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ORIGINAL ARTICLE

Relation of the incidence of congenital variations and anomalies with intracranial aneurysms in intracranial arteries

İntrakranyal arterlerde konjenital varyasyon ve anomalilerin sıklığı ve intrakranyal anevrizmalar ile birlikteliği

Yeliz Aktürk[1], Mehmet Murat Fırat[2], Mehmet Emrah Güven[3], Murat Beyhan[4]

[1] Dışkapı Yıldırım Beyazıt Training and Research Hospital, Department of Radiology Ankara, Turkey
[2] Ankara Guven Hospital Department of Radiology, Ankara, Turkey
[3] Izmir Egepol Hospital Department of Radiology, İzmir, Turkey
[4] Tokat State Hospital Department of Radiology, Tokat, Turkey
Corresponding author: Yeliz Aktürk, Dışkapı Yıldırım Beyazıt Training and Research Hospital, Department of Radiology Ankara, Turkey
Email: yelizakturk@yahoo.com

Özet

Amaç: İntrakranyal vasküler yapıların gelişiminde pek çok embriyolojik basamak bulunur ve konjenital varyasyonları ve anomalileri sık görülür. Serebrovasküler görüntüleme çalışmalarının doğru yorumlanması için bu anomali ve varyasyonların görülme sıklıklarının, lokalizasyonlarının ve eşlik edebilecek patolojilerin ayırdedilebilmesi gereklidir. Çalışmamızda; 2004-2009 yılları arasında yapılmış olan serebral anjiogramları retrospektif olarak değerlendirerek konjenital varyasyon ve anomalilerin sıklığını ve intrakranyal anevrizmalar ile birlikteliğini araştırmayı hedefledik.

Yöntemler: Ocak 2007 ile Nisan 2009 tarihleri arasında Gaziosmanpaşa Üniversitesi Tıp Fakültesi Radyodiagnostik Anabilim Dalı'nda, çeşitli nedenlerle serebral anjiografisi yapılmış olguların arşiv görüntüleri iki radyolog tarafından, birlikte retrospektif olarak incelenmiştir. Toplam 254 olguda intrakraniyal arter varyasyon-anomalileri ve eşlik eden intrakranyal arter anevrizmaları saptanmıştır.

Bulgular: İntrakranyal arterlerde en sık görülen varyasyonlar anterior serebral arter A1 segmentinin hipoplazisi-agenezisi idi. Bunu posterior komünikan arterde infundibülar dilatasyon ve fetal orijinli posterior serebral arter izledi. A1 hipoplazisi-agenezisi izlenen olgularda anevrizma görülme oranı %22,5, orta serebral arterde trifürkasyon varyasyonu olan olgularda anevrizma görülme oranı %18,2 idi.

Sonuç: Akım hemodinamiği, intrakranyal sakküler anevrizmaların oluşumunda önemli etkilere sahiptir. İntrakranyal vasküler yapıların konjenital anomalilerinde hemodinamik strese bağlı anevrizma gelişim insidansı artmaktadır. Anevrizmaların patogenezinde yer alan hemodinamik faktörler hakkında bilgi sahibi olmak için varyasyonlar ve anevrizmaların yeri arasındaki ilişkiyi incelemek gerekir. Olgu sayımız sınırlı olmakla birlikte bu oranlar, varyasyonu olmayan hastalar ile karşılaştırıldığında varyasyonlu olgularda anevrizma görülme sıklığının arttığını düşündürmektedir.

Anahtar kelimeler: konjenital varyasyon, intrakranial anevrizma, anjiografi

Abstract:

Objectives: There exists many embryological steps during the development of intracranial vascular structures. Therefore, the congenital variations and anomalies of these structures are often encountered. For a proper evaluation of cerebrovascular imaging studies it is essential to know the frequencies of these anomalies and variations, their locations and the possible accompanying patologies should be distinguished. We aimed to analyse the frequency of congenital variations and anomalies and their association with intracranial aneurysms by evaluating the cerebral angiographic images.

Methods: The cerebral angiograms taken for various reasons in the Radiology Department, Faculty of Medicine, Gaziosmanpasa University, between January 2007 and April 2009, were evaluated in a retrospective fashion. We investigated the association of intracranial aneurysms and congenital variations in addition to the influence of variations on aneurysm formation. A total of 254 cases with intracranial artery variation-anomaly and coexisting intracranial artery aneurysm were included in the study.

