Spontaneous Mirror Dissections of Cervicocephalic Arteries

Pathomechanical Considerations

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Summary

While so-called twin or mirror aneurysms constitute an established subgroup of multiple aneurysms, simultaneous spontaneous mirror dissections of cervicocephalic artery have not yet been reported as a particular entity. Among the patients treated at our institution since 1989, we identified 74 patients with spontaneous, non-traumatic dissections. Six of these cases presented with simultaneous bilateral dissections and four of the six patients had mirror dissections. Acute or chronic headache was present in all four cases. Additional clinical presentations consisted of impaired consciousness, cranial nerve palsy, and tinnitus. Angiography revealed irregular stenosis, dilatation or aneurysms located in the cervical ICA (internal carotid artery), VA (vertebral artery), or MCA (middle cerebral artery) without evident location bias. Although mirror dissections seems to be an exceptional finding, they may shed light on the vulnerability of different arterial segments to specific diseases. Similar to arterial aneurysm formation, pathogenesis of mirror dissection may involve an underlying "shared defect" in the endothelial cells, since these cells demonstrate a bilateral distribution during embryological development. This particular distribution therefore also provides a chronicle trail of the first trigger striking during embryonic development and demonstrates the segmental vulnerability to highly specific triggers.

Introduction

Intracranial twin or mirror aneurysms are defined as saccular-type aneurysms located at symmetric sites of corresponding arteries. This aneurysm type is encountered in approximately 10% of patients with multiple aneurysms and constitutes a well established subgroup of multiple aneurysms¹⁻³. These mirror aneurysms presumably have a different etiology than other non-mirror like multiple intracranial saccular aneurysms and demonstrate a tendency of early rupture without extrinsic risk factors⁴. Similarly, spontaneous dissections of cervicocephalic vessels occur bilaterally in between 10 and 40 percent of all patients harbouring this disease^{5,6}. However, up to now there are no reports on the mirror occurrence of these spontaneous, non-traumatic dissections. The aim of this paper is, therefore, to determine the rate of occurrence of mirror dissections of cervicocephalic arteries, i.e. dissections in homologous arterial segments on both sides and to describe the clinical and pathomechanical considerations of this type of vascular disease.

Methods

Patients were selected after a retrospective search through the neurovascular databank of the Bicêtre hospital from 1989 to January 2006 employing the search term "dissection". Pa-

tients with traumatic dissections and saccular aneurysms including mirror aneurysms, were excluded from the study. 74 cases with dissections of the cervicocephalic artery were found, of which 23 presented with fusiform lumen. Six patients harbored bilateral dissections and four were found to have bilateral mirror dissections. Mirror dissection was defined as simultaneous dissections at symmetric sites of corresponding cervicocephalic arteries and segments. Clinical and imaging findings of these four patients were reviewed based on their clinical charts, MRI and DSA data.

Case Reports

Case 1

A 57-year-old male with poorly controlled hypertension presented with persistent bitemporal pulsatile headache since six weeks and sudden onset of left-sided ptosis. Recent infection was denied, and family history was unremarkable. Physical examination revealed a left-sided ptosis and neck hyperesthesia. A clinical diagnosis of Horner's syndrome and ICA dissection on the left side was made. However, MRI revealed an intramural hematoma of the cervical and

Figure 1 Case 1. MRI (A-D, T1-weighted fat-suppressed images (SPIR) pre contrast) reveals an intramural crescent shaped hyperintensity indicative of mural hematoma on the cervical segments of both internal carotid arteries (ICA). Lateral projection of the left (E) and right (F) common carotid artery angiography demonstrates irregular stenosis and dilatation of the ICA lumen which confirmed the diagnosis of bilateral mirror-like segmental ICA dissections. A pseudoaneurysm was opacified near the cranial base on the left ICA.

petrous segment of the ICA bilaterally, suggestive of bilateral internal cerebral artery (ICA) dissections. Angiography of the left ICA demonstrated an irregular stenosis beginning several centimeters above the left common carotid artery bifurcation and ending at the cavernous segment. An aneurysm was present in the cervical segment of the left ICA. On the right side, a symmetrical irregular cervical ICA stenosis, followed by irregular dilatation of the petrous segment, was revealed (Figure 1). Bilaterally, the vertebral arteries were normal except that the left one originated directly from the aortic arch.

