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CLINICAL AND RADIOLOGICAL FINDINGS IN CERVICOCRANIAL ARTERY DISSECTION

Z. Betül YALCINER*, Göksel BAKAC TUMAY**, Sema DEMIRCI BRANDMEIER***, Melek KANDEMIR*, Hayriye KUCUKOGLU****, Cengiz DAYAN*****, Nurhak DEMIR*****

*Bayindir Hospital Icerenkoy, Department of Neurology, Istanbul, TURKEY
**Kadikoy Florance Nightingale Hospital, Department of Neurology, Istanbul, TURKEY
***Medical Faculty of Istanbul Medipol University, Department of Neurology, Istanbul, TURKEY
****Bakirkoy Mazhar Osman State Training and Research Hospital for Psychiatric and Neurological
Diseases, 2nd Clinic of Neurology, Istanbul, TURKEY
****Bakirkoy Mazhar Osman State Training and Research Hospital for Psychiatric and Neurological
Diseases, 1st Clinic of Neurology, Istanbul, TURKEY

***** Medical Faculty of Dokuz Eylul University, Department of Neurology, Izmir, TURKEY

ABSTRACT

INTRODUCTION: Cervicocranial artery dissections account for 25% of ischaemic strokes in young adults. Numerous risk factors that can damage the structure of the vessel walls have been identified. Dissections can present with various forms of stroke, and are diagnosed using magnetic resonance angiography or digital subtraction angiography. In our study, we aimed to evaluate the clinical and radiological characteristics of cervicocranial artery dissections.

METHODS: We retrospectively analysed 45 patients, twenty-nine (64%) males and 16 (36%) females, pre-diagnosed with dissection in the Angiography Unit of the Bakirkoy Mazhar Osman State Training and Research Hospital for Psychiatric and Neurological Diseases. The mean age of females and males was 35 and 39 years, respectively.

RESULTS: Forty-one (91%) patients presented with transient ischaemic attack and/or ischaemic stroke. Isolated 12th nerve paralysis was found in one patient, and headache was the only symptom in another. Digital subtraction angiography was performed in all the patients, and bilateral dissection was detected in six patients. From the angiography results, three different types of vessel pathologies were detected: 1- long-segment irregularities (65%), 2- tapering occlusion (35%) and 3- pseudoaneurysm (16%). Radiological follow-up was done for 19 (42%) patients; eight of them completely recovered showing normal imaging findings.

DISCUSSION and CONCLUSION: Dissection can present with a broad clinical spectrum, and it should be suspected in cases with unusual neck movement and mild-to-severe trauma. Digital subtraction angiography is still the best modality for the diagnosis of dissection

Keywords: Cervicocranial artery dissection, aetiology, imaging findings.

SERVİKOKRANİYAL ARTER DİSEKSİYONLARINDA KLİNİK VE RADYOLOJİK BULGULAR

ÖZET

GİRİŞ ve AMAÇ: Servikokraniyal arter disseksiyonları genç erişkinlerdeki iskemik inmelerin %25'inden sorumludur. Damar duvarı yapısına zarar veren çok sayıda risk faktörü tespit edilmiştir. Disseksiyonlar çeşitli inme formlarında görülebilirler ve manyetik rezonans anjiografi veya dijital substraksiyon anjiografi tetkikleri ile tanı konulabilir. Çalışmamızda servikokraniyal arter disseksiyonlarının klinik ve radyolojik özelliklerinin değerlendirilmesi amaçlanmıştır. YÖNTEM ve GEREÇLER: Bakirkoy Mazhar Osman Ruh Sagligi ve Hastaliklari Egitim ve Arastirma Hastanesi anjiografi ünitesinde disseksiyon ön tanısı almış 45 hasta, 29 (%64) erkek ve 16 (%36) kadın, retrospektif olarak incelenmiştir. Ortalama yaş kadın ve erkeklerde sırasıyla 35 ve 39 idi.

