Nonatherosclerotic Vascular Disease: Biological and Pathological Basis

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Abstract

Atherosclerosis is the main cause of vascular disease in most cases; in about 10 % of cases, carotid artery disease is related to nonatherosclerotic causes, including several unfrequent pathologies such as Takayasu arteritis, giant cell arteritis, fibromuscular disease, moyamoya syndrome, arterial dissection and extracranial carotid aneurysm. These entities are discussed in the present chapter, with a special focus on pathogenesis. Indeed, the aetiology of these diseases is in most cases not completely known, since related to several factors (genetic, immune and infectious). Early diagnosis, usually leading to a good patient's outcome, is usually achieved after clinical examination and imaging tests.

Keywords

Arterial dissection • Carotid artery disease • Extracranial carotid aneurysm • Fibromuscular dysplasia • Giant cell arteritis • Moyamoya syndrome • Nonatherosclerotic disease • Takayasu arteritis

Introduction

Atherosclerosis is identified as the main cause of vascular disease in around 90 % of cases; in the remaining 10 % of cases, nonatherosclerotic causes include several less common entities, such as Takayasu arteritis, giant cell arteritis,

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fibromuscular disease, moyamoya syndrome, arterial dissection, and extracranial carotid aneurysm. The pathogenesis of these diseases is in most cases unclear or even related to several factors, such as genetic, immune, and infectious, often associated with triggering events. The correct diagnosis is currently achieved after clinical examination and imaging tests; although US, CT, and MRA are useful, catheter angiography represents the gold standard in diagnosing most of these diseases. The majority of patients have a good outcome if the specific disease is diagnosed early.

Takayasu Arteritis (TA)

Takayasu arteritis (TA) is a granulomatous arteritis affecting the aorta and its branches [1]. The first case was reported by Takayasu in 1905 [2]; lately, the disease was more comprehensively described as "pulseless disease" by Shimizu and Sano in 1951 [3]. It generally affects women in the first fourth decades of life (nine females/one male), with a general incidence of 2.6 cases per million per year in the USA and major prevalence in patients of Asiatic origin [4].

The etiology of TA is still unclear; the underlying pathologic process is inflammatory, with several etiologic factors, either infective or autoimmune, having been proposed. The most likely hypothesis is that an unknown stimulus triggers the expression of the 65 kDa heat-shock protein in the aortic tissue which induces the major histocompatibility class I chain-related A (MICA) on vascular cells. The T cells and NK cells recognize MICA on vascular smooth muscle cells and release perforin, resulting in acute vascular inflammation; pro-inflammatory cytokines are also released and increase the recruitment of mononuclear cells within the vascular wall. Th1 lymphocytes drive the formation of giant cells through the production of interferon-γ and activate macrophages with release of VEGF resulting in increased neovascularization and PDGF, resulting in smooth muscle migration and intimal proliferation. The inflammatory cellular infiltrate mainly involves the media and adventitia, usually characterized by three stages: [5, 6].

1. A systemic stage, characterized by signs and symptoms of an acute inflammatory condition, such as fever, arthralgia, anemia, and increased erythrocyte sedimentation rate
2. A vascular inflammatory stage, when vascular stenosis and less frequently aneurysms occur, with corresponding signs and symptoms (stroke, transitory ischemic attacks, hypotensive ischemic retinopathy with visual symptoms, vertebrobasilar ischemia, hypertensive encephalopathy)
3. A burned-out stage, when fibrosis sets, usually associated with remission According to the American College of Rheumatology (ACR) [7], the criteria for assessing the diagnosis are:

- Age at disease onset <40 years
- Claudication of extremities
- Decreased brachial artery pulse
- BP difference >10 mmHg
- Bruit over subclavian arteries or aorta
- Arteriogram abnormality, represented by arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes (changes usually focal or segmental)

A patient shall be said to have TA if at least three of these six criteria are present.

According to the classification proposed by Hata et al. [8], TA can be divided into six types based on angiographic involvement:

- Type I – branches of the aortic arch
- Type IIa – ascending aorta, aortic arch, and its branches
- Type IIb – Type IIa region plus thoracic descending aorta
- Type III – thoracic descending aorta, abdominal aorta, renal arteries, or a combination
- Type IV – abdominal aorta, renal arteries, or both
- Type V – entire aorta and its branches

Hence, only types I, IIa, IIb, and V usually involve the carotid arteries, whereas the other types more commonly involve the thoracic descending and abdominal aorta and the renal arteries.

