical approaches are sometimes feasible. Increasingly, novel endovascular techniques, including placement of flow-diverting stents and stent-assisted coiling, can successfully treat these aneurysms.

Key points

- Fusiform and dolichoectatic (nonsaccular) aneurysms can present with hemorrhage, ischemia, or symptoms related to compression of cranial nerves or brain structures, but are also often asymptomatic and detected incidentally.
- Fusiform and dolichoectatic aneurysms can occur from a variety of etiologies, including atherosclerosis, arteritis, connective tissue disorders, and as a primary disorder termed "dilatative arteriopathy."
- Management of these aneurysms is challenging, but individualized surgical and endovascular approaches, selectively applied to symptomatic or larger aneurysms, can be effective in preventing growth or rupture.
- Application of advanced endovascular techniques, including flow-directing stents and stent-assisted coiling, has increasingly allowed successful treatment of complex nonsaccular aneurysms.

Historical note and terminology

An aneurysm is a pathologic, localized blood vessel dilatation, which is called "saccular" when the inflow and outflow points are in common and "nonsaccular" when arterial dilatation is greater than 1.5 times normal without a clearly defined neck (ie, the inflow and outflow points are longitudinally separate) (Flemming et al 2004). "Fusiform" refers to nonsaccular, spindle-shaped aneurysms with focal circumferential dilatation whereas "dolichoectatic" aneurysms are predominantly elongated and tortuous with a uniform enlarged circumference (dolichos=long; ectasia=distended).{embed="pagecomponents/media_embed" entry_id="9065"} Some authors use these terms interchangeably (Steel et al 1982), but important differences in their clinical features and pathophysiology can be distinguished. A third subtype, "transitional", has been proposed, which features focal circumferential dilatation, elongation, and displacement. Notably, the recent trend in study of dolichoectatic and fusiform aneurysms focuses on dilatation as the main pathologic feature, and consequently, the term "dilatative arteriopathy" has gained currency (Caplan 2005; Lou and Caplan 2010).

Classically, 2 forms of nonsaccular aneurysms are separated by their pathophysiology: (1) acute dissecting and (2) chronic fusiform or dolichoectatic aneurysms (Nakatomi et al 2000). Acute dissecting aneurysms will not be addressed in this article. These aneurysms, as well as those occurring from infection or neoplastic infiltration of the arterial wall, are addressed under separate headings.

Some radiologists describe nondistended tortuous and elongated arteries commonly visualized on CT and MRI as dolichoectatic, but distension is much more closely correlated with hemorrhage than elongation, making the distinction

clinically relevant. Aneurysms greater than 2.5 cm in diameter are referred to as "giant" (Wehman et al 2006). When a giant aneurysm partially thromboses, revealing tortuous vascular channels, it is called a “giant serpentine” aneurysm (Segal and McLaurin 1977; Sari et al 2006). Additional names for nonsaccular intracranial aneurysm of the basilar artery include ectasia, S-shaped aneurysm, wandering basilar artery, tortuous basilar artery, cirsoid aneurysm, megadolichovascular malformation, megadolichobasilar artery, dolichomegavertebralis anomaly, and aneurysmal malformation (Flemming et al 2004).

Moniz provided the first angiographic demonstration of a dolichoectatic aneurysm in 1934, but the clinical condition had been previously described by Dandy in 11 instances in the vertebrobasilar circulation and in 6 instances in the internal carotid artery circulation (Moniz 1934; Dandy 1944).

Clinical manifestations

Presentation and course

Fusiform or dolichoectatic aneurysms may present with headache or other symptoms related to compression of adjacent brain structures, with infarction or transient ischemic attacks, or with rupture and hemorrhage. Often, however, they are discovered incidentally. Specific symptoms depend on aneurysm location.

Intracranial nonsaccular aneurysms occur in both the vertebrobasilar and internal carotid artery circulations. Although generally thought to be more frequent in the former, the series of Anson and colleagues suggests that both circulations are affected approximately equally, fusiform aneurysms being more common in the internal carotid artery distribution and dolichoectatic aneurysms in the vertebrobasilar distribution (Shokunbi et al 1988; Anson et al 1996).

In the anterior circulation, the middle cerebral artery is most commonly affected. These aneurysms occur more commonly in elderly adults, with a markedly increased frequency in men in the vertebrobasilar circulation and a slight preponderance for women in the internal carotid artery circulation. Giant fusiform aneurysms occur less frequently in the anterior circulation (20%) than in the posterior circulation (80%) where the basilar trunk and posterior cerebral arteries are most frequently involved (Drake and Peerless 1997). Occasionally, multiple fusiform intracranial aneurysms or a combination of saccular and fusiform aneurysms occur in the same person (Anson et al 1996; Josephson and Johnston 2005).

Transient ischemic attacks, cerebral infarction, or hemorrhage are common presenting symptoms (Echiverri et al 1989; Anson et al 1996). Hemorrhage occurs in 15% to 20% of cases, usually into the subarachnoid space, whereas intraparenchymal hemorrhage is infrequent (Anson et al 1996). Typically the onset is abrupt, followed by improvement or stuttering progression. Minor asymptomatic perianeurysmal hemorrhages are frequently noted on MRI and at autopsy, but their prognostic importance is unknown. At other times, artery-to-artery embolization causes transient or permanent ischemic symptoms (Cohen et al 1980; Steel et al 1982). Hemodynamic factors resulting in sluggish blood flow and red-cell sludging can also cause intermittent ischemic symptoms. Dissection into the wall of the aneurysm sometimes occurs, resulting in obstruction of branch vessels, headache, and further dilatation of the aneurysm. Tandem atherosclerotic stenotic lesions are sometimes present and responsible for symptoms (Little et al 1981). Headache is a common symptom of large fusiform aneurysms. The pain is often severe, lateralized to the side of the aneurysm, occipital with basilar artery aneurysms, retro-orbital or frontal with internal carotid artery aneurysms, and may antedate other neurologic symptoms by days or months. Sometimes the headache is positional. The mechanism of the headache is generally attributed to traction on nearby pain-producing structures and is relieved by separating the aneurysm from these tissues (Little et al 1981). Nausea, vomiting, and neck stiffness strongly suggest subarachnoid hemorrhage.