BULGULAR: Hastaların 41'i (%91) geçici iskemik atak ve/veya iskemik inme ile prezente olmuştur. İzole 12. kraniyal sinir felci bir hastada gözlenmiş, bir başkasında ise baş ağrısı tek semptom olarak görülmüştür. Tüm hastalara dijital substraksiyon anjiografi yapılmış ve 6 hastada bilateral disseksiyon saptanmıştır. Anjiografi sonuçlarında 3 farklı damar

Corresponding author: Melek Kandemir, MD. Bayindir Hospital Icerenkoy, Department of Neurology, Istanbul, Turkey.

Telephone: +90216 575 26 66-3373 E-mail: kandemirmelek@yahoo.com

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patolojisi saptandı: 1- uzun segment düzensizlikleri (%65) 2- kama şeklinde tıkanma (%35) ve 3- yalancı anevrizma (%16). Hastaların 19'u (%42) radyolojik olarak takip edilebilmiş, bunlardan sekizinde radyolojik bulgular tamamen normale dönmüştür.

TARTIŞMA ve SONUÇ: Disseksiyon geniş bir klinik spektrumla prezente olabilir ve alışılmışın dışında boyun hareketi ve ılımlıdan şiddetliye kadar çeşitli düzeyde travma öyküsü olanlarda akla gelmelidir. Disseksiyon tanısında dijital substraksiyon anjiografisi hala en iyi yöntemdir.

Anahtar Sözcükler: Servikokraniyal arter diseksiyonları, etiyoloji, görüntüleme bulguları.

INTRODUCTION

Cervicocranial artery dissections (CCADs) account for 2% of all ischaemic strokes and 25% of the ischaemic strokes in young adults under the age of 45 years (1, 2, 4, 6, 8–11, 15, 20, 22, 24–28, 30-37). Although numerous risk factors that can damage the structure of the vessel walls have been identified, such as hypertension, smoking, hyperhomocysteinaemia, migraine. infection. hyperlipidaemia, alpha-1 antitrypsin deficiency, connective tissue disease (Ehlers-Danlos and Marfan syndromes, fibromuscular dysplasia, i.e., FMD) and trivial/minor trauma chiropractic applications and hyperextension/flexion of neck), the reasons for CCAD and its pathogenesis still remain unclear (1, 7, 10, 15, 17, 26, 28, 29, 32, 34–36, 38).

Artery dissection starts with a rupture in the intimal layer in the weakest part of the vessel. Due to the high pressure in the vessel, the rupture extends into the tunica intima endothelial cells and media layer. The extension of the rupture along the vessel causes the formation of a cleavage and a false lumen, associated to it, resulting in stenosis and occlusion of the lumen. Thrombus formation in the dissection area is common; on the other hand, a sub-adventitial dissection can transform into a pseudoaneurysm (6, 10, 20, 22, 27).

CCADs can present with completed stroke, transient ischaemic attack (TIA), subarachnoid haemorrhage (SAH), or isolated cranial nerve palsy (1, 9, 30). Two possible mechanisms have been put forward for the development of stroke. The first is related to stenosis and occlusion of the real lumen, while the second is related to microemboli detached from the thrombus. If the dissection extends intracranially, the bleeding of the pseudoaneurysm may cause SAH. Further, the development of isolated cranial nerve palsies is associated with aneurysm compression or ischaemia (6, 10, 20, 22, 27).

In this study, we aimed to evaluate the clinical and radiological characteristics of CCAD, which can appear in various forms.

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MATERIAL AND METHODS

We retrospectively analysed the clinical and radiological characteristics of 45 patients prediagnosed with dissection between 1998 and 2004 in the Angiography Unit of Bakirkoy Mazhar Osman State Training and Research Hospital for Psychiatric and Neurological Diseases. Age, gender and risk factors of the patients were recorded. However, sufficient information was not obtained regarding the complaints and risk factors of eight patients. The images were taken in the cranial and cervical anterio-posterior, oblique and lateral planes using a digital subtraction angiography (DSA) Siemens Polystar device (Berlin, Germany). The findings were classified based on the anatomic region of the dissection. Nineteen patients were followed up, and DSA was performed in 12 of these patients. Further, control imaging with magnetic resonance angiography (MRA) was performed in 7 of the followed-up patients.