Angiography is the gold standard for diagnosis, even though color Doppler imaging can be useful in the first part of the diagnostic process, showing the mural thickening of the common carotid arteries (hypoechoic in the early stages and then hyperechoic after the development of fibrosis). CT, and especially MRA due to the young age of these patients, is also extremely helpful, even more during the follow-up [9]. If possible, whole-body MRI technique should be employed in order to exclude other localizations of disease [10].

TA is a chronic relapsing and remitting disorder. The overall 10-year survival rate is approximately 90 %, this rate being reduced in case of major complications, such as stroke, intracranial hemorrhage, and graft stenosis and/or occlusion. For this reason, strict management of traditional cardiovascular risk factors is mandatory in order to minimize secondary cardiovascular complications. Approximately 20 % of patients have a monophasic and self-limited disease; in the remaining 80 % of patients, TA requires immunosuppressive treatment, resulting in remission in around 60 % of cases [11].

Giant Cell Arteritis

Giant cell arteritis (GCA), also known as temporal arteritis, is the most common form of vasculitis occurring in adults, with a prevalence of persons with active or remitted GCA of 200 cases per 100,000 population aged 50 years or older. The female-to-male ratio is 3.7:1 [12]. It is a granulomatous arteritis affecting large- or medium-sized vessels, usually the terminal branches of carotid arteries (more frequently the temporal and ophthalmic arteries). The clinical manifestations of giant cell arteritis result from two different processes. Inflammatory cells infiltrate the arterial wall and cause structural damage, eventually leading to vascular complications. In the majority of patients, a systemic inflammatory syndrome is also present. The systemic and the vascular components of giant cell arteritis seem to have different underlying pathogenetic mechanisms: the vascular inflammation results from abnormal adaptive immune responses, whereas the systemic inflammation likely depends on an excessively activated innate immune system [13]. Despite increased understanding of the inflammatory cascade responsible for the disease process, the initial event triggering the cascade remains uncertain; the adventitia is more likely considered as the site of initial immunologic injury [14].

According to the American College of Rheumatology [15], the criteria for assessing the diagnosis are:

- Age at disease onset >=50 years
- New headache
- Temporal artery abnormality (temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries)
- Elevated erythrocyte sedimentation rate
- Abnormal artery biopsy (biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells)

A patient shall be said to have GCA if at least three of these five criteria are present.

The disease usually has an acute or subacute start, characterized by symptoms related to the acute inflammatory status (fever, night sweats) and headache, jaw pain, and blurred or double vision; if the disease is not early diagnosed, complications may occur, such as blindness and less frequently stroke [9].

The diagnostic process starts with clinical examination and imaging studies. US findings can show swelling of the vessel's wall as a hypoechoic dark halo around the color-coded flow in the artery; the disease is segmental; therefore, its visualization is suitable for localization of the biopsy [16]. CT and MR imaging can show an

abnormal wall enhancement of the affected arteries after contrast media injection in the acute phase, whereas vascular stenosis and aneurysm can be observed in the subacute-chronic phase. In the chronic phase, the differential diagnosis between GCA and atherosclerotic disease may not be easy, but the different localization of the abnormalities (temporal and ophthalmic arteries for GCA, ubiquitarious for atherosclerosis) can lead to the correct diagnosis. Standard test for definitive diagnosis is biopsy of the temporal artery, being more samples needed because the inflammation usually does not involve all parts of the artery [10].

The prognosis for patients with untreated GCA is extremely poor; these patients may suffer blindness or death from myocardial infarction, stroke, or dissecting aortic aneurysm. On the contrary, with prompt and adequate therapy, full recovery is the rule. Symptoms from temporal arteritis usually improve within days of treatment with corticosteroids, except for those symptoms related to an effective vision damage that occurred before initiation of therapy, which are often irreversible. The mean duration of treatment is 2 years [17].

Fibromuscular Dysplasia (FMD)

Fibromuscular dysplasia (FMD) is an angiopathy affecting medium-sized arteries, more frequently in young women of childbearing age. Among patients with identified FMD, renal involvement occurs in 60–75 % and cerebrovascular involvement in 25–30 %; involvement of visceral arteries and arteries of the limbs is less commonly observed (about 9 % and 5 %, respectively); in the case of cerebrovascular localization, the internal carotid artery is more frequently affected (C2 segment) [9, 18].