Results: The most common variation in intracranial arteries was the agenesis or hypoplasia of the A1 segment of anterior cerebral artery. This was followed infundibular dilatation of posterior communicating artery and fetal type posterior cerebral artery. The incidence of aneurysm was 22.5% in patients who had A1 hypoplasia-agenesia and 18.2% in patients who had trifurcation of middle cerebral artery.

Conclusion: Flow haemodynamics take an important part in the formation of intracranial saccular aneurysms. The incidence of aneurysm formation increases in congenital anomalies of intracranial vascular structures. It is necessary to explore the relationship between variations and aneurysms in order to understand the haemodynamic factors which take place in the pathogenesis of aneurysms. These ratios lead us to think that the incidence of aneurysms increases in cases which have variations when compared to those which have not.

Key words: congenital variation, intracranial aneurysm, angiography

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INTRODUCTION

In order to accurately comment on cerebrovascular images, vascular anatomy of the central nervous system should be known in detail and anatomic variations mimicking pathological conditions should be differentiated [1]. Since many embriyological steps take part in the development of Willis polygon, variations and anomalies are commonly encountered in each branch [2]. In relation to these variations, hemodynamic stresses and media layer defects at the vascular junctions generate a predisposition to aneurysm formation. People with Willis polygon asymmetry or anomaly develop intracranial aneurysms more frequently [3]. Berry or saccular aneurysms are associated with vascular anomalies and increased flow rates [4,5]. Flow hemodynamics affect the formation, growth, and shaping of the intracranial aneurysms. Congenital anomalies of the intracranial vascular structures increase the incidence of hemodynamic stress-related aneurysm formation [6,7,8,9]. In order to gain further insight about the hemodynamic factors involved in the pathogenesis of aneurysms, the relationship between arterial variations and location of aneurysms should be investigated [10].

The aim of this study is to investigate relation of the incidence of congenital variation and anomalies with intracranial aneurysms by retrospectively evaluating cerebral angiograms taken between 2007-2009.

METHODS

The cerebral angiograms taken for various reasons in the Radiology Department, Faculty of Medicine, Gaziosmanpasa University, between January 2007 and April 2009, were evaluated in a retrospective fashion.

We investigated the association of intracranial aneurysms and congenital variations in addition to the influence of variations on aneurysm formation.

The entire angiographic images were obtained via DSA GE Innova 3100 (Milwaukee, USA). The archival images were assessed by the GE Adventix 4.3 workstation (Milwaukee, USA). Statistical analyses were conducted with the chi-square test.

Fenestration is described as a vessel with normal origin and position, having double lumens at some parts; duplication is supplying a certain area with two independent arteries originating from the same location; agenesis is the absence of a certain artery; posterior communicating artery (PComA) infundibular dilatation is a triangular-shaped dilatation less than 3 mm wide; dilatation of posterior cerebral artery (PCA) is the presence of a right or left P1 segment having a diameter >3.5 mm; persistant trigeminal artery, originating from the proximal segment of the internal carotid artery (ICA) located anteriorly, penetrating the cella turcica close to the clivus or a persistant embryological artery following a course lateral to the cella turcica and joining with the upper portion of the basilar artery (BA); hypoplasia of the anterior cerebral artery (ACA) -A1 segment is the presence of a right or left A1 segment having a diameter ≤1.5 mm; ACA-A1 segment agenesis is the presence of an A1 segment failing to fill; azygos ACA is the presence of A1 segments joining in the midline and proceeding as one artery while this single artery supplies both ACAs; bihemispheric ACA, both ACAs are present, however, one of them is dominant and supplies the contralateral artery only partially; middle cerebral artery (MCA) trifurcation, MCA-M2 segment ends by splitting into 3 main truncs; fetal origin of the PCA, PComA appears larger than the P1 segment and supplies the occipital lobe.

RESULTS

A total of 254 cases with intracranial artery variation-anomaly and coexisting intracranial artery aneurysm were included in the study. While 150 patients (59.1%) were female, 104 (40.9%) were male. The age range was 17-84

years (mean, 42.5 years) for women and 29-79 years (mean, 45 years) for men.

The retrospective assessment of the angiographic images revealed PComA infundibular dilatation, fetal origin of the PCA, and ACA A1 segment hypoplasia as the most common anomalies in the intracranial arteries (Table 1). No statistically significant difference was observed between men and women with regard to the incidence of arterial variations (p>0.05).