Our study is a retrospective analysis of the angiography results that were obtained for diagnostic purposes in the Angiography Unit. Therefore, we did not require study approval from the Ethical Board, although general consent was obtained from each hospitalized patient.

RESULTS

Of the 45 patients included in the study, 16 (36%) were female and 29 (64%) were male, with mean ages of 35.8 years (range, 19–69 years) and 39.6 years (range, 18–56 years), respectively. Table shows the risk factors, clinical presentations and imaging findings with respect to the dissection types. Eleven patients had a history of trivial/minör trauma (hyperextension/flexion/rotation of the neck, minor traffic accidents, or massage, etc.) 25 days prior to their complaint.

In 24 (65%) of the 37 patients, there were complaints of headache and/or neck pain, correlating with the clinical findings, and these complaints were more common in the patients

with vertebral artery dissections (VAD) compared to those with internal carotid artery dissections (ICAD).

Forty-one (91%) patients presented with TIA and/or cerebrovascular accident in the form of ischaemic stroke. Six patients presented with TIA, six with ischaemic stroke development followed by TIA and 29 with completed ischaemic stroke. Furthermore, SAH was detected in one of the ischaemic stroke patients, and both SAH and VAD were detected in two patients overall.

Of the patients with DSA, 20 (44%) had ICAD and 25 (56%) had VAD. Bilateral dissection (Figure I) was present in six patients. In total, 55 vessel pathologies were detected, with more than one vessel affected in the same patient or more than one pathology detected in the same vessel in some patients. Three different types of vessel pathologies were detected: 1- long-segment irregularities (Figure II) 2- tapering occlusion (Figure II and III) 3- pseudoaneurysm (Figure IV).

In 36 (65%) vessels, long-segment stenosis and irregularities were detected; further, occlusion and pseudoaneurysm were detected in 19 (35%) and nine (16%) vessels, respectively. The findings are summarized in Table.

Radiological follow-up was performed for 19 (42%) patients. Eight patients showed normal radiological findings, (Figure V) with average follow-up period of 3.5 months (1–6 months), no recurrence was observed in any of the 19 patients.

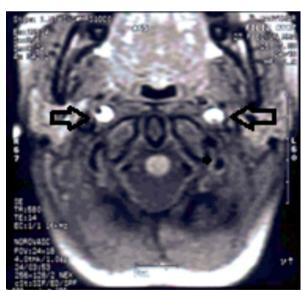


Figure I. Axial fat-saturated T1-weighted magnetic resonance imaging shows bilateral internal carotid artery dissections.



Figure II. Digital subtraction angiography shows long-segment irregularities in the carotid arteries (fibromuscular dysplasia).

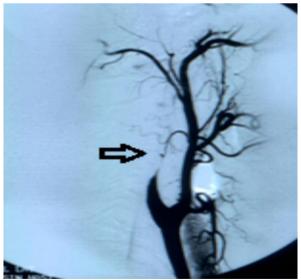


Figure III. Digital subtraction angiography shows tapering occlusion of the left carotid artery.

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Figure IV. Digital subtraction angiography shows posterior inferior cerebellar artery dissection and pseudoaneurysm.

DISCUSSION

CCADs are one of the rare causes of stroke. Nevertheless, they are responsible for 10%-25% of the strokes seen in adults younger than 45 years of age (1, 2, 4, 6, 8-11, 15, 20, 22, 24-28, 30-37). In accordance, the average age of the patients in our study was under 40 years. Further, previous studies have shown that CCAD is more common in males than in females. In our study, the number of males was about twice that of females. Although there is no definite distinction among dissections in terms of gender, similar to the findings of previous studies, the females in our study were slightly younger than the males (3, 24, 25, 30). In some studies, it has been claimed that multiple vessel dissection is more common in females (3). However, in our study, they were more common in males.

