Carotid artery dissection and sports

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Summary

Carotid artery dissection (CAD) is a potentially life-threatening disease and mainly affects young and middle-aged people. In recent years dissections have been diagnosed more frequently, probably because new imaging techniques allow more reliable diagnosis. The cause of so-called spontaneous CAD is largely unexplained. It most likely involves an underlying abnormality of the vessel wall and triggering factors such as infection, minor trauma or sporting activities. Various sporting activities have been reported in association with CAD. Nevertheless, the causal relationship between sporting activities and CAD often remains doubtful. Traumatic CAD mainly complicates severe blunt head or neck traumas, which are often due to motor vehicle or sport accidents.

The clinical presentations of CAD are highly variable. Headache, neck and facial pain, pulsatile tinnitus, Horner's syndrome, cranial nerve palsies, stroke, TIA, retinal infarction, or transient monocular blindness, may be present in isolation or in various combinations. Ischaemic symptoms are often preceded by local symptoms or signs. The knowledge and recognition of these symptoms and the often subtle signs are the keys to an early diagnosis and antithrombotic treatment of ICAD before severe ischaemic complications occur. The long-term prognosis of CAD is favourable in the majority of patients. Clinical functional outcome mainly depends on the initial stroke severity. Symptomatic recurrent dissections and recurrent strokes are rare.

Introduction

Cervical artery dissection and cardiac embolism are the most frequent causes of stroke in young and middle-aged adults.

Carotid artery dissection (CAD) is considered to be either spontaneous or traumatic in origin. Traumatic CAD mainly complicates severe blunt head or neck traumas, which are often due to motor vehicle or sports accidents. The term "blunt cerebrovascular injury" is used in the literature for patients who developed lesions of the carotid or vertebral artery after a blunt trauma [1].

So-called spontaneous CAD occurs without a precipitating event or may be preceded by a minor trauma, sporting or recreational activities, sneezing, coughing, vomiting, brisk head movements or extensive or prolonged hyperextension and rotation of the neck. These activities are not usually followed by CAD and are therefore considered to be triggering events. Nevertheless, the causal relationship between minor trauma and sporting activities and CAD often remains doubtful, and patients with dissection following minor trauma or sporting activities are usually classified as spontaneous CAD. However, the classification of CAD into traumatic and spontaneous forms may be arbitrary in some cases.

Epidemiology

Carotid and vertebral artery dissection accounts for up to 25% of strokes in patients younger than 45 years of age [2].

The incidence of carotid artery dissection was reported to be 2.6–2.9 per 100 000/year [3, 4].

However, the true incidence of CAD is likely to be higher because these studies did not take into account dissections without ischaemic events (up to 30% of carotid dissections). Furthermore, an unknown number of dissections occur asymptomatically and dissections may be overlooked because the clinical manifestations are not necessarily familiar to many physicians. The mean age is between 39 and 45 years. Men are on average five years older than women. In 10 to 20% of patients multivessel CAD occur simultaneously. Multivessel dissections are more frequent in women than in men [5].

The true incidence of CAD after blunt cervicocerebral trauma is underrecognised since symptoms and signs of CAD may be masked by the symptoms of underlying head or spine trauma.

Blunt carotid artery injury was first described in 1872, and there were only 96 cases reported up to 1980 [6]. Studies from the late 1980s reported an incidence of 0.08% [1]. Later on, the awareness of the severe complications of traumatic CAD such as embolic stroke increased. Subsequent studies have detected more pa-

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tients with traumatic CAD, reporting incidence rates of up to 0.5% for all blunt trauma victims and up to 0.67% for motor vehicle accident patients [7, 8]. The largest series to date consisted of 18,233 patients with blunt trauma [9]. In the first retrospective part of the study over a period of six years the incidence of blunt cerebrovascular injury was 0.1%. The subsequent prospective study period over 2.5 years revealed a 0.86% blunt cerebrovascular injury rate. Carotid dissection was present in the majority of these patients. Another retrospective study evaluated the medical records of 3,342 patients admitted to a trauma centre [10]. The incidence of internal carotid dissection was 0.21% for all trauma patients and 3.2% for patients with severe injuries.