When the internal carotid artery is affected, the supraclinoid portion is most frequently involved, followed by the petrous segment. Often the aneurysm is contiguous, involving both the internal carotid artery and middle cerebral artery (Little et al 1981). When the supraclinoid internal carotid artery is aneurysmal, compression of the optic nerve against the dural fold that overlies it can result in decreased visual acuity, scotoma, optic atrophy, or even the Foster-Kennedy syndrome. Occasionally, a large aneurysm mimics a pituitary tumor by compressing the pituitary gland and optic chiasm, producing a bitemporal hemianopsia (Hilton and Hoyt 1966). These compressive symptoms are usually both insidious and progressive. In rare instances, aneurysms of the cavernous carotid artery rupture into the sphenoid sinus and present as recurrent epistaxis (Pritz 1994).

Compression of adjacent structures, particularly the cranial nerves or the brainstem, is the cause of symptoms in about one third of cases (Anson et al 1996). Cranial nerve compression can result from a tortuous dolichoectatic basilar artery or frank aneurysmal dilatation (Smoker et al 1986; Pico et al 2015). Cranial nerves that are frequently compressed include (1) the trigeminal nerve with associated trigeminal neuralgia or trigeminal neuropathy or both (Echiverri et al 1989; Anson et al 1996), (2) the facial nerve with hemifacial spasm or facial paralysis or both, and (3) the oculomotor nerve with pupillary or eye movement dysfunction. One series using CT to pursue a diagnosis of hemifacial spasm revealed a dolichoectatic vertebrobasilar artery in 36 of 46 patients (Digre et al 1988). Infrequently, cranial nerves VIII, IX, X, XI and XII are affected, either singly or in combination, together with long-tract findings. In a series of 20 patients with vertebrobasilar dolichoectasia, half the patients had an isolated 3rd, 6th, or 7th nerve palsy, whereas the other half of the patients had multiple neurologic deficits, including combinations of compressive cranial neuropathies, both compressive and ischemic deficits, and hydrocephalus (Smoker et al 1986).

An aneurysmal basilar artery can compress the brainstem, resulting in pontomedullary and cerebellar signs including downbeat nystagmus and internuclear ophthalmoplegia mimicking a posterior fossa mass lesion (Jacobson and Corbett 1989). Motor weakness without sensory signs is frequent. Dysphagia, hiccups, and orthostatic hypotension occur infrequently (Echiverri et al 1989). Rarely, obstructive hydrocephalus is present (Yu et al 1982; Gelal et al 2002; Siddiqui et al 2008). Large aneurysms frequently have intraluminal thrombus seen either on MRI or at surgery. Such a thrombus can propagate, occlude penetrating vessels (especially those originating from the basilar artery), and result in brainstem ischemia (Echiverri et al 1989; Anson et al 1996). Rarely, the entire basilar artery may thrombose, leading to massive brainstem ischemia (Watanabe et al 1994).

Prognosis and complications

The prognosis of incidentally discovered fusiform and dolichoectatic aneurysms is starting to be defined. When the only symptoms are those of isolated cranial nerve compression, the prognosis is often good, but sometimes symptoms can progress and multiple cranial neuropathies and brainstem compression can develop. If intraluminal thrombus is also present, the risk of stroke is high and the prognosis is guarded. Some authors have quoted mortality rates as high as 65% to 100% for untreated giant aneurysms at 2 years (Gonzalez et al 2006), and subarachnoid hemorrhage is reported in 18% to 40% of patients in some series of dolichoectatic and fusiform aneurysms (Hayes et al 1967; Little et al 1981; Nishizaki et al 1986; Anson et al 1996). These data are almost certainly influenced by a significant selection bias and, therefore, should be interpreted cautiously.

More recent series have shown evidence of a more positive prognosis overall. Persons with internal carotid artery circulation aneurysms fare better than those with vertebrobasilar system disease (Anson et al 1996), and patients with aneurysms of the posterior cerebral arteries fare better than patients with aneurysms of the proximal vertebrobasilar trunk (Coert et al 2007). A report of 159 patients with vertebrobasilar nonsaccular intracranial aneurysms reported an annual hemorrhage rate of 0.9%, increasing to 2.3% annually among the subset of nonsaccular aneurysms that were either fusiform (primarily dilatative) or transitional subtypes. The overall death rate was 41% during a mean 4.4-year follow-up (Flemming et al 2004). In the same series, the 1- and 10-year risk of cerebral infarction was 2.7% and 15.9%, respectively, and the risk of recurrent cerebral infarction was 6.7% per patient year. Median survival was 7.8 years, with death most often due to cerebral ischemia (Flemming et al 2005). Growth in aneurysm diameter is correlated with an increasing number of symptoms, a tendency for increased subarachnoid hemorrhage risk, and a more than 10-fold higher 5 year mortality rate (56.5% vs. 3.7%, p=0.004) (Mangrum et al 2005). In another series of 156 patients with nonsaccular intracranial vertebrobasilar aneurysms of the dolichoectatic subtype (not including spindle-shaped fusiform or transitional subtypes) who were followed for an average of almost 12 years, 38% suffered an ischemic stroke and 13% an intracranial hemorrhage (Passero and Rossi 2008). Among this series of patients, there were 3 subarachnoid hemorrhages and 15 intracerebral hemorrhages during a mean follow-up of almost 10 years, for crude incidence rates of 2.2 per 1000 person-years for subarachnoid hemorrhage and 11 per 1000 person-years for intracranial hemorrhage (Passero et al 2005).

Another series reported retrospective natural history data from 121 patients with unruptured intradural fusiform aneurysms (Sacho et al 2014). Importantly, in this series atherosclerotic aneurysms were separated by well-defined criteria from nonatherosclerotic aneurysms. For the 96 patients followed with nonatherosclerotic aneurysms, the prognosis was relatively good; over 193 person-years of follow-up, 1 rupture occurred (with fatal outcome), and 8 patients showed aneurysm progression (for a rate of 1.6% per year). Symptomatic presentation and diameter larger than 7 mm were risk factors for progression. In contrast, in 25 patients with atherosclerotic aneurysms, 65% exhibited

progression (at a rate of 12% per year), and 5 died due to aneurysm rupture or ischemic stroke (a rate of 5.2% per year), frequencies that were significantly higher than in the nonatherosclerotic aneurysm group.

Clinical vignette

Case 1. A 59-year-old male with hypertension, diabetes, and dyslipidemia, with history of coronary disease treated with coronary bypass grafting, and with history of abdominal aortic aneurysm repair, presented with recurrent posterior circulation ischemic events, improving with intravenous tissue plasminogen activator on one occasion and intravenous heparin therapy on the second and third occasions. He was left with infarction in the territory of the left posterior cerebral artery, with right hemianopsia, and verbal memory deficits. CT scanning of brain revealed an ectatic basilar artery with mural calcifications (A). CT angiography demonstrated basilar artery dolichoectasia (B). He recovered and continued with stable deficits through 2 years of follow-up, on warfarin.{embed="pagecomponents/media_embed" entry_id="16907"}

This case demonstrates the risks of ischemic infarction that accompany the common form of basilar artery dolichoectasia in patients with typical atherosclerotic risk factors.