Various risk factors have been identified for the aetiology of CCAD, including diseases related to connective tissue, environmental factors (infection), trivial/minor traumas and atherosclerotic risk factors (1, 7, 10, 15, 17, 26, 28, 29, 32, 34–36, 38). The risk factors identified in our study were hypertension, hyperlipidaemia, migraine, smoking, alcoholism, FMD and trivial/minor trauma.

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Table. Clinical and angiographic findings with respect to dissection types.

-		ICAD*	VAD† Total	
		(n)	(n)	(n)
Gender (M/F)		11/9	18/7	45
Risk factors	Smoking	1	9 ′	10
	Alcohol	1	3	4
	Migraine	3	4	7
	Hypertension	0	5	5
	Hyperlipidaemia	2	4	6
	Headache/Neck Pain	9	15	24
	Minor Trauma	3	8	11
Clinical	TIA‡	5	1	6
presentation	Stroke	14	21	35
	SAH§	0	2	2
	Isolated CN∥ XII Palsy	1	0	1
	Headache (only)	0	1	1
DSA [¶] findings	Extracranial Dissection	24	22	46
	Intracranial Dissection	19	13	9
	Bilateral Dissection	4	2	6
	Long-Segment Irregularity	15	12	27
	Tapering Occlusion	9	10	19
	Pseudoaneurysm	5	2	7

*ICAD: Internal carotid artery dissection; †VAD: Vertebral artery dissection; †TIA: Transient ischemic attack; §SAH: Subarachnoid hemorrhage; ||CN: Cranial nerve; †DSA: Digital subtraction angiography

Histories of massage, hyperextension/ flexion/rotation of the neck and minor traffic accidents were considered as trauma, up to 25days prior to the dissection. Twenty-four percent of the patients had a history of minor trauma. The frequency of minor trauma as a risk factor for CCAD has not been confirmed in the literature. However, a previous study showed a rate of minor trauma history of at least 40% in the 30-day period before CCAD (13). Severe trauma may directly result in vessel occlusion; on the other hand, it has been claimed that minor trauma can be a mechanical trigger, resulting in dissection only in the presence of an impairment that causes the vessel blood to become fragile on the ground (13, 17, 21, 29, 34). Therefore, it has been suggested in some studies that mechanical triggers be used as risk factor rather than minor trauma (13, 17, 29). However, in order to clarify the relationship between minor trauma and dissection, systematic studies involving large numbers of cases need to be conducted.

In our study, the most common complaints of the patients were headache and/or neck pain (65%), and these complaints were more common in patients with VAD, similar to results of several other studies (1, 4, 14, 19). Headache was the only complaint of one of our patients with VAD.



Figure V. One month later, spontaneous occlusion of posterior inferior cerebellar artery was observed in the same patient as that in Figure IV.

Dissection with isolated headache is reportedly rare; however, it is most commonly seen in patients with VAD (1, 12, 18). It has also been reported that pain could be unilateral or bilateral in dissections, even it is unilateral (1, 19).

Dissection can result in two situations depending on the localisation of the haematoma on the wall of the artery: 1) haematomas in the subintimal layer narrow the artery lumen and increase the risk of thrombosis, resulting in temporary or completed ischaemic symptoms. 2) intramural haematomas settle subadventitially more frequently, causing the vessel wall to expand eccentrically. In these cases, as the haematoma gradually affects the artery lumen, cranial nerve involvement is observed rather than cerebral ischaemic findings (6, 10, 16, 20, 22, 27). Cranial nerve involvement is associated with ischaemia due to compression or interruption in the nourishing vessels of the nerve, and is seen in 10% of the extracranial carotid dissections (9, 21). In our study, there was one patient with isolated 12th nerve involvement due to extracranial ICAD. If a sub-adventitially expanding dissection extends to the intracranial segment, it may also cause SAH. SAH is more common in intracranial dissections because there is no external elastic membrane in the adventitia layer of the intracranial arteries (30, 31). VAD presenting with SAH has been reported more often than CAD (10, 30). In our study, SAH was detected in three patients, one with stroke and the other two in association with VAD.