The etiology of FMD is not known, even though several factors have been considered as involved in its pathogenesis. More in detail, hormonal factors such as estrogen have been proposed; however, although in the US Registry 91 % of registrants were female, clear supporting epidemiological evidence for the role of female hormones beyond the sex and age distribution of FMD has not been described yet. Although no etiologic genes for FMD have been identified, evidence supports a genetic basis for susceptibility to FMD; a few authors reported the potential inheritance pattern to be autosomal dominant with variable penetrance. However, several factors have limited the identification and characterization of genes contributing to FMD, such as disease rarity, variable phenotype, and gene-environment interactions [19]. FMD is usually described in terms of the affected arterial layer and the composition of the lesions; depending on the type of FMD, the narrowing (stenosis) of the artery is caused by an excess of either the fibrous or muscular components of the arterial wall [18].

Many people with FMD do not have any symptoms; symptoms can occur if the stenosis is severe enough to restrict blood flow through the affected artery or if dissection occurs. Symptoms of FMD in the carotid artery include headaches, ringing or "swishing" noise in the ears, or light-headedness; advanced cases of FMD can cause stroke or a transient ischemic attack [9].

FMD is often accidentally diagnosed when the beaded appearance in the arteries is observed during examinations performed for other reasons. Noninvasive imaging studies such as duplex ultrasound, MRA, and CTA can be used to confirm the diagnosis of FMD and determine the extent of the lesions. In general, angiographic studies are performed only when the diagnosis is not clear or if the patient requires a therapeutic procedure such as a balloon angioplasty [10].

The most common type of FMD is the medial fibrodysplasia (75–80 % of cases), affecting the tunica media; it is characterized by areas of vessel stenosis alternating with areas of ectasia, resulting in a classic "beads on a string" appearance on angiograms. Intimal and perimedial fibroplasias are less common (10 % of cases, respectively), caused by collagen deposits in the intima and in the outer portion of the tunica media; in the intimal type, a concentric, smooth, narrowing (without beads) appearance of arteries can be observed,

whereas the perimedial type has a "beads on a string" appearance, but with "beads showing a smaller diameter in comparison to the normal vessel" [20].

When a patient is diagnosed with FMD in the carotid arteries, additional imaging studies may be obtained to evaluate the other blood vessels, especially vertebral arteries, Willis circle, and renal arteries.

When FMD is present without any symptoms, it usually does not require intervention; risk factors for vascular disease, such as high blood pressure, diabetes, and high cholesterol, should be evaluated and treated, and imaging studies should be performed at regular intervals to evaluate the disease progression. Angioplasty is recommended for patients with FMD of the internal carotid artery who experience TIAs or stroke related to severe arterial narrowing. Stenting is rarely necessary only, in the case of carotid or vertebral artery dissection or carotid aneurysm. Surgery depends upon the location and the extent of disease and consists in removing or bypassing the affected portion of the artery to restore normal blood flow; reconstructive surgery may be recommended for patients with an aneurysm of the internal carotid or vertebral arteries [19, 21].

Moyamoya Syndrome

Moyamoya is a disease causing intimal thickening of the walls and stenosis of the terminal branches of the internal carotid arteries bilaterally; the term "moyamoya" (Japanese for "puff of smoke") refers to the angiographic appearance of abnormal vascular collateral networks that develop adjacent to the stenotic vessels [22, 23]. Moyamoya disease occurs primarily in Asians; the female-to-male ratio of moyamoya disease is 1.8:1. The disease has two peaks of incidence in the first and fourth decades of life [8, 24]. The cause of moyamoya disease is not clear. The disease is believed to be hereditary; a few authors suggested that the transmission may be autosomal dominant with incomplete penetrance based on age and genomic imprinting factors [25].

According to the Classification of the Japanese Health Ministry, there are four clinical forms of the moyamoya disease: ischemic, hemorrhagic, epileptic, and "other." Clinical presentation in children is usually ischemic, related to occlusion of internal carotid artery or one of the branches of the Willis circle, resulting in paresis, sensory impairment, involuntary movements, headaches, dizziness, or seizures; mental retardation is often present. In adults, the hemorrhagic form, especially with subarachnoid hemorrhage, is more frequently observed as a result of hemorrhage of fragile vessels [9, 26].