Case 2. A 36-year-old athletic female, without known vascular risk factors, presented during the third trimester of pregnancy with headache, slurred speech, and “twisted mouth.” Head CT scan showed an extra-axial mass compressing the left cerebral peduncle (A), recognized by CT angiography (B) as a partially thrombosed fusiform aneurysm of the left posterior cerebral artery. There was also tortuosity and ectasia of the basilar and left internal carotid arteries (C). Six years later, the vascular mass had expanded (D), producing severe midbrain compression and hydrocephalus, with spastic quadriparesis. She was treated with ventricular drainage and clipping of the left posterior cerebral artery proximal to the aneurysm.{embed="pagecomponents/media_embed" entry_id="16908"}

Dilatative arteriopathy in cases like this, with severe progressive multifocal ectasia of cerebral arteries in the absence of a detectable generalized vasculopathy, can produce deficits due to cranial nerve and parenchymal brain compression as well as risks of ischemia or hemorrhage.

Biological basis

Etiology and pathogenesis

Nonsaccular aneurysms of fusiform, dolichoectatic, or transitional forms can arise from a wide variety of causes (Table 1). In the past it has been generally accepted that fusiform or dolichoectatic aneurysms commonly develop due to severe, complicated, atherosclerotic weakening the arterial wall (Weeler 1992; Vinters et al 2004). In particular, basilar artery dolichoectasia often appears in the setting of generalized atherosclerosis, as in vignette Case 1 (above). Scholarship has emphasized the association of dolichoectasia with microvascular disease (Gutierrez et al 2011; Pico et al 2015). Fusiform aneurysms can also likely result from focal intracranial dissections or other arteriopathies (Mizutani and Aruga 1992; Anson 1998; Zhang et al 2016). It is helpful to recognize that dolichoectasia may be a final common pathway of any arterial wall disease that disrupts the structural integrity of the tunica media (Pico et al 2015).

Table 1. Underlying Causes of Fusiform and Dolichoectatic Aneurysms

- Atherosclerosis
- Dissection
- Dilatative arteriopathy
- Infective endocarditis
- Collagen vascular disease (Marfan syndrome, Ehlers-Danlos syndrome)
- Polycystic kidney disease
- HIV vasculopathy
- Pseudoxanthoma elasticum
- Fibromuscular dysplasia
- Neurofibromatosis
- Sickle cell disease
- Alpha-1-antitrypsin deficiency
- Alpha-glucosidase deficiency
- Giant-cell arteritis
- Metastasis (choriocarcinoma, atrial myxoma)

Genetics. In some cases, a clear genetic basis for nonsaccular aneurysms can be identified. In autosomal dominant polycystic kidney disease, dolichoectasia occurs in about 2% of cases (Schievink et al 1997). Rarely, in younger patients, fusiform aneurysms are due to congenital defects in the media connective tissue or smooth muscle cells, such as occurs in Marfan syndrome, Ehlers-Danlos syndrome, pseudoxanthoma elasticum, fibromuscular dysplasia, neurofibromatosis, sickle cell disease, alpha-1-antitrypsin deficiency, and alpha-glucosidase deficiency (Makos et al 1987; Schievink et al 1994; Caplan 2005). In one report of 3 patients, late-onset Pompe disease (alpha-glucosidase deficiency) was associated with dolichoectatic aneurysms (Laforet et al 2008).

Other causes of nonsaccular aneurysms. Dolichoectasia, particularly of the vertebrobasilar and carotid arteries, can be a relatively common manifestation in patients with atherosclerotic disease or cerebral microvascular disease. Fusiform aneurysms sometimes arise due to focal injury to the arterial wall due to spontaneous dissection or inflammation. Isolated cases associated with giant cell arteritis have been reported. Rare cases occur due to bacterial or fungal infection in the arterial wall, termed “infective aneurysms.” Interestingly, there are a handful of case reports of fusiform intracranial aneurysms in children and adults with HIV infection (Tipping et al 2006). Rare involvement of the arterial wall with metastatic neoplasms can also sometimes result in focal mural weakening and fusiform aneurysm formation. Finally, focal fusiform aneurysms or progressive transitional aneurysms can often be found without any identified risk factors or cause, suggesting the presence of an underlying idiopathic arteriopathy, as in the clinical vignette, case 2 above.

Pathology and pathophysiology. Dolichoectasia of intracranial arteries, particularly involving the basilar artery, is common in stroke patients, with reported frequency as high as 6% in a series of consecutive autopsies of patients with stroke; in the particular context of paramedian pontine infarction, it was thought to be the underlying cause in a full 31% of cases (Kwon et al 2009). Most of these patients have atherosclerotic risk factors. During the process of chronic atherosclerosis, fibrous tissue may replace smooth muscle and elastic fibers of the internal elastic lamina, allowing vessels to stretch in length and width. In support of this concept, an animal model employing injection of elastase into the cisterna magna of mice successfully induced intracranial dolichoectasia (Dai et al 2015). When widening is extreme, the artery balloons into a fusiform aneurysm, whereas if both lengthening and widening occur, the aneurysm becomes dolichoectatic. Because the aneurysmal dilatation is circumferential, there is no discrete aneurysm neck, and there is no preference for localization at arterial branch points.

Another mechanism potentially of relevance to dolichoectasia is the process of compensatory enlargement of atherosclerotic arteries known as “adaptive remodeling.” This has been described in various arterial beds, including the coronary arteries (Glagov et al 1987) and cervical arteries. Adaptive remodeling produces enlargement of the external wall of the artery compensating for mural plaque, variably resulting in net preservation, constriction, or dilatation of the arterial lumen. Adaptive remodeling is thought to be driven by increased wall shear stress and is mediated by endothelial signaling factors, including nitric oxide production and cytokine production. Breakdown of the internal elastic lamina can be documented experimentally due to high flow-induced arterial enlargement (Masuda et al 1999). Such mechanisms may also underlie arterial dolichoectasia in atherosclerosis.

