Ultrasound in spontaneous cervical artery dissection

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Summary
Spontaneous cervical artery dissection is caused by a hematoma in the arterial wall. Recent research revealed that the most likely pathophysiological key mechanism is rupture of a vas vasorum resulting in a bleeding into the medio-adventitial borderzone [1]. The expansion of the hematoma into the arterial lumen can secondarily lead to a rupture of the tunica intima with a high risk of thrombus formation and embolic cerebral infarction [2]. Moreover the expansion of the hematoma causes an arterial stenosis or arterial occlusion with the risk of hemodynamic impairment. The risk of an ischemic stroke in the course of a dissection is thought to be about 70% for dissections of the internal carotid artery (ICA) [3] and about 80% for dissections of the vertebral artery (VA) [4]. The annual incidence of dissections of the ICA has been estimated to be 2.5—3/100,000 and for the VA 0.97—1.5/100,000 [5,6]. Although dissections as such are rare they are a frequent etiology of stroke in children and young adults. Approximately 25% of the strokes in patients younger than 50 are caused by dissections with a peak age between 40 and 45 years [7—16].

Introduction
Spontaneous cervical artery dissection is caused by a hematoma in the arterial wall. Recent research revealed that the most likely pathophysiological key mechanism is rupture of a vas vasorum resulting in a bleeding into the medio-adventitial borderzone [1]. The expansion of the hematoma into the arterial lumen can secondarily lead to a rupture of the tunica intima with a high risk of thrombus formation and embolic cerebral infarction [2]. Moreover the expansion of the hematoma causes an arterial stenosis or arterial occlusion with the risk of hemodynamic impairment. The risk of an ischemic stroke in the course of a dissection is thought to be about 70% for dissections of the internal carotid artery (ICA) [3] and about 80% for dissections of the vertebral artery (VA) [4]. The annual incidence of dissections of the ICA has been estimated to be 2.5—3/100,000 and for the VA 0.97—1.5/100,000 [5,6]. Although dissections as such are rare they are a frequent etiology of stroke in children and young adults. Approximately 25% of the strokes in patients younger than 50 are caused by dissections with a peak age between 40 and 45 years [7—16].

Due to the technical improvement of the ultrasound devices the investigation of the brain supplying arteries is nowadays an established and indispensable diagnostic tool in the detection and monitoring of spontaneous dissection.

Ultrasound in spontaneous dissection of the internal carotid artery

The ultrasound investigation should include the complete anterior circulation, i.e. both common carotid arteries (CCA), both external carotid arteries (ECA) and both ICA. Moreover, the flow velocities and flow properties of the intracranial portion of the ICA, the anterior (ACA), and
middle cerebral arteries (MCA) should be documented and the collateral flow pathways including the periorbital arteries if present. For the extracranial parts of the arteries, a high frequency linear transducer (≥7.5 MHz) should be used. The use of a sector probe for the distal portion of the ICA is strongly recommended, as the stenosis is frequently located much further distally to atherothrombotic disease [17,18]. For the intracranial arteries, a phased array transducer (≥2 MHz) is recommended.

The ultrasound investigation usually reveals absent or only mild atherosclerosis due to the fact that dissections occur in middle aged people [3,19–22]. A higher incidence of kinking or coiling of arteries has been reported in patients with cervical artery dissection [23]. However, other investigators could not confirm this arterial elongation as a regular finding in this patient group [24]. In patients with fibromuscular dysplasia, a known risk factor for cervical artery dissection [25], irregular wall thickening, multisegmental stenosis or an aberrant course of the ICA are frequently found [26,27].

The typical angiographic signs of an ICA dissection have first been described at first in conventional transfemoral angiography restricted to intraluminal pathologies [28]

- Smooth or slightly irregular tapered stenosis
- Various types of ICA occlusion
- Rattail-shaped tapered occlusion
- Flame-like occlusion
- False or double lumen
- Saccular or fusiform aneurysmal dilatation (pseudoaneurysm)
- Irregular dilatation
- Intimal flap

B-mode ultrasound investigation also visualizes the arterial wall and the surrounding tissue. The typical direct finding of a dissection of the ICA is the detection of a wall thickening of low echogenicity caused by the intramural hematoma with adjacent thrombotic material leading to a stenosis of this artery [17,22,29] (see Fig. 1).