Although it is not useful for determining artery wall thickness and configuration in the diagnosis of the dissection, DSA is widely accepted as a gold standard imaging technique (4, 10, 22, 27, 28, 31, 33, 37). As vertebral arteries (VAs) are smaller, deeper and with frequent anatomical variation and surrounding tissues, DSA is more useful in the diagnosis of VAD (1, 22, 31). Angiographic findings vary depending on the degree of luminal stenosis and the depth of the localized false lumen in the vessel wall (31). The 'string sign', which refers to the tapering of the eccentric stenosis that originates from the distal end of the carotid bulb, is the hallmark of ICADs. Focal stenoses in the distal part of dilatations are known as 'string and pearl signs'. Pathognomonic findings, such as double lumen or intimal flap are rarely observed (10, 21, 33), and the most common findings in DSA, in descending order of frequency, are: irregular stenosis, occlusion and pseudoaneurysm. Pseudoaneurysm occurs if the dissection is sub-adventitial. the pseudoaneurysm is thrombosed, it may not be visible initially (27, 37). Our findings, in the descending order of frequency, were irregular stenosis, occlusion and pseudoaneurysm, Fortyfour percent of our patients had ICAD and 56% had VAD. Many previous studies have reported that ICADs are more common than VADs (20, 33). In a study by Maruvama et al., VADs were more common, similar to the findings of our study (23).

In recent years, non-invasive imaging modalities have gradually replaced DSA. However, DSA is still the best technique in cases where filiform stenosis with very slow blood-flow, FMD and complications of intracranial dissection need to be imaged, as well as in cases where noninvasive techniques fail to confirm the results. Doppler duplex ultrasound has low sensitivity in detecting VAD findings, which are especially difficult to distinguish from low-grade stenosis and deeper structures in the neck. MRA or computed tomography angiography, together with magnetic resonance imaging (MRI), play important roles in this differential diagnosis, and can be used to view haemorrhages associated with dissection and the ischaemic area. In MRI, the increased signal showing the narrowness of the eccentric lumen and the intramural haematoma (hyper intense image in T1 sequences) pathognomonic finding for ICAD (1, 31, 35) (Figure I). Due to the tissues surrounding VAs, the

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detection of intramural haematomas in VAs depends on the affected artery segment. Therefore, MRI is more sensitive for diagnosing ICAD (31). In our study, 31 patients were subjected to MRI, and the findings were normal in seven of these patients. None of the patients were subjected to MRA for diagnostic purposes. Noninvasive methods are normally preferred in the follow-up for dissection (20, 33). In our study, we used DSA as the primary method for diagnostic imaging.

Dissection is a dynamic process, and stenosis can quickly develop into occlusion, or the opposite may happen, resulting in occlusion recanalization (31, 37). Although documentation of the follow-up of the dissection cases in previous studies was insufficient, in the cases that were followed up it has been reported that the majority of the occlusions were recanalised, while 46% of the pesudoaneurysms were permanent (27, 31). The recurrence rate is very low in dissections and is reported to be 2% for the first month and 1% after 1 year. Connective tissue disease, FMD and having a family history of dissection increase the likelihood of recurrence (5, 10, 24, 31, 33). In our study, only 19 patients were followed up, and recurrence was not observed in any of them. There was complete recovery of vessel pathology, as observed by DSA or MRA, in eight patients.

Dissection is an important and curable cause of stroke in young adults. It can present with a broad clinical spectrum, from completed stroke to SAH, TIA, isolated headache, or cranial nerve involvement. In particular, patients with limited findings, such as isolated cranial involvement and/or headache/neck pain must be carefully examined, along with other risk factors. Whether there has been a recent history of extraordinary neck movement or mild-to-severe trauma must also be ascertained. Additionally, dissection is a dynamic process and, consequently the findings may vary depending on the process and the imaging technique chosen. Therefore, to achieve definitive diagnosis, the most appropriate imaging modality must be chosen depending on the patient's overall clinical findings. We believe that despite being an invasive technique, DSA is still the best modality for the diagnosis of dissection.

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