Table1. Intracranial congenital variations and anomalies

Infundibular dilatation of the PComA	48	18.9
Fetal origin of the PCA	43	16.9
Hypoplasia of the ACA A1 segment	43	16.9
Double SCA	29	11.4
Agenesis of the ACA A1 segment	28	11
Trifurcation of the MCA	22	8.7
Azygos ACA	12	4.7
Bihemispheric ACA	11	4.3
Infundibulary dilatation of the PCA	8	3.1
Fenestration of the BA	6	2.4
One trunc origin of the SCA-PCA	5	2
VA ending with the PICA	4	1.6
Persistant trigeminal artery	2	0.8

PComA: posterior communicating artery, ACA: anterior cerebral artery, A1: A1 segment of the anterior cerebral artery, SCA: superior cerebellar artery, MCA: middle cerebral artery, PCA: posterior cerebral artery, BA: basilar artery, VA: vertebral artery, PICA: posterior inferior cerebellar artery.

We evaluated A1 segment hypoplasia, agenesis, azygos ACA, as well as ACA and anterior communicating artery (AComA) aneurysms accompanying bihemispheric variations (Table 2).

Table 2. Anterior cerebral artery-A1 segment hypoplasia, agenesis, azygos anterior cerebral artery variations and aneurysms

	Number of the cases	Location of the aneurysms			
		AComA	ACA A1 segment	Pericallosal artery	Azygos ACA
Hypoplasia of the A1 segment	43	9	1		
Agenesis of the A1 segment	28	4		1	1
Azygos ACA	12				2

AComA: anterior communicating artery, ACA: anterior cerebral artery, A1: A1 segment of the anterior cerebral artery .

Furthermore, we evaluated the distribution of intracranial aneurysms coexisting with all types of variations (Table 3).

Table 3. Locations of intracranial aneurysms with congenital variations and anomalies

Variations and anomalies	Number of the cases	Location of the aneurysms						
		AComA	ACA	MCA	ICA	PCA	PComA	BA
Hypoplasia of the A1	43	9	1	6	5			
Agenesis of the A1	28	4	2	1	2			
Azygos ACA	12		2	5	2			
Bihemispheric ACA	11			1				
Unilateral fetal origin of the PCA	33	4		6	3			1
Bilateral fetal origin of the PCA	10	1		1	1			
Unilateral infundibular dilatation of the PComA	34	10		10	1		3	1
Bilateral infundibular dilatation of the PComA	14				2			1
Trifurcation of the MCA	22			5				
Infundibular dilatation of the P1	8		2		1			2
One trunc of the SCA-PCA	5			3				
Fenestration of the BA	6							1
VA ending with the PICA	3							1
Persistan trigeminal artery	2							

ACA: anterior cerebral artery, A1: A1 segment of the anterior cerebral artery, PCA: posterior cerebral artery, P1: P1 segment of the posterior cerebral artery, PComA: posterior communicating artery, MCA: middle cerebral artery, SCA: superior cerebellar artery, BA:basilar artery, VA: vertebral artery, PICA: posterior inferior cerebellar artery.

In cases with ACA A1 segment hypoplasia and agenesis, the incidence of coexisting aneurysms in ACA and AComA was 22.5%.

Five cases displayed intracranial aneurysm accompanying MCA trifurcation. The aneurysms were ipsilateral to the trifurcation in 4 cases and in the contralateral MCA at the bifurcation level in one case. Among cases with MCA trifurcation, the incidence of ipsilateral MCA aneurysm was 18.2%.

Seventeen of the fetal origin of the PCA cases (39.5%) had coexisting aneurysms. One case had fetal origin of the PCA accompanied by basilar artery fenestration, and there was a

kissing aneurysm over the fenestration bed of the basilar artery.

Two cases (4.2%) of PComA infundibular dilatation had aneurysmatic dilatation combined with infundibular dilatation. The aneurysmatic dilatation was on the right in both cases.

DISCUSSION

Since there are many embryological steps in the development of intracranial arteries, the variations and anomalies involving the branches of Willis polygon are particularly common [2]. Willis polygon including all components and lacking a hypoplastic component account only for 20-25% of all cases. About 50% of the variations are observed in the posterior circulation [11].

In this study, the most common variation in the intracranial arteries was infundibular dilatation in the PComA (18.9%), followed by fetal origin of the PCA (16.9%) and ACA A1 hypoplasia (16.9%). However, when A1 segment hypoplasia was evaluated in combination with A1 segment agenesis, it was the most common congenital variation, being present in 27.9% of the cases.

In the literature, the most common variation of ACA-AComA complex is the hypoplasia-agenesis of the A1 segment [12]. The incidence of hypoplastic-aplastic A1 segment is 5-18% [13,14]. In the present study, the indicence of hypoplasia-agenesis in the ACA A1 segment was higher than the rate reported in the literature [13,15].