Pathogenesis

CAD is thought to arise from an intimal tear leading to penetration of circulating blood into the vessel wall and the formation of a haematoma with variable longitudinal or circular extension. In addition, some CAD may be due to a primary intramural haematoma. Subintimal dissection tends to cause luminal narrowing or occlusion, whereas subadventitial dissection may cause a dissecting aneurysm. Combined forms with stenosis and aneurysmal dilatation may occur. There are three main consequences of dissection:

– Compression or stretching, due to the enlarged artery or aneurysm, causes local symptoms such as pain, Horner's syndrome and cranial nerve palsies.
– Retinal or focal cerebral ischaemia caused by embolisation of thrombus overlying the dissection to the retinal artery or the intracranial vessels. Less frequently, if the collateral circulation is insufficient, hypoperfusion may lead to haemodynamically induced infarction.
– Subadventitial rupture of the dissected artery (only in intracranial dissections) can cause subarachnoid haemorrhage. This may be because the wall of the intracranial segment is thinner than that of the extracranial arteries.

Aetiology of traumatic carotid dissection

Crissey et al. described four mechanisms leading to injury of the carotid artery: 1) neck hyperextension associated with rotation, 2) direct blow to the neck, 3) blunt intra-oral trauma, and 4) basilar skull fracture involving the carotid canal [11]. A subsequent study emphasised the predominant role of neck hyperextension with rotation, which is particularly common in motor vehicle accidents [12]. Basilar skull fracture, through the petrous segment of the carotid canal is a frequent cause of unrecognised carotid artery injuries [13, 14].

The main trauma leading to CAD is a motor vehicle accident [15]. Patients with combined head, facial and cervical spine injuries are at increased risk for CAD.

Table 1
Sports, recreational activities and other conditions considered as potential precipitating mechanisms of carotid artery dissection.

Sporting activities: karate, judo, skiing, football, rugby, yoga, jogging, ice hockey, swimming, horse riding, golf tennis, cycling, basket ball, diving…

Recreational activities: roller coaster, bungee jumping

Other conditions: hair dresser or dentist (neck hyperextension), prolonged telephone call with forced head rotation

Medical interventions: intubation, catheter angiography, bronchoscopy, chiropractor manipulation

Delivery, post-partum period

Coughing, vomiting, sneezing

CAD has been reported in association with various sports injuries, sporting and recreational activities. The most frequent sporting activities preceding CAD include judo, skiing, yoga, ice hockey, rowing, wrestling, horse riding, soccer and jogging (table 1). CAD has also been thought to be roller coaster induced or triggered by a prolonged phone call with forced head rotation [16, 17].

Aetiology of spontaneous carotid artery dissection

An underlying abnormality of the vessel wall is thought to be involved in the pathogenesis of spontaneous CAD. Reports of familial CAD and dissection associated with vessel tortuosity, fibromuscular dysplasia, intracranial aneurysms, aortic root enlargement, and hereditary connective tissue disorders support the concept that predisposing genetic abnormalities play some role. Skin biopsies of patients with carotid artery dissections have shown ultrastructural abnormalities in the dermal connective tissue, including enlarged or irregular collagen fibrils and pronounced elastic fibre fragmentation, suggesting a predisposing systemic disorder [18]. In an ultrasound study, common artery diameter change during the cardiac cycle was significantly higher in carotid dissection than in controls. These findings all suggest that a generalised defect of the extracellular matrix is present in at least some patients with spontaneous CAD. However, extensive genetic studies for mutations in extracellular matrix molecules have been negative [19].

CAD is often preceded by a minor head or neck trauma and in 25% to 58% of the patients by a usually mild, mainly respiratory infection [20]. These observations strongly suggest that mechanical factors and inflammation may be involved in the pathogenesis of spontaneous CAD. However, the pathogenic links between inflammation and CAD remain widely unknown.

Clinical manifestations

The clinical manifestations of traumatic and spontaneous CAD are highly variable. It is unclear, whether the frequency of some clinical symptoms and signs differ between traumatic and spontaneous CAD. Headache,

Table 2
Clinical findings in patients with carotid artery dissection.

Local symptoms or signs	Ischaemic manifestations
Headache (often frontotemporal), facial pain (often periorbital, retroorbital), anterior neck pain	Amaurosis fugax
Horner's syndrome	Retinal infarction (rare)
Pulsatile tinnitus	TIA
Cranial nerve palsies (mainly cranial nerves IX to XII)	Stroke (mainly in the middle cerebral artery territory)

neck and facial pain, pulsatile tinnitus, Horner's syndrome, cranial nerve palsies, stroke, TIA, retinal infarction, amaurosis fugax may be present in isolation or in various combinations (table 2) [5]. The classical triad of unilateral headache and/or neck pain and an ipsilateral Horner's syndrome followed by ischaemic symptoms from the ipsilateral hemisphere or retina occurs in less than one third of patients [21]. Ischaemic symptoms are often preceded by local symptoms or signs. The knowledge and recognition of these often subtle symptoms and signs is the key to an early diagnosis and treatment of CAD before ischaemic complications occur.