In the affected cerebral vessels, pathological examinations do not show atherosclerotic or inflammatory lesions. Currently, the major proteins believed to be involved in the pathogenesis are vascular endothelial growth factor (VEGF), basic fibroblast growth factor (bFGF), hepatocyte growth factor (HGF), transforming growth factor-β (TGF-β), and granulocyte colony-stimulating factor (G-CSF) [27]. The cause of stenosis is the overgrowth of the smooth muscle layer, with thrombotic changes. The disease leads to different degrees of stenosis and occlusions of arteries of the anterior part of the Willis circle and to the development of the collateral vasculature. The vessels of the collateral circulation are formed as a result of the widening of the existing vessels or development of new perforating arteries. There are three main pathways of collateral circulation – parenchymal, meningeal, and transdural. In the vessels of the collateral circulation, there may appear thrombotic changes, which are the cause of ischemic symptoms. An increased blood flow through thin collateral walls during stress, as well as the presence of microaneurysms, is the probable cause of intracranial hemorrhages [28].

Before CTA or MRA were introduced to a wide clinical practice, the final diagnosis of the vascular changes was based on cerebral angiography. Although CT examination is sufficient to diagnose ischemic or hemorrhagic stroke in the course of the disease, MRA can be considered as the first imaging technique for definitive diagnosis, considering the young age of the patients [29]. To support the diagnosis, the following findings should be observed:

- Stenosis or occlusion at the terminal portion of the internal carotid artery or the proximal portion of the anterior or middle cerebral arteries
- Abnormal vascular networks in the vicinity of the occlusive or stenotic areas
- Bilaterality of the findings (although some patients may initially present with unilateral involvement)

Mortality rates from moyamoya disease are around 10 % in adults and 4 % in children. About 50–60 % of affected patients experience a gradual deterioration of cognitive function, likely due to recurrent strokes. The outcome of the disease depends on the severity of the hemorrhage, the prognosis on recurrent attacks. Cases of mild clinical course are normally treated conservatively. In severe cases, surgery is indicated, including direct anastomoses, indirect procedures, and combined therapies [30].

Extracranial Internal Carotid Artery Aneurysms

Aneurysms of the extracranial internal carotid artery are extremely rare, being more commonly observed in males (prevalence ranging from 0.1 % to 3.7 %); most of them are of atherosclerotic origin. In case of nonatherosclerotic etiology, they can be congenital (due to collagen disorders, FMD), infectious (TBC, HIV, mycotic), and posttraumatic. True aneurysms must be differentiated from pseudoaneurysms, which are usually iatrogenic (due to carotid puncture or endarterectomy). For a definitive diagnosis of aneurysm, the diameter of the ectatic artery must increase of at least 50 %, if compared to the normal vessel diameter [9, 10].

Most of these aneurysms are asymptomatic, whereas TIA or stroke is the most common cause of hospital admission, related to embolization from the aneurysmal contents. Spontaneous rupture or bleeding is very rare, but fatal complications are seen particularly in cases with mycotic aneurysms [31].

The mainstay of treatment of extracranial carotid artery aneurysms is surgical repair; it consists in resecting the portion of the carotid artery involved with the aneurysm and performing a bypass from the normal artery below the aneurysm to the normal artery above the aneurysm. Stroke and cranial nerve injuries (particularly XII pair) are potential risks for surgical treatment of aneurysms with a diameter of more than 3 cm and/or extending to the skull base; endovascular treatment may be preferred in appropriate cases as an alternative to surgical therapy; since there is no risk of cranial nerve injury, it allows to treat lesions that are hard to reach surgically, and there is no need for general anesthesia [31].