In contrast to atherosclerotic stenosis which is predominantly located at the proximal part of the ICA, the stenosis due to dissection is found primarily in the distal part of the ICA [21,30]. Therefore it is often helpful to examine the distal part of the ICA with a sector probe especially in patients with a short neck, a prominent mandibular angle or a high bifurcation of the carotid artery. The detection rate of an intramural hematoma in the ICA by ultrasound is about 15–25% [17,22,29,31] (Fig. 2).

Another direct ultrasound sign of spontaneous cervical artery dissection is a “double lumen” which is found very rarely in the ICA. It is a result of a ruptured Tunica intima due to the space occupying intramural hematoma. The sonographic detection rate varies between 0 and 2% [17,31].

More diagnostic sensitivity is achieved when performing a duplex sonography with measurement of the blood flow velocity and with graduation of stenosis. Due to the fact that a stenosis caused by a dissection is located at the more distal part of the ICA this arterial segment has to be investigated with a sector probe more often. The sector probe has a lower spatial resolution with a lower chance to detect the intramural hematoma directly. In summary the detection of a stenosis in an arterial segment usually not affected by atherosclerosis is the most frequent finding. On the basis of the European Carotid Surgery Trial (ESCT) criteria which considered the percentage of local diameter reduction, a focal increase of the blood flow velocity of more than 120 cm/s is the cut-off for a 50% stenosis. Most of the published ultrasound studies have used the ESCT criteria and therefore it has to be kept in mind that the actual most widely accepted North American Symptomatic Carotid Endarterectomy Trial (NASCET) classification refers to the distal diameter reduction which leads to lower degrees of stenosis [3,18,32]. In one of the largest patient series on 181 patients and 200 dissections of the ICA, stenoses of the ICA have been found according to the ESCT criteria in 88% of the patients (stenosis ≤50% in 8%, stenosis 51–80% in 9%, stenosis >80% or occlusion in 71% of the cases) [17].
Due to the distal location of ICA dissection sometimes only indirect signs are detectable with ultrasound. These indirect signs comprise:

(a) increased pulsatility upstream or decreased pulsatility downstream to the suspected lesion. This is detectable in about 77% of cases
(b) >50% difference in the blood flow velocity compared to the corresponding arterial segments of the artery under study, detectable in about 73%
(c) detection of intracranial collateral flow, detectable in about 38% [31].

Taken the indirect and direct signs together, pathologic ultrasound findings suggestive for ICA dissection can be detected in 80—96% of all cases [18,31,33]. However, clinical aspects are also very important. In patients with local symptoms only (new onset of so far unknown head and or neck ache (painful) Horner’s syndrome, pulsatile tinnitus, palsies of the caudal cranial nerves (No IX—XII), or rarely palsies of the Nerves Nos. III, IV, VI), the ultrasound investigation is much less sensitive [3].

In summary the ultrasound investigation has a high sensitivity in detecting pathologic findings in patients with ICA dissection. However, it is not the sole investigation to verify the diagnosis of dissection especially in patients with local symptoms only.

**Ultrasound in spontaneous dissection of the vertebral artery**

The ultrasound investigation of the vertebral artery (VA) should include all segments, the origin and pre-vertebral part of the artery (V0/V1 segment), the part between the foramina of the transverse processes (V2 segment), the atlas loop (V3 segment) and the intracranial part (V4 segment). The V1 and V2 segment is normally investigated with a linear probe. The origin of the VA is sometimes not accessible with the linear probe especially in obese patients, and an investigation with a sector probe is superior. This is also the case when the V3 segment with its curved course is investigated. The V4 segment should be investigated via the transnuchal approach with a phased array transducer.