Local symptoms and signs

The most common symptom is headache of acute onset. It is usually an early manifestation, most frequently lo-

Figure 1
Most frequent locations of headache, facial pain and neck pain in patients with carotid artery dissection (courtesy to Professor Matthias Sturzenegger, Department of Neurology, University Hospital Berne).
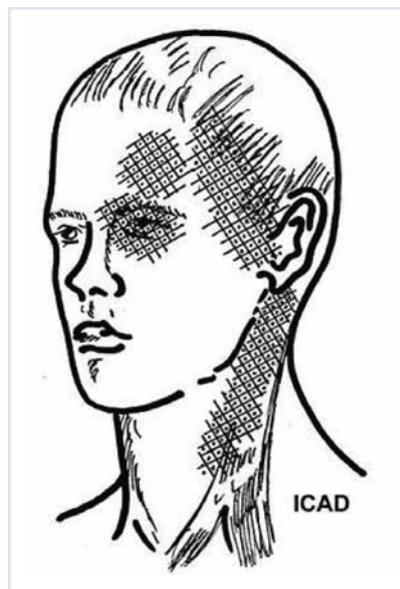

Figure 2
Left-sided Horner's syndrome with miosis and ptosis in a patient with internal carotid artery dissection.

calized in the periorbital and fronto-temporal regions and/or in the upper anterolateral cervical region, ipsilateral to the dissection (fig. 1) [22]. Occasionally patients complain of occipital headache, entire hemicrania or bilateral pain. In about a sixth of the patients the pain involves the neck, face and head simultaneously. In more than 70% the pain is severe. Occasionally it is like a thunderclap. It has also been reported to mimic migraine or cluster headache.

Ipsilateral Horner's syndrome is reported in up to 50% of patients (fig. 2). It is due to compression, stretching or hypoperfusion of the sympathetic fibres within the carotid wall. A painful Horner's syndrome of acute onset is almost pathognomonic of carotid dissection. Carotid dissection leads to cranial nerve palsies in about 10% of patients. The hypoglossal nerve is most commonly affected, followed by cranial nerves IX, X, XI and V. The involvement of various combinations of nerves has been described. The oculomotor and facial nerves may be involved in rare cases. One possible mechanism leading of cranial nerve palsy is compromise of the vasa nervorum. Direct compression of the cranial nerves by the mural haematoma is another possible explanation.

Dissection with stenosis may lead to pulsatile tinnitus due to the propagation of the murmur of the carotid stenosis but this is present in less than one fourth of patients

Retinal and cerebral ischaemia

Transient monocular blindness ipsilateral to the affected carotid artery, due to embolism or impaired blood flow to the retina, is a frequent warning symptom. It is very suggestive of dissection when associated with acute ipsilateral facial pain or headache. Persisting visual loss due to central retinal artery occlusion or anterior ischaemic optic neuropathy is rare [23].

The frequency of cerebral ischaemia varies from 50% to 90%, mainly depending on delay in diagnosis and patient selection. Local symptoms, amaurosis fugax and/or transient ischaemic attacks precede the stroke in the majority of patients. Most cerebral infarcts are located in the middle cerebral artery territory and occur within the first week of local symptom onset. However late infarcts, more than a month after symptom onset, have been reported. CAD with ischaemic events has a higher prevalence of >80% stenosis or occlusion or intracranial obstruction, and is less frequently associated with Horner's syndrome and cranial nerve palsy, compared with dissection without ischaemic events [24].

Diagnosis

Investigations should be performed as an emergency when CAD is suspected. Magnetic resonance imaging (MRI) in combination with magnetic resonance angiography (MRA) is a reliable, non-invasive method for the diagnosis of CAD. Axial T1-weighted fat-

suppressed techniques can show the intramural haematoma as a crescent-shaped hyperintensity that surrounds the narrowed lumen (fig. 3). MRI and MRA can also define the location and length of the dissection and the degree of stenosis. The typical location of CAD is different from the location of atheromatous plaques. It is generally located two centimetres downstream from the carotid bulb and often extends until the entrance of the internal carotid artery into the osseous part. However, MRI and MRA also have some limitations. The differentiation between intramural haematoma and intraluminal thrombus can be difficult, and hyper acute haematoma may remain unrecognised because of iso-intensity to the surrounding tissue [25].