Craniocervical Arterial Dissection

Craniocervical artery dissection (CCAD) is one of the major causes of ischemic symptoms in young adults. It has a prevalence of 2.6 cases per 100, 000 persons per year; vertebral artery dissections are less common than carotid artery dissections. Genetic factors such as constitutional weakness of the arterial wall (such as in FMD or in monogenic connective tissue disease, mainly Ehlers-Danlos syndrome or Marfan's syndrome) might have a role in the pathophysiology of CCAD; environmental factors such as minor trauma act as a trigger [32]. Arterial dissections begin with a tear in the intima or media resulting in bleeding in the arterial wall; expansion of the wall by intramural blood causes compression and narrowing of the lumen, contributing to the formation of an intraluminal thrombus. The intramural hematoma can create a false lumen that might reconnect with the true lumen and forms parallel flow; alternatively, wall rupture through the adventitia causes extraluminal bleeding. CCAD can be asymptomatic and discovered during routine examinations. The most common symptom is pain in the head and neck and in the region of dissections, usually following a minor trauma. If the dissection compromises the arterial lumen or causes thrombus formation, clinical symptoms are related to ischemia; the closer the dissection to the brain is, the higher the possibility of brain infarction is present. On the contrary, if the dissection is more extracranial, the probability of local symptoms from space-occupying lesions is higher;

bleeding in the subadventitial wall results in compression of the adjacent structures, such as lower cranial nerves. Patients with subadventitial intracranial dissections often present with subarachnoid hemorrhage [32, 33].

CCAD can be noninvasively diagnosed by performing MRA and CTA [34]; color Doppler flow imaging showed good results in visualization of dissections, even though the main limitation is represented by intracranial dissection (which is the most common site of localization). The traditional method for CCAD diagnosis is catheter angiography that may show irregular luminal narrowing occlusion, pseudoaneurysm, intimal flap, double lumen, or distal branch occlusion [35].

There is no general consensus regarding optimal management of internal carotid artery dissection, but the choice among medical, endovascular, and surgical options may depend on the type of and anatomic location of dissection; while surgery has a limited role, candidates for angioplasty and stent placement include patients with persistent ischemic symptoms despite adequate anticoagulation, patients with a contraindication to anticoagulant therapy, and patients with significantly compromised cerebral blood flow [36, 37]. Overall, the prognosis for spontaneous internal carotid artery dissection is favorable, with about 75 % of patients making a good recovery; the reported mortality is less than 5 %. The functional outcome is also generally good, and recurrence of cerebral ischemia and carotid artery dissection is rare; the risk of recurrence is highest in the first month and then remains around 1 % per year for about a decade. Headache may persist for a long time, in some cases for years, after the dissection [38].

Summary

Atherosclerosis is the main cause of vascular disease, being responsible in around 90 % of cases of carotid disease; in the remaining cases, nonatherosclerotic causes such as Takayasu arteritis, giant cell arteritis, fibromuscular disease, moyamoya syndrome, arterial dissection and extracranial carotid aneurysm must be taken into account. These less common diseases have some similarities, such as an unclear pathogenesis and a good outcome if the specific disease is diagnosed early.

References

1. Jennette JC (1994) Nomenclature of systemic vasculitis. Arthritis Rheum 37:187–192
2. Numano F et al (1996) Takayasu arteritis-five doctor in the history of Takayasu arteritis. Int J Cardiol 54:S1–S10
3. Shimizu K, Sano K (1951) Pulseless disease. J Neuropathol Clin Neurol 1.7–1.47
4. Hall S et al (1985) Takayasu arteritis. A study of 32 North American patients. Medicine 64:89–99
5. Arnaud L, Haroche J, Mathian A (2011) Pathogenesis of Takayasu's arteritis: a 2011 update. Autoimmun Rev 11(1):61–67
6. Matsunaga N, Hayashi K, Sakamoto I et al (1997) Takayasu arteritis: protean radiologic manifestations and diagnosis. Radiographics 17:579–594
7. Arend WP et al (1990) The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 33:1129–1134
8. Hata A, Noda M, Moriwaky R et al (1996) Angiographic findings of Takayasu arteritis: new classification. Int J Cardiol 54(suppl):S155–S163
9. Bartels E, Bartels S, Poppert H (eds) (2012) New trends in neurosonology and cerebral hemodynamics-an update. Perspect Med 1:244–249
10. Catalano C, Anzidei M, Napoli A (editors): (2013) Cardiovascular CT and MR imaging. Springer-Verlag Mailand Italia Srl; ISBN 978-88-470-2868-5.
11. Phillip R, Luqmani R (2008) Mortality in systemic vasculitis: a systematic review. Clin Exp Rheumatol 26(5 Suppl 51):S94–S104
12. Salvarani C, Gabriel SE, O'Fallon WM, Hunder GG (1995) The incidence of giant cell arteritis in Olmsted County, Minnesota: apparent fluctuations in a cyclic pattern. Ann Intern Med 123(3):192–194
13. Weyand CM, Goronzy JJ (2003) Giant-cell arteritis and polymyalgia rheumatica. Ann Intern Med 139:505–515
14. Weyand CM, Goronzy JJ (2000) Pathogenic principles in giant cell arteritis. Int J Cardiol 75(Suppl 1):S9–S15, discussion S17–S9
15. Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH et al (1990) The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 33:1122–1128
16. Schmidt WA, Kraft HE, Vorpahl K (1997) Color duplex ultrasonography in the diagnosis of temporal arteritis. N Engl J Med 337:1336–1342