In analogy to the ICA dissection, the intramural hematoma of a VA dissection can cause an echolucent wall thickening and sometimes a double lumen. These signs can be found in 10—20% [31] (see Fig. 3).

A stenosis or occlusion can be more often diagnosed by detecting a focal increase of the blood flow velocity especially in the V3 segment where it is nearly impossible to visualize the arterial wall with the B mode ultrasound. Comparable to the findings in ICA dissection, a stenosis or occlusion due to dissection occurs in nearly 80% [31]. The corresponding indirect signs such as increased or decreased pulsatility or a blood flow velocity difference of >50%, are more difficult to interpret since the VA can be hypoplastic or is ending in the posterior inferior cerebellar artery [35].

A proximal arterial occlusion may be overlooked when the V4 segment is filled with an orthograde flow via cervical collaterals [36]. Comparable to ICA dissections, the predilection site for VA dissection is different from atherosclerotic lesions. The dissections occur primarily in the V2 and V3 segment [4] whereas the atherosclerotic disease is mostly found in the V0 or V4 Segments [37]. The overall sensitivity of the ultrasound investigation in detecting pathologies suggestive of a VA dissection varies from 70 to 92% [18,31,38].

In 8—13% the ultrasound investigation reveals normal findings despite MRI proven ICA or VA cervical artery dissection. The reason for this is usually a dissection in the distal part of the ICA especially at the base of the skull where the resolution of the B-mode is not high enough to detect the intramural hematoma directly. Another reason for failure found in ICA and VA dissection is a mild stenosis of <50% without hemodynamic flow changes [18,31].

**Recanalization of dissected arteries**

Hemodynamic relevant stenosis and arterial occlusion are frequently found in cervical artery dissection. The recanalization rate of ICA or VA occlusion can be easily monitored by ultrasound and varies between 42 and 72% and occurs within 6 weeks to 18 months [20,39—41]. The improvement rate of stenotic or occluded arteries is about 69% within the first 6 months after dissection. Afterwards, the improvement rate is much lower (19%). A complete recanalization without any stenosis after 6 months is achieved in 39% [40]. Beyond 9 months, further recanalization is only rarely seen (1%) [41].

**Recurrence of dissection**

So far, a recurrence of dissection between 2 days and 8.6 years has been reported and frequencies vary between 0 and 8% [3,6,10,11,42—44]. In a recent study with repetitive MRI-investigations in a group of 36 patients, a much higher recurrence rate could be found. A new dissection in a formerly unaffected artery was diagnosed in 19% between 1 and 4 weeks, and in another 6% of patients within 5—7 months...
Spontaneous cervical artery dissection

This remarkable finding has been reproduced in a much larger cohort of 76 patients with 105 dissections. The patients have been investigated with repetitive ultrasound daily during the hospitalization, then every month during the first 6 months and afterwards every 6 months with a mean follow-up of 58 months. A recurrent dissection in a formerly unaffected artery has been detected in 20 arteries (26.3%) during the stay in hospital. Afterwards a recurrence was found only in 2 (2.7%) previously affected arteries [41]. Therefore, the recurrence rate is much higher than previously thought and varies from 19 to 26% in the acute phase of the disease.

Summary

Due to the high sensitivity in detecting pathologic findings, ultrasound is an essential investigation method for both the ICA dissection and VA dissection because it can be quickly performed, it has a high availability and it is non-invasive. However, the diagnosis should be confirmed by MR-imaging because this is the method of choice to detect the intramural hematoma [45,46]. We recommend using both methods complementarily. Ultrasound is the most practical method for monitoring of hemodynamics in dissection and follow-up investigations to detect recurrent dissections which are more than twofold more frequent than previously thought.

Authors’ contributions

All authors have contributed substantially to the manuscript. They drafted and revised it together and gave final approval to its submission.

Conflict of interests

Dr. Dittrich and Dr. Ritter have no conflict of interest.

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