Other investigations include digital subtraction angiography, multisection computed tomography (CT) and CT angiography and extracranial and transcranial Duplex ultrasound. Digital subtraction angiography is invasive and not always available on an emergency basis and has therefore been replaced by MRI/MRA as the diagnostic gold standard. Nevertheless, catheter angiography may be necessary in some rare cases with doubtful MRI findings. CT and CT angiography have been shown to be reliable in diagnosing CAD in small case series. However, experience with CT and CTA is limited, and therefore most centers use MRI/MRA as the diag-

Figure 3
Cervical axial MRI with fat-suppression technique showing a characteristic hyperintense semilunar intramural haematoma in the prepetrosal segment of the left internal carotid artery.

nostic method of choice. Duplex ultrasound is often used as a screening method in patients with clinical suspicion of CAD. However, carotid ultrasound shows normal findings in up to 31% of the patients, especially when only local clinical symptoms or signs are present [26]. Another major shortcoming of ultrasound is the lack of specificity, stressing the need for confirmation of the diagnosis by MRI/MRA. In the rare cases were an intimal flap or a double lumen can be visualised by duplex sonography confirmation by MRI is not mandatory. In summary, combined MRI/MRA is the most important diagnostic tool for CAD. However, it should be remembered that each of the above mentioned investigations may be normal, stressing the need for combining or repeating investigations in some patients.

Treatment

To date, there are no evidence-based guidelines for the treatment of CAD. There is a broad consensus that antithrombotic treatment should be given immediately after diagnosis of CAD. The main goal of antithrombotic treatment is to prevent retinal and cerebral embolism from the dissected artery. This concept is supported by recent studies suggesting that most strokes in patients with CAD are of embolic rather than of haemodynamic origin [27, 28].

However, there is much debate as to whether CAD patients should be treated with anticoagulants or antiplatelet agents [29]. For a controlled randomised trial comparing aspirin with anticoagulation a sample size of more than 2000 patients using clinical endpoint events has been suggested. Such a large trial is unlikely to be completed within a few years.

Our usual treatment of acute CAD is intravenous heparin followed by oral warfarin with a target international normalised ratio of 2.0 to 3.0 for 3 to 6 months. In patients with large cerebral infarcts or infarcts with haemorrhagic transformation we give aspirin for 10 to 21 days depending on the size of the infarct and then switch to anticoagulants for 3 to 6 months. We then either stop all antithrombotic treatment if the artery has completely normalised or we switch to antiplatelet therapy such as aspirin 100 mg daily for long-term prevention when there is an underlying arterial disease such as fibromuscular dysplasia or a persistent stenosis or occlusion of the dissected vessel.

Some centres prefer aspirin to anticoagulants in acute CAD, especially in patients with isolated local symptoms or signs and in patients with low-grade carotid stenosis.

For intracranial dissection we avoid anticoagulants and use aspirin because of the danger of subarachnoid haemorrhage, although there are no controlled trials. In the case of subarachnoid haemorrhage we do not give any anthrombotic treatment.

In patients with acute ischaemic stroke following

CAD and marked neurological deficits, thrombolysis has been shown to be feasible [30]. However, patients only exceptionally present within three hours of symptom onset. Endovascular treatment with balloon dilatation and stenting has been used successfully in rare selected patients with progressive or fluctuating symptoms of ischaemia despite anticoagulants. Surgical treatment has a high complication rate, including stroke, early and delayed graft re-occlusion, inability to access the injured segment, and lesions of cranial nerves and is therefore not recommended for treatment of CAD [31].

Prognosis

The long-term prognosis of CAD is favourable in the majority of patients. Clinical functional outcome mainly depends on the initial stroke severity. Including asymptomatic and monosymptomatic cases, the mortality rate is less than 5%. Nearly three fourths of the patients recover without significant disability [5].

Traumatic CAD seems to have a less favourable outcome than spontaneous CAD [32]. However, to date direct comparisons between outcome of traumatic and spontaneous CAD are lacking.

Several studies have reported a less than 1% annual rate of recurrent stroke or recurrent CAD [33, 34]. Symptomatic recurrent CAD is uncommon and mainly occurs in a different artery and rarely in the same artery. Risk factors for recurrent dissection are not clearly defined, but a family history of arterial dissection has been reported to be associated with a higher risk of recurrent CAD [35].

Asymptomatic early recurrent dissections seem to be more frequent than expected. A recent small study using repeated MRIs has shown mainly asymptomatic recurrent dissections in initially uninvolved cervical arteries in 9 of 36 patients [36].

On follow-up ultrasound or MRA the majority of CAD with stenosis, and some occlusive dissections, recanalise – mainly within the first 3 to 6 months. Persistent occlusion or stenosis is reported in 15 to 40% of patients [37].

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