Because AComA has a collateral function between the bilateral ACAs, the blood flow is under the influence of pressure difference of the ICAs. The presence of different pressures in the A1 segments of the ACA affects the blood flow in the AComA. Therefore, the asymmetry in the ACA proximal segments increases the AComA aneurysm incidence [16]. The hemodynamic effect of the pulsatile flow in the large A1 segment has an impact on the AComA anterior wall and this is a very common aneurysm site [17]. Yasargil et al. reported that 80% of AComA aneurysms are accompanied by A1 segment hypoplasia or agenesis [18]. Wilson et al. found ACA A1 segment hypoplasia in 85% of the AComA aneurysms [19]. Karazincir et al. conducted a retrospective study and observed the coexistence of A1 segment hypoplasia-agenesis in 50% of AComA aneurysm cases [6]. In our study, 22.5% of the A1 hypoplasia-agenesis cases had an aneurysm in the ACA and AComA. In this regard, our results were consistent with the literature, indicating an association between A1 hypoplasia-agenesis and the incidence of ACA and AComA aneurysms.

In the presence of azygos ACA, subsequent to the hemodynamic changes, the incidence of saccular aneurysm formation in the distal ACA, particularly at the pericallosal and callosomarginal artery bifurcation, is high [6]. Ohno et al. performed a large study and found the incidence of coexisting azygos ACA and distal ACA aneurysms as 8-9% [20]. Huber et al. found aneurysm in 7 (41.1%) of 17 cases with azygos and bihemispheric ACA variations [21]. In the present study, 2 (16.7%) of the 12 azygos ACA cases demonstrated an aneurysm in the azygos artery. Our sample size was not adequate to perform a statistical analysis, however, this rate suggests that azygos ACA variation is a risk factor for aneurysm formation in the ACA.

In our study, 22 cases (8.7%) showed trifurcation in the MCA. Four cases (18.2%) manifested an ipsilateral intracranial aneurysm at the level of trifurcation. Although our sample size was not adequate to reach a conclusion, the results indicate that trifurcation variation increases the risk of MCA aneurysm.

According to some authors, infundibulum is a potentially weak spot and can be regarded as a preaneurysmal lesion. A defect in the elastic layer may grow or become an aneurysm under the influence of hemodynamic effects such as carotid artery occlusion or hypertension [22,23]. In the literature, a long follow up is

recommended particularly for patients with contralateral ICA-PComA aneurysm, young patients, hypertensive cases, as well as cases with a well developed PComA, and those with small irregularities in the wall [24]. In our study, 2 cases manifested aneurysmatic dilatation in the presence of infundibular dilatation. Those cases did not manifest hemorrhage and are still followed up by our clinic.

To our knowledge, there is no study in the literature reporting that fetal origin of the PCA variation increases the incidence of aneurysm. In our cases, with the exception of 1 case involving the coexistence of fetal origin of the PCA and basilar artery fenestration in addition to kissing aneurysm in the basilar artery, no posterior system aneurysm accompanying this variation was determined.

The wall of a fenestrated artery exhibits normal structural properties. The medial wall includes focal defects at both ends of the fenestration and causes a predisposition to aneurysm formation. While the incidence of basilar fenestration and aneurysm coexistence is 7%, the incidence of fenestration in cases with an aneurysm at the vertebrobasilar junction is 35% [25]. In our study, we determined one case with kissing aneurysm formation associated with fenestration. Another case had dolichoectasia in the basilar artery. We were unable to evaluate the relationship between BA fenestration and the incidence of arterial aneurysm due to small sample size.

In the present study, two cases (0.8%) showed persistant trigeminal artery variation. The incidence of this variation in the literature is noted as 0.06-0.6%. At clinical settings, intracranial aneurysms and persistant trigeminal artery are associated in 14% of cases. However, only 1% of aneurysms are located in the persistant trigeminal artery [26]. In our study, none of the two cases displayed a coexisting aneurysm.

In conclusion; congenital anomaly and variations increase the incidence of saccular aneurysm by inducing various hemodynamic stresses. In order to reveal the hemodynamic factors taking part in the underlying pathological mechanisms of aneurysms, variations should be well described and the correlations between variation-anomalies and location of aneurysm should be studied.

In this study, the incidence of ACA A1 segment hypoplasia-agenesis was higher than those reported in the literature.

Our results were consistent with the literature reporting higher risk for patients with ACA A1 segment hypoplasia-agenesis, azygos ACA, and MCA trifurcation variations as compared to those with no variation.

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