There were no signs of atherosclerosis or generalized vessel wall disease (such as fibromuscular dysplasia) present. Therapy was started with anticoagulation medication followed by antiplatelet therapy. The patient showed no change of symptoms at 3 month follow-up and aspirine was still kept at the time of this report.

Case 2

A 51-year-old female presented with complaints of regular migraine attacks after having fallen down the stairs four years prior to admission. One month before admission, she suf-

Figure 2 Case 3. (A) CT scan reveals an extensive frontotemporal hematoma and diffuse SAH. AP view of right (B) and left (C) ICA angiography demonstrates a pseudoaneurysm on the right MCA and a fusiform aneurysm on the left, thereby indicating bilateral segmental dissections of the MCA. Notice the diffuse irregular stenosis in the anterior circulation bilaterally, corresponding to atheromatous changes as verified during surgery.

ferred from a sudden onset of severe headache during work, that was followed by bilateral neck pain. The severity of the pain fluctuated and one week later, she experienced a right pulsatile tinnitus which was audible on auscultation. Cranial CT and cervical ultrasound were normal, however, MRI, including MRA, revealed bilateral cervical dissections. The tinnitus alleviated after admission but a visual field defect emerged. Angiography demonstrated symmetric aneurysms on the arterial wall of the right and left ICAs, with a diameter of 2-3 mm. The patient was treated with aspirine; control angiography one year later revealed complete obliteration of the dissecting aneurysm on the right ICA. The left one remained unchanged. The patient was free of symptoms at three years follow-up and aspirine treatment was maintained.

Case 3

A 60-year-old female with well controlled hypertension presented with a sudden onset of headache and subsequent loss of consciousness. Physical examination revealed GCS (Glasgow coma scale) 6, a right-sided mydriasis and left hemiplegia. Cranial CT revealed a large right frontotemporal hematoma and diffuse subarachnoid hemorrhage. Immediate angiography revealed irregular stenosis of the right middle and anterior cerebral artery. A pseudoa-

neurysm in the right MCA bifurcation was believed to be the cause of hemorrhage. A fusiform dilatation accompanied with irregular stenosis of the left middle and anterior cerebral artery was found and, in addition, a partially thrombosed basilar tip aneurysm was revealed (Figure 2). The patient was sent for surgical evacuation of the hematoma and clipping of the presumably ruptured right MCA aneurysm. During operation, a diffuse atheroma, corresponding to the diffuse vascular stenosis on the angiogram, was noted. The right middle sylvian aneurysm turned out to be a dissecting aneurysm with a wide neck. The hematoma was evacuated and the aneurysm was clipped even though intra-operative rupture had occurred when dissecting the false sac of the aneurysm. The patient failed to recover from the primary hemorrhage and died two weeks later. No autopsy could be obtained.

Case 4

A 40-year-old obese female presented with sudden onset of headache and right-sided hemiparesis. Her medical history included systemic lupus erythematoses, thrombocytopenia and right leg phlebitis complicated by chronic infection. She was treated with steroids and anticoagulation. Mental retardation was present since birth but had never been further evaluated. The patient had a sister with dysthy-

roidea. On physical examination, a mild right hemiparesis and a left facial paralysis were found. She was confused when admitted and presented with a GCS of eleven. Cranial CT revealed intraventricular hemorrhage and moderate hydrocephalus. Immediate angiography revealed a pseudoaneurysm with evident contrast stagnation at the C1-level of the left vertebral artery. The right vertebral artery showed tandem pearl and string sign from the level of C2 to the proximal intradural segment. Bilateral renal angiogram was normal. Thus, a bilateral vertebral dissection was diagnosed and the hemorrhage was believed to result from iatrogenic over-anticoagulation, which was verified by lab work-ups. The patient was therefore treated conservatively. She recovered quickly from the event with a GCS of 15 three days later and was transferred to the referring hospital. A further follow-up is not available for this patient.