Some authors, alternatively, have emphasized the preferential association of intracranial dolichoectasia with signs of lacunar stroke and cerebral microvascular disease. The Etude du Profil Genetique de l'Infarctus Cerebral (GENIC) study suggested that dilatative arteriopathy occurs as part of a systemic vascular ectatic disease and is associated with cerebral microvascular disease, distinct from atherosclerosis and involving primarily the tunica media and the extracellular matrix (Pico 2005; Pico 2007). Dysfunctional matrix metalloprotein activation and imbalance between protease and antiprotease activities in the extracellular matrix may lead to loss of elastic fibers, with disruption of the internal elastic lamina and consequent excessive dilatation (Pico 2007; Lou and Caplan 2010; Gutierrez et al 2011). This process has been termed dilatative arteriopathy (Lou and Caplan 2010). Atherosclerotic plaque found in the affected basilar arteries has been considered by these authors a secondary effect of the arteriopathy and dilatation (Pico 2007; Lou and Caplan 2010). It is clear that the high degree of overlap between risk factors for atherosclerosis and those for microvascular disease leads to a frequent co-occurrence of these conditions, and it may be that typical intracranial dolichoectasia in vasculopathic patients arises from a mix of etiologies.

One retrospective clinicopathologic series of 16 chronic fusiform aneurysms described a characteristic histological pattern (Nakatomi et al 2000). There was suggestion of a progression starting with (1) fragmentation of the internal elastic lamina with consequent intimal hyperplasia followed by (2) neoangiogenesis within the hyperplastic intima, coinciding with gadolinium enhancement of the aneurysm wall when the diameter is larger than 12 mm. As the

aneurysm progresses and neoangiogenesis continues, there is (3) intramural hemorrhage with thrombus formation followed by (4) ensuing hematoma recanalization, usually in aneurysms that have grown larger than 20 mm.

Other causes of structural defects in the arterial wall leading to aneurysm formation that have been described include connective tissue replacement, deficient elastin, fibromuscular dysplasia, degeneration of the internal elastica lamina, fibrous and collagenous replacement of the media, and inflammation (Hirsch and Roessmann 1975; Paulson et al 1978). The internal elastic lamina is of variable thickness and may contain abrupt gaps. In some locations the artery is supported only by adventitia, making these likely rupture points (Shokunbi et al 1988). Thickened, hypertrophic, fibrous bands, and occasionally the absence of intima have been described (Stehbens 1972; Segal and McLaurin 1977). Widespread deficiencies in the muscularis and internal elastic lamina may be found, resulting in multiple intracranial and extracranial aneurysms (Little et al 1981; Nishizaki et al 1986; Braunsdorf 1987). In a series of 362 patients with alpha-1-antitrypsin deficiency, the finding of ruptured intracranial aneurysms among 3 and spontaneous cervical artery dissection in 1 suggests that an imbalance in proteolytic enzymes may contribute to development of ruptured intracerebral aneurysm and dissection (Schievink et al 1994). Fusiform basilar artery aneurysms and alpha-glucosidase deficiency (Pompe disease) have been reported among 3 siblings, 1 of whom had a temporal artery biopsy that revealed glycogenous deposition in smooth muscle (Makos et al 1987). One series proposed recurrent medial dissection as the primary etiology for fusiform aneurysm formation (Day et al 2003). Giant fusiform aneurysms developing in children in angiographically normal appearing vessels after balloon occlusion treatment have been described (Johnston et al 1998). Some giant fusiform aneurysms originate from saccular aneurysms (Drake and Peerless 1997). The origins of many of these giant aneurysms probably lie in a hitherto undiscovered arteriopathy.

Epidemiology"

Population-based studies of the incidence or prevalence of nonsaccular aneurysms are unavailable. However, available data suggest a sharp distinction in prevalence between focal fusiform aneurysms and dolichoectasia.

In 2 large autopsy series totaling over 16,000 cases, 15 (less than 0.1%) fusiform aneurysms were discovered (Housepian and Pool 1958; Hayes et al 1967). In autopsy series, these aneurysms accounted for 5% to 9% of all aneurysms (Housepian and Pool 1958; Shokunbi et al 1988). In Yu's large series of 50,000 angiograms, 31 fusiform aneurysms were discovered (0.06%), 14 of which were in the internal carotid artery circulation, 8 in the vertebrovascular circulation, and 9 in both (Yu et al 1982). Thus, true fusiform aneurysms are relatively uncommon. In contrast, in a study of 381 consecutive autopsies of patients with stroke, 23 cases (6%) had intracranial dolichoectasia, primarily involving the basilar artery, designated as “dilatative arteriopathy” by these authors (Pico 2007).

Prevention

No specific means of preventing the formation of fusiform and dolichoectatic aneurysms is known. Given the strong association between these aneurysms and vascular disease, strict control of atherogenic and arteriosclerotic risk factors, such as hypertension, hyperlipidemia, hyperglycemia, and cigarette smoking, might delay progression of these nonsaccular aneurysms.

Differential diagnosis

An abrupt onset of severe headache with or without focal neurologic symptoms suggests intracranial hemorrhage. Hemorrhage has sometimes been considered less likely with fusiform or dolichoectatic aneurysms than with saccular aneurysms. However, hemorrhages without question can occur. In a long-term prospective study of 156 patients with vertebrobasilar dolichoectasia followed for an average of 9 years, 6 subarachnoid hemorrhages and 26 intraparenchymal hemorrhages occurred (Passero 2005).

When multiple cranial neuropathies and headache are present, tuberculous, carcinomatous, and other basal granulomatous meningitides must be considered. Neuroimaging is mandatory, and when normal, lumbar puncture is required to rule out subarachnoid hemorrhage or a CSF inflammatory process.

Transient ischemic attacks and stroke can be caused by large fusiform or dolichoectatic aneurysms. The results of a complete work up for other causes of stroke must be considered in reaching this conclusion. In such instances, investigation will frequently demonstrate intraluminal thrombus when the aneurysm is the cause for symptoms.

Giant nonsaccular aneurysms can mimic tumors by producing headache, seizures, hydrocephalus, and focal neurologic

signs. Optic atrophy in one eye and papilledema in the other suggests the Foster Kennedy syndrome, whereas hypopituitarism and a bitemporal visual field defect can mimic a pituitary neoplasm. Pressure on isolated cranial nerves can result in trigeminal neuralgia, hemifacial spasm, or other cranial neuropathies for which the differential diagnosis is broad. There is no simple clinical method to differentiate these disorders, but neuroimaging usually easily confirms the diagnosis of aneurysm. Occasionally, misinterpretation of a vascular mass as a neoplasm on brain imaging leads to a disastrous biopsy procedure.