17. Borchers AT, Gershwin ME (2012) Giant cell arteritis: a review of classification, pathophysiology, geoepidemiology and treatment. Autoimmun Rev 11(6–7):A544–A554
18. Luscher TF, Lie JT, Stanson AW et al (1987) Arterial fibromuscular dysplasia. Mayo Clin Proc 62(10):931–952
19. Olin JW, Gornik HL, Bacharach JM et al (2014) Fibromuscular dysplasia: state of the science and critical unanswered questions: a scientific statement from the American Heart Association. Circulation 129(9):1048–1078
20. Begelman SM, Olin JW (2000) Fibromuscular dysplasia. Curr Opin Rheumatol 12(1):41–47
21. Olin JW, Pierce M (2008) Contemporary management of fibromuscular dysplasia. Curr Opin Cardiol 23(6):527–536
22. Kuroda S, Houkin K (2008) Moyamoya disease: current concepts and future perspectives. Lancet Neurol 7(11):1056–1066
23. Scott R, Smith ER (2009) Moyamoya disease and moyamoya syndrome. N Engl J Med 360:1226–1237
24. Uchino K, Johnson A, Claiborne S, Tirschwell DL (2005) Moyamoya disease in Washington State and California. Neurology 65:956–958
25. Mineharu Y, Takenaka K, Yamakawa H et al (2006) Inheritance pattern of familial moyamoya disease: autosomal dominant mode and genomic imprinting. J Neurol Neurosurg Psychiatry 77(9):1025–1029
26. Shamim S, Kumar J, Jamalvi SW et al (2008) Moya Moya disease in a child. J Coll Physicians Surg Pak 18(4):252–253
27. Weinberg DG, Arnaout OM, Rahme RJ et al (2011) Moyamoya disease: a review of histopathology, biochemistry, and genetics. Neurosurg Focus 30(6):E20
28. Lim M, Cheshier S, Steinberg G (2006) New vessel formation in the central nervous system during tumor growth, vascular malformations, and Moyamoya. Curr Neurovasc Res 3(3):237–245
29. Tarasów E, Kułakowska A, Łukasiewicz A et al (2011) Moyamoya disease: diagnostic imaging. Pol J Radiol 76(1):73–79
30. Burke GM, Burke AM, Sherma AK et al (2009) Moyamoya disease: a summary. Neurosurg Focus 26(4):E11
31. Yetkin U, Yürekli, Gürbüz A (2006) Extracranial internal carotid artery aneurysms. Internet J Thorac Cardiovasc Surg 10(2)
32. Debette S, Leys D (2009) Cervical artery dissections: predisposing factors, diagnosis and outcome. Lancet Neurol 8:668–678
33. Caplan LR (2008) Dissections of brain-supplying arteries. Nat Clin Pract Neurol 4:34–42
34. Debette S, Provenzale JM (2009) MRI and MRA for evaluation of dissection of craniocerebral arteries: lessons from the medical literature. Emerg Radiol 16:185–193
35. Houser OW, Mokri B, Sundt TM et al (1984) Spontaneous cervical cephalic arterial dissection and its residuum: angiographic spectrum. Am J Neuroradiol 5:27–34
36. Baumgartner RW (2010) Management of spontaneous dissection of the cervical carotid artery. Acta Neurochir Suppl 107:57–61
37. Xianjun H, Zhiming Z (2013) A systematic review of endovascular management of internal carotid artery dissections. Interv Neurol 1(3–4):164–170
38. Schievink WI (2001) Spontaneous dissection of the carotid and vertebral arteries. N Engl J Med 344(12):898–906