Discussion

Bilateral cervicocephalic artery dissections may be considered uncommon but are not rare. About 5 to 10% of carotid and 38% of vertebral artery dissections are bilateral^{7,8}. In our series we found bilateral spontaneous dissection in 8.1% of all patients harbouring this disease. A high number of these patients presented with mirror dissections in homologous arterial segments of both sides as a specific type of bilateral spontaneous dissections. This mirror bilateral dissection has been reported previously in three cases with strangulation trauma for the cervical ICA, although its true incidence might be higher since trauma, in particular strangulation might be hidden by the patient⁹. Spontaneous symmetrical dissection has not yet been reported upon, even not in patients with underlying generalized vessel wall diseases such as present in collagen disorders, fibromuscular dysplasia or neurofibromatosis. Although the number of overall cases is limited, such a high rate, as reported in our series is unlikely to occur just by chance alone or due to a referral bias of our hospital, and, therefore, should raise more attention regarding possible pathomechanisms of this particular type of dissection.

Mirror dissections were present in our series both in the ICA and the vertebral system without a preferenced location concerning the af-

fected segment or the affected vessel. None of these patients harboured a vessel wall disease. Although the number of cases in this manuscript is too small to draw any definite conclusions, mirror dissections seem to differ from mirror aneurysms in regard of the geotropism: although the latter can be found in different vascular segments of the intracranial arterial system, there seems to be a predominance of mirror aneurysms in the middle cerebral artery which has not been found in mirror dissections.

An underlying vessel wall defect, initialized by endothelial dysfunction¹⁰, has widely been accepted in spontaneous multivessel dissection¹¹⁻¹², although the exact type of arteriopathy remains unknown in most cases¹³⁻¹⁵. As pointed out above, these vessel wall defects do not give rise to bilateral mirror type of dissections. Therefore, in the case of spontaneous mirror dissections, the mechanism for symmetric involvement of vessels and sparing of certain segments is elusive. A study on segmental identity and vulnerability in cerebral arteries might shed light on this issue¹⁶. From an embryological point of view, the morphologically contiguous cerebral arteries are actually consisted by segments with different evolutionary backgrounds. Such differences are imprinted on the arterial anatomy and potentially readable. In other words, these invisible heterogenous discontinuous segments establish an apparently homogenous anatomical, histological and hemodynamic system. Thus, two gross mechanisms can be created to explain segmental impact: 1) a defect occurs early in a precursor vessel cell, thus the structural vulnerability has a clonal character, which can subsequently be revealed by a second non-specific trigger. 2) a group of normal cells share a specific common recognisable identity and a second trigger that is specific to this identity can strike the particular segment. The latter is involved in the immune type of recognition and autoimmune diseases, whereas the former might play a role in mirror dissections. This second trigger might be associated to infections, since clinical practice reveals an epidemic occurrence of patients presenting with dissections.

It seems obvious that, during cephalic vasculogenesis, a defect harboured by embryonic cells with bilateral distribution (such as mesodermal/neural crest stem cells) might be transmitted to the next generation of cells. The de-

scendant cells might be topographically rearranged and regrouped to symmetric vessel segments on different sides, thereby explaining the mirror appearance¹⁷. Over time, the dormant defect within the vulnerable endothelial cell might get symptomatic due to secondary

triggering factors such as chronic inflammation, atherosclerosis, or the role played by vasa vasorum. In this meaning, mirror dissections not only reveal segmental vulnerability in cerebral arteries, but is also a timemarker of the first trigger striking.

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