Diagnostic workup

The appropriate initial workup depends on the mode of presentation. Symptoms suggesting subarachnoid hemorrhage should be evaluated with rapid nonenhanced brain CT scan. In situations where subarachnoid hemorrhage is strongly suspected and radiologic tests are negative for blood, lumbar puncture should be performed, provided there is no evidence of mass effect in the posterior fossa. The initial CT scan is often immediately followed by CT angiography, which can detect dolichoectatic arteries and fusiform aneurysms as well as most saccular aneurysms larger than 2 mm in size. In contrast, presentation with cranial nerve palsy or signs of parenchymal compression should usually prompt brain MRI scanning (without and with intravenous contrast) and MR angiography (MRA).

MRI with MRA is also the test of choice to identify and characterize most incidentally discovered proximal fusiform and dolichoectatic aneurysms. It will demonstrate the dilated, tortuous vessel as a signal void region corresponding to the artery (Iwama et al 1990). Compared to catheter angiography, MRI/MRA has the advantage of more accurately determining the true size of aneurysm, because catheter angiography can only outline the residual lumen without characterizing the vessel wall. Additionally, effects on adjacent structures and the presence of intraluminal thrombus will typically be better demonstrated on MRI (Martin 2011). MRI may also assist in differentiating a primary fusiform aneurysm from one that is secondary to arterial dissection, where an intimal flap, wall hematoma, or double lumen may be demonstrated (Iwama et al 1990). High field strength MRI scanning is increasingly used to perform vessel wall imaging, with remarkable resolution.

CT angiography can provide many of the same advantages as MR angiography, with equal or better resolution for definition of the vascular lumen, and high sensitivity to mural changes, particularly for mural calcifications. CT angiography is also becoming increasingly useful in assessing distal aneurysms.

In light of development of increasingly sophisticated endovascular interventions for both dissections and nonsaccular intracranial aneurysms, diagnostic catheter angiography may be needed to help decide what therapeutic approach should be pursued.

Management

There is a lack of prospective trials on the management of fusiform aneurysms. It is thought that in older patients with asymptomatic fusiform aneurysms, conservative management should include antiplatelet agents and vascular risk-reduction measures such as blood pressure control and cholesterol-lowering therapy. In the setting of cerebral ischemia, either antiplatelet or anticoagulant therapy may be considered. Theoretically, anticoagulant therapy might be more effective than antiplatelet therapy at reducing ischemic recurrence in the setting of a large dilated aneurysm with thrombus present, but a potential increased bleeding risk may offset this benefit. There have been no randomized trials comparing these 2 different treatment approaches, and thus, treatment decisions must be based on clinical judgment, weighing individual patient characteristics.

When the aneurysm is responsible for severe headache, cranial nerve compression, brain compression, or hydrocephalus, surgical decompression followed by direct treatment of the aneurysm can be considered. Similarly, in the setting of hemorrhage, efforts to secure the aneurysm should generally be undertaken if feasible. Finally, aneurysm enlargement over serial imaging studies has been shown to predict rupture and death (Mangrum et al 2005) and usually signals the need to consider intervention. It is widely accepted that surgical and endovascular treatment of fusiform and dolichoectatic aneurysms is difficult technically and must be individualized, depending on location, size, symptoms, and flow characteristics.

Surgical treatments of nonsaccular aneurysms include (1) direct clipping, which is difficult because these aneurysms do not have a neck, and which usually refers to external refashioning with clips, (2) resection and re-anastomosis, with or without additional bypass, if only a short length is involved, or the artery is redundant, (3) trapping and distal

bypass, particularly for the anterior circulation, (4) proximal occlusion, (5) transposition, (6) thrombectomy, endarterectomy and aneurysmorrhaphy, and (7) reinforcement of the aneurysm dome by wrapping. Reinforcement by wrapping and proximal ligation are the least successful techniques because rebleeding and symptom progression rates are high when the aneurysm itself is not eliminated. These methods are, therefore, reserved for aneurysms that cannot be eliminated from the circulation. If tortuous vessels cause brainstem compression in the absence of intraluminal thrombus, the artery may be mobilized and transposed. Aneurysms of the internal carotid artery circulation are easier to treat than those in the vertebrobasilar distribution, but supraclinoid carotid aneurysms may be particularly difficult because of the important branch arteries and perforators that must remain patent. Similarly, in giant aneurysms, perforating arteries to deep structures are frequently involved and must be preserved. Sometimes these arteries may function as collaterals that are only visualized after vessel occlusion, and not preoperatively (Drake and Peerless 1997).

With the development of increasingly sophisticated catheters, detachable balloons, coils, and stents, endovascular approaches to fusiform aneurysm treatment have become increasingly viable as alternatives to open surgical management (Higashida et al 1991; Higashida et al 1997; Zhang et al 2016). Unlike narrow-necked saccular aneurysms, fusiform aneurysms are extremely difficult to treat with coil embolization alone due to the width of or lack of a well-defined aneurysm neck, which limits coil retention and results in incomplete thrombosis. Sometimes the parent artery must be sacrificed to address an otherwise untreatable aneurysm. Successful endovascular occlusion of the internal carotid artery for a cavernous or paraclinoid internal carotid artery aneurysm proximal to the origin of the ophthalmic artery has been reported (Higashida et al 1991; Aymard et al 1992). Proximal occlusion of an aneurysmal posterior cerebral artery can also be undertaken (as in clinical vignette case 2). In the vertebrobasilar circulation, treatment is particularly difficult when the basilar artery is involved. A balloon test occlusion of one or both vertebral arteries, or of the basilar artery, preceded by a test of functionality of the posterior communicating artery, should be attempted to document whether collaterals are adequate and occlusion can be tolerated (Aymard et al 1992). In some cases, a multimodality combination of microsurgical and endovascular approaches can be successfully applied in complex intracranial aneurysms not amenable to endovascular or microsurgical approaches alone (Lawton et al 2003).

The advent of balloon-assisted coiling, stent-assisted coiling, balloon-in-stent-assisted coiling, and even stent-in-stent remodeling has advanced the ability of the interventionalist to treat aneurysms with circumferential dilatation or wide necks (Coert et al 2007; Lubicz et al 2008). In particular, stent-assisted coiling, excluding the coil mass from the arterial lumen with a stent, has become a frequent solution to this problem. Sometimes multiple overlapping stents are required for complex aneurysms (Won et al 2015). Procedure-related complication rates can be high (19% in this series).

Recent years have also seen increasing use of a flow-diverting stents with closed-cell mesh designs that direct blood flow away from the aneurysm while preserving flow in the parent vessel and adjacent branches.{embed="pagecomponents/media_embed" entry_id="16909"} When flow diversion is successful, angiographic stasis occurs in the aneurysm, preventing rupture and causing the aneurysm to decrease in size over time (Pumar et al 2013). These flow-diverting stents have been approved for use in European countries for some years and more recently were approved in the United States. They can be used in both anterior and posterior circulation fusiform aneurysms, and both elective aneurysm treatment and treatment of ruptured aneurysms can be accomplished with flow-diversion techniques. Even aneurysms in small distal vessels, such as the distal anterior cerebral artery, can sometimes be successfully treated with flow-diverting stents (Nossek et al 2017). A systematic review found reports of a total of 1704 aneurysms (81% saccular and 18% fusiform) treated with flow-diverter devices, with more than an 80% rate of final complete occlusion, and with a neurologic morbidity rate of 3.5% and a mortality rate of 3.4% (Briganti et al 2015).

Outcomes

Surgical treatment of fusiform aneurysms is generally reserved for larger, more often symptomatic or progressing aneurysms; therefore, outcomes cannot be directly compared to results observed in aneurysms treated expectantly. Reported case series reflect the complexity and difficulty of surgical treatment of these cases. One surgical case series reported a 30% complication rate, with no operative mortality, in 40 surgically-treated patients followed over a mean period of 2.8 years; 58% made good recoveries (Glasgow Outcome Scale score 1), 20% had moderate disabilities (Glasgow Outcome Scale score 2), 8% had severe disabilities (Glasgow Outcome Scale score 3), and 15% died (Anson et al 1996). A report found that surgical or endovascular treatment of unruptured intradural fusiform aneurysms in 23

patients was associated with death in 3 patients (13%) and mild resultant disability in another 3 patients (13%) (Sacho et al 2014). Lin and colleagues, reporting early United States experience with use of the “Pipeline” flow diversion stent in the treatment of 26 patients with ruptured aneurysms, including 6 fusiform aneurysms, described a periprocedural complication rate of 19% and a complete occlusion rate of 78% in follow-up angiograms (Lin et al 2014). These studies demonstrate both the considerable morbidity and the encouraging success that can be achieved in the treatment of these challenging aneurysms.

Special considerations

Pregnancy

No clear influence of pregnancy on aneurysm appearance or progression has been described.

Anesthesia

Anesthesia techniques similar to those used for other difficult aneurysm surgeries should be used. Electroencephalographic and evoked potential monitoring and barbiturates titrated to burst suppression must be considered. When prolonged interruption of blood flow is likely, hypothermic circulatory arrest may be necessary (Wascher 1995).

References cited

Anson JA. Treatment strategies for intracranial fusiform aneurysms. Neurosurg Clin N Am 1998;9:743-53. PMID 9738104

Anson JA, Lawton MT, Spetzler RF. Characteristics and surgical treatment of dolichoectatic and fusiform aneurysms. J Neurosurg 1996;84:185-93. PMID 8592220

Aymard A, Hodes JE, Rufenacht D, Merland JJ. Endovascular treatment of a giant fusiform aneurysm of the entire basilar artery. AJNR Am J Neuroradiol 1992;13 (4):1143-6. PMID 1636527

Braunsdorf WE. Fusiform aneurysm of basilar artery and ectatic internal carotid arteries associated with glycogenosis type 2 (Pompe's disease). Neurosurgery 1987;21(5):748-9. PMID 3480453

Briganti F, Leone G, Marseglia M, et al. Endovascular treatment of cerebral aneurysms using flow-diverter devices: a systematic review. Neuroradiol J 2015;28(4):365-75. PMID 26314872

Caplan LR. Dilatative arteriopathy (dolichoectasia): What is known and not known. Ann Neurol 2005;57(4):469-71. PMID 15786452

Coert BA, Chang SD, Do HM, Marks MP, Steinberg GK. Surgical and endovascular management of symptomatic posterior circulation fusiform aneurysms. J Neurosurg 2007;106(5):855-65. PMID 17542530

Cohen MM, Hemalatha CP, D'Addario RT, Goldman HW. Embolization from a fusiform middle cerebral artery aneurysm. Stroke 1980;11:158-61. PMID 7368242

Dai D, Kadirvel R, Rezek I, Ding YH, Lingineni R. Kallmes D. Elastase-induced intracranial dolichoectasia model in mice. Neurosurgery 2015;76(3):337-43. PMID 25599211

Dandy WE. Intracranial Arterial Aneurysms. New York: Hafner, 1944.

Day AL, Gaposchkin CG, Yu CJ, Rivet DJ, Dacey RG Jr. Spontaneous fusiform middle cerebral artery aneurysms: characteristics and a proposed mechanism of formation. J Neurosurg 2003;99(2):228-40. PMID 12924694

Digre KB, Corbett JJ, Smoker WR, McKusker S. CT and hemifacial spasm. Neurology 1988;38(7):1111-3. PMID 3386831

Drake CG, Peerless SJ. Giant fusiform intracranial aneurysms: review of 120 patients treated surgically from 1965 to 1992. J Neurosurg 1997;87:141-62. PMID 9254076

Echiverri HC, Rubino FA, Gupta SR, Gujrati M. Fusiform aneurysm of the vertebrobasilar arterial system. Stroke

1989;20:1741-7. PMID 2595737

Flemming KD, Wiebers DO, Brown RD Jr, et al. Prospective risk of hemorrhage in patients with vertebrobasilar nonsaccular intracranial aneurysm. J Neurosurg 2004;101(1):82-7. PMID 15255255

Flemming KD, Wiebers DO, Brown RD Jr, et al. The natural history of radiographically defined vertebrobasilar nonsaccular intracranial aneurysms. Cerebrovasc Dis 2005;20(4):270-9. PMID 16123548

Gelal F, Vidinli BD, Yurt A, Cakir Y, Uygur M. Giant fusiform basilar artery aneurysm causing obstructive hydrocephalus. Skull Base 2002;12(4):197-200. PMID 17167680

Glagov S, Weisenberg E, Zarins CK, Stankaunvicius R, Kolettis GJ. Compensatory enlargement of human atherosclerotic coronary arteries. N Engl J Med 1987; 316:1371-5. PMID 3574413

Gonzalez NR, Duckwiler G, Jahan R, Murayama Y, Vinuela F. Challenges in the endovascular treatment of giant intracranial aneurysms. Neurosurgery 2006;59(5 Suppl 3):S113-24. PMID 17053594

Gutierrez J, Sacco RL, Wright CB. Dolichoectasia-an evolving arterial disease. Nat Rev Neurol 2011;7(1):41-50.** PMID 21221115

Hayes WT, Benhardt H, Young JM. Fusiform arteriosclerotic artery aneurysm of the basilar artery. Five cases including two ruptures. Vasc Surg 1967;1:171-8. PMID 5299951

Higashida RT, Halbach VV, Dowd CF, Barnwell SL, Hieshima GB. Intracranial aneurysms: interventional neurovascular treatment with detachable balloons-results in 215 cases. Radiology 1991;178:663-70. PMID 1994399

Higashida RT, Smith W, Gress D, et al. Intravascular stent and endovascular coil placement for a ruptured fusiform aneurysm of the basilar artery. Case report and review of the literature. J Neurosurg 1997;87(6):944-9. PMID 9384409

Hilton GF, Hoyt WF. An atherosclerotic chiasmal syndrome. JAMA 1966;196:1018-20. PMID 5295932

Hirsch CS, Roessmann U. Arterial dysplasia with ruptured basilar artery aneurysm: report of a case. Hum Pathol 1975;6:749-58. PMID 1183998

Housepian EM, Pool JL. A systematic analysis of intracranial aneurysms from the autopsy file of the Presbyterian Hospital, 1914-1956. J Neuropathol Exp Neurol 1958;17:409-23. PMID 13564252

Iwama T, Andoh T, Sakai N, Iwata T, Hirata T, Yamada H. Dissecting and fusiform aneurysms of vertebro-basilar systems. MR imaging. Neuroradiology 1990;32:272-9. PMID 2234385

Jacobson DM, Corbett JJ. Downbeat nystagmus associated with dolichoectasia of the vertebrobasilar artery. Arch Neurol 1989;46:1005-8. PMID 2775004

Johnston SC, Halbach VV, Smith WS, Gress DR. Rapid development of giant fusiform cerebral aneurysms in angiographically normal vessels. Neurology 1998;50(4):1163-6. PMID 9566418

Josephson SA, Johnston SC. Multiple stable fusiform intracranial aneurysms following atrial myxoma. Neurology 2005;64(3):526. PMID 15699386

Kwon HM, Kim JH, Lim JS, Park JH, Lee SH, Lee YS. Basilar artery dolichoectasia is associated with paramedian pontine infarction. Cerebrovasc Dis 2009;27(2):114-8. PMID 19039214

Laforet P, Petiot P, Nicolino M, et al. Dilative arteriopathy and basilar artery dolichoectasia complicating late-onset Pompe disease. Neurology 2008;70(22):2063-6. PMID 18505979

Lawton MT, Quinones-Hinojosa A, Sanai N, Malek JY, Dowd CF. Combined microsurgical and endovascular management of complex intracranial aneurysms. Neurosurgery 2003;52:263-75. PMID 12535354

Lin N, Brouillard AM, Keigher KM, et al. Utilization of Pipeline embolization device for treatment of ruptured intracranial aneurysms: US multicenter experience. J Neurointerv Surg 2014;pii:neurintsurg-2014-011320. PMID 25230839

Little JR, St Louis P, Weinstein M, Dohn DF. Giant fusiform aneurysms of the cerebral arteries. Stroke 1981;12:183-8. PMID 7233461

Lou M, Caplan LR. Vertebrobasilar dilatative arteriopathy (dolichoectasia). Ann N Y Acad Sci 2010;1184:121-33.** PMID 20146694

Lubicz B, Collignon L, Lefranc F, et al. Circumferential and fusiform intracranial aneurysms: reconstructive endovascular treatment with self-expandable stents. Neuroradiology 2008;50(6):499-507. PMID 18365185

Makos MM, McComb RD, Hart MN, Bennett DR. Alpha-glucosidase deficiency and basilar artery aneurysm: report of a sibship. Ann Neurol 1987;22(5):629-33. PMID 3322184

Mangrum WI, Huston J, Link MJ, et al. Enlarging vertebrobasilar nonsaccular intracranial aneurysms: frequency, predictors, and clinical outcome of growth. J Neurosurgery 2005;102:72-9. PMID 15658099

Martin AJ, Hetts SW, Dillon WP, Higashida RT, Halbach V, Dowd CF, Lawton MT, Saloner D. MR imaging of partially thrombosed cerebral aneurysms: characteristics and evolution. AJNR Am J Neuroradiol 2011;32:346-51. PMID 21087941

Masuda H, Zhuang Y-J, Singh TM, Kawamura K, Murakami M, Zarins CK, Glagov S. Adaptive remodeling of internal elastic lamina and endothelial lining during flow-induced arterial enlargement. Arterioscler Thromb Vasc Biol 1999;19:2298-307. PMID 10521357

Mizutani T, Aruga T. Dolichoectatic intracranial vertebrobasilar dissecting aneurysm. Neurosurgery 1992;31:765-73. PMID 1407466

Moniz E. L'Angiographie Cerebrale. Paris: Masson & Cie, 1934:230-46.

Nakatomi H, Segawa H, Kurata A, et al. Clinicopathological study of intracranial fusiform and dolichoectatic aneurysms : insight on the mechanism of growth. Stroke 2000;31(4):896-900. PMID 10753995

Nishizaki T, Tamaki N, Takeda N, Shirakuni T, Kondoh T, Matsumoto S. Dolichoectatic basilar artery: a review of 23 cases. Stroke 1986;17:1277-81. PMID 3810731

Nossek E, Zumofen DW, Setton A, et al. Treatment of distal anterior cerebral artery aneurysms with the Pipeline Embolization Device. J Clin Neurosci 2017;35:133-8. PMID 27863970

Passero SG, Calchetti B, Bartalini S. Intracranial bleeding in patients with vertebrobasilar dolichoectasia. Stroke 2005;36(7):1421-5. PMID 15976311

Passero SG, Rossi S. Natural history of vertebrobasilar dolichoectasia. Neurology 2008;70:66-72. PMID 18166708

Paulson G, Boesel C, Evans W. Fibromuscular dysplasia. Arch Neurol 1978;35:287-90. PMID 646681

Pico F, Labreuche J, Amarenco P. Pathophysiology, presentation, prognosis, and management of intracranial arterial dolichoectasia. Lancet Neurol 2015;14(8):833-45.** PMID 26194931

Pico F, Labreuche J, Seilhean D, Duyckaerts C, Hauw JJ, Amarenco P. Association of small-vessel disease with dilatative arteriopathy of the brain: neuropathologic evidence. Stroke 2007;38(4):1197-202. PMID 17322076

Pico F, Labreuche J, Touboul PJ, Leys D, Amarenco P. Intracranial arterial dolichoectasia and small-vessel disease in stroke patients. Ann Neurol 2005;57(4):472-9. PMID 15739178

Pritz MB. Ruptured nontraumatic fusiform aneurysm of the cavernous carotid presenting with multiple episodes of epistaxis. Surg Neurol 1994;42:293-6. PMID 7974122

Pumar JM, Arias-Rivas S, Rodríguez-Yáñez M, et al. Using Leo Plus stent as flow diverter and endoluminal remodeling in endovascular treatment of intracranial fusiform aneurysms. J Neurointerv Surg 2013;5 Suppl 3:iii22-7. PMID 23585639

Sacho RH, Saliou G, Kostynskyy A, et al. Natural history and outcome after treatment of unruptured intradural fusiform

aneurysms. Stroke 2014;45:3251-6. PMID 25205312

Sari A, Kandemir S, Kuzeyli K, Dinc H. Giant serpentine aneurysm with acute spontaneous complete thrombosis. AJNR Am J Neuroradiol 2006;27(4):766-8. PMID 16611761

Schievink WI, Prakash UB, Piepgras DG, Mokri B. Alpha 1-antitrypsin deficiency in intracranial aneurysms and cervical artery dissection. Lancet 1994;343(8895):452-3. PMID 7905956

Schievink WI, Torres VE, Wiebers DO, Huston J 3rd. Intracranial arterial dolichoectasia in autosomal dominant polycystic kidney disease. J Am Soc Nephrol 1997;8:1298-303. PMID 9259357

Segal HD, McLaurin RL. Giant serpentine aneurysm: report of two cases. J Neurosurg 1977;46(1):115-20. PMID 830809

Shokunbi MT, Vinters HV, Kaufmann JC. Fusiform intracranial aneurysms. Clinicopathologic features. Surg Neurol 1988;29:263-70. PMID 3353838

Siddiqui A, Chew NS, Miszkiel K. Vertbrobasilar dolichoectasia: a rare cause of obstructive hydrocephalus: a case report. Br J Radiol 2008;81(964):e123-6. PMID 18344271

Smoker WR, Corbett JJ, Gentry LR, Keyes WD, Price MJ, McKusker S. High-resolution computed tomography of the basilar artery: 2. Vertebrobasilar dolichoectasia: clinical-pathologic correlation and review. AJNR 1986;7(1):61-72. PMID 3082145

Steel JG, Thomas HA, Strollo PJ. Fusiform basilar aneurysm as a cause of embolic stroke. Stroke 1982;13:712-6. PMID 7123606

Stehbens, WE. Pathology of the cerebral blood vessels [by] William E. Stehbens. St. Louis: C. V. Mosby, 1972.

Tipping B, de Villiers L, Candy S, Wainwright H. Stroke caused by human immunodeficiency virus-associated intracranial large-vessel aneurysmal vasculopathy. Arch Neurol 2006;63(11):1640-2. PMID 17101835

Vinters HV, De Girolami U, Hauw JJ. Vascular pathology. In: Gray F, De Girolami U, Poirier J, editors. Escourolle & Poirier: Manual of Basic Neuropathology, 4th edition. Philadelphia: Butterworth Heinemann, 2004:82.

Wascher T. Hypothermic circulatory arrest. In: Spetzler RF, Carter P, Hamilton M, editors. Neurovascular Surgery. New York: McGraw-Hill, 1st ed. 1995;191-209.

Watanabe T, Sato K, Yoshimoto T. Basilar artery occlusion caused by thrombosis of atherosclerotic fusiform artery of the basilar artery. Stroke 1994;5:1068-70. PMID 8165681

Weeler RD. Atherosclerotic fusiform aneurysms. In: Adams JH, Duchen LW, editors. Greenfield's neuropathology. New York: Oxford University Press, 5th ed. 1992.

Wehman JC, Hanel RA, Levy EI, Hopkins LN. Giant cerebral aneurysms: endovascular challenges. Neurosurgery 2006;59(5 Suppl 3):S125-38. PMID 17053595

Won YS, Rho MH, Chung EC, et al. Multiple overlapping stent-assisted coiling of complex aneurysms: a single-center experience. Neurol Res 2015;37(3):189-96. PMID 25213599

Yu Y, Moseley I, Pullicino P, McDonald W. The clinical pictures of ectasia of the intracerebral arteries. J Neurol Neurosurg Psychiatry 1982;45:29-36. PMID 7062068

Zhang Y, Tian Z, Sui B, et al. Endovascular treatment of spontaneous intracranial fusiform and dissecting aneurysms: outcomes related to imaging classification of 309 cases. World Neurosurg 2016. [Epub ahead of print] PMID 27890754

**References especially recommended by the author or editor for general reading.

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ICD and OMIM codes

ICD codes

ICD-9:
Unruptured cerebral aneurysm: 437.3
Ruptured cerebral aneurysm: 430
Congenital brain aneurysm: 747.81

ICD-10:
Cerebral aneurysm, nonruptured: I67.1
Subarachnoid haemorrhage, unspecified: I60.9
Congenital cerebral aneurysm (nonruptured): Q28.3

Profile

Age range of presentation

02-05 years
06-12 years
13-18 years
19-44 years
45-64 years
65+ years

Sex preponderance

Carotid distribution:
female>male, >1:1

Vertebrobasilar circulation:
male>female, >1:1

Family history

none

Heredity

none

Population groups selectively affected

none selectively affected

Occupation groups selectively affected

none selectively affected

Differential diagnosis list

saccular aneurysms
tuberculous
carcinomatous
basal granulomatous meningitides
tumors of the brain
migraine
transient ischemic attacks
stroke

stroke-in-evolution
trigeminal neuralgia
hemifacial spasm
pituitary mass

Associated disorders

Atherosclerosis
Hypertension
Diabetes
Dissection
Hyperlipidemia
Saccular aneurysms
Fibromuscular dysplasia
Polycystic kidney disease
Marfan syndrome
Pseudoxanthoma elasticum
Alpha-glucosidase deficiency
Tuberous sclerosis
Neurofibromatosis
Giant-cell arteritis
Syphilis
Coarctation of the aorta

Other topics to consider

Carotid transient ischemic attacks
Intracranial atherosclerosis
Spontaneous carotid and vertebral artery dissection
Unruptured cerebral aneurysms
Vertebrobasilar transient ischemic attacks