

Bilateral Internal Carotid Artery Hypoplasia and Multiple Posterior Circulation Aneurysms. Importance of 3DCTA for the Diagnosis

Bilateral İnternal Carotid Arter Hipoplazisi ve Multipl Posterior Sirkülasyon Anevrizması. Tanıda 3DCTA'nın Önemi

ABSTRACT

We present a case with bilateral internal carotid artery hypoplasia and multiple posterior circulation aneurysms who was diagnosed following a subarachnoid hemorrhage. The patient was admitted to our clinic with a history of sudden and severe headache with short-term loss of consciousness and being unable to open the right eyelid five days ago. Nuchal rigidity and right partial ophthalmoplegia were found during the examination. Computed tomography revealed a subarachnoid hemorrhage. Digital subtraction angiography showed bilateral internal carotid artery hypoplasia while three-dimensional computed tomographic angiography showed bilateral internal carotid artery hypoplasia and multiple posterior circulation aneurysms. The aneurysms arising from the right posterior cerebral artery (P1 segment) and left superior cerebellar artery region were clipped using the right modified pterional approach. Asymptomatic unilateral or bilateral internal carotid artery hypoplasia may not be an important problem. However, other concurrent anomalies may be potentially life-threatening. These aneurysms must be treated due to the marked hemodynamic stress even if they have not ruptured and are asymptomatic. It may not be possible to see the aneurysm with digital subtraction angiography in these cases due to superimposition. Three-dimensional computed tomographic angiography provides more detailed diagnostic information.

KEYWORDS: Aneurysm, Internal carotid artery hypoplasia, Computed tomographic angiography, Angiography, Artery hypoplasia

ÖZ

Bu yazıda, bilateral internal carotid artery hipoplazisi ve multipl posterior sirkülasyon anevrizması olan ve subaraknoid kanama sonrası tanı alan bir olgu sunuldu. Hasta beş gün önce oluşan ani ve şiddetli baş ağrısı, kısa süreli bilinç kaybı ve sağ göz kapağını açamama yakınması ile kliniğimize kabul edildi. Muayenede ense sertliği ve sağda parsiyel oftalmopleji saptandı. Bilgisayarlı tomografide subaraknoid kanama saptandı. Dijital subtraksiyon anjiyografide bilateral internal carotid arter hipoplazisi ve yine üç boyutlu bilgisayarlı tomografik anjiyografide bilateral internal carotid arter hipoplazisi ve multipl posterior sirkülasyon anevrizması saptandı. Hasta sağ modifiye pterional yaklaşımla opere edilerek sağ posterior serebral arter ve sol serebellar arter bölgesinden doğan anevrizmalar klibe edildi. Asemptomatik unilateral veya bilateral internal carotid artery hipoplazisi önemli bir problem olmayabilir. Ancak birlikte olduğu diğer anomalilerle birlikte yaşamı tehdit edici etkisi olabilir. Bu anevrizmalar kanamamış-asemptomatik olsalar bile hemodinamik stres yüksekliği nedeni ile tedavi edilmelidir. Bu olguların tanısında burada sunulan olguda olduğu gibi dijital subtraksiyon anjiyografide süperimpozisyon nedeni ile anevrizma görülmeyebilir. Bu nedenle tanıda üç boyutlu bilgisayarlı tomografik anjiyografi daha yeterli bilgiler verir.

ANAHTAR SÖZCÜKLER: Anevrizma, İnternal carotid arter hipoplazisi, Bilgisayarlı tomografik anjiyografi, Anjiyografi, Arter hipoplazisi

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Received : 15.12.2008

Accepted : 11.03.2009

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INTRODUCTION

Internal carotid artery (ICA) hypoplasia is a rare congenital anomaly. Bilateral hypoplasia has been encountered in approximately 20 of the 100 cases reported so far (2,3,7-9,13,14,22). The actual number is probably higher as most cases are asymptomatic (2). The intracranial aneurysm rate was also found to be higher than the general population in these cases (1,9,12,22). Digital subtraction angiography (DSA) may be inadequate in diagnosing bilateral ICA hypoplasia cases due to superimpositions as the complete polygon of Willis receives its blood supply from the vertebral arteries only. More detailed diagnostic images may therefore be obtained with three-dimensional computed tomographic angiography (3DCTA). We present a case diagnosed with bilateral ICA hypoplasia and multiple posterior circulation aneurysm together with subarachnoid hemorrhage (SAH) who underwent surgery for the aneurysms. We also review the literature and emphasize the importance of 3DCTA.

CASE REPORT

A 49-year-old male patient presented at another healthcare center 5 days ago with sudden and severe headache and short-term loss of consciousness together with being unable to open the right eyelid. The patient was referred to our clinic from this center and admitted. He was conscious, oriented and cooperating during the examination (Yaşargil SAH Grade 2b). Right palpebral ptosis was present. No other pathology was detected in system examination. Noncontrast cranial CT showed a local hemorrhage in the right ambient and interpeduncular cisterns (Figure 1). Xanthochromia was found during the lumbar puncture (LP). DSA showed hypoplastic right and left ICA with a contrast injection to the common carotid arteries (CCA's) and it was only possible to observe the external carotid arteries (ECA) (Figure 2 A,B). During vertebral angiography, the posterior cerebral arteries (PCA's), anterior cerebral arteries (ACA's), middle cerebral arteries (MCA's) and the ICA supraclinoidal segments were observed to receive their blood supply from the vertebral artery. It was not possible to observe an aneurysm due to superimpositions (Figure 3 A,B).

3DCTA showed hypoplasia of both ICA's (Figure 4 A). 3DCTA also revealed two aneurysms originating from the right PCA (P1) and between the left superior cerebellar artery and left PCA (Figure 5 A,B). The patient was operated on. The aneurysms on both sides were clipped using a right modified pterional (anterior

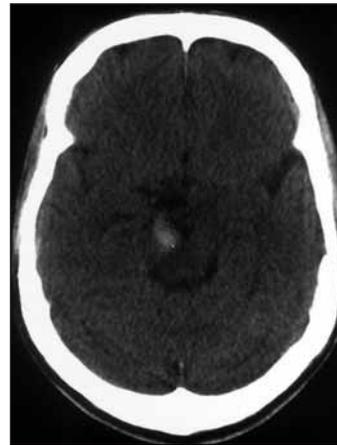


Figure 1: Non-contrast cranial CT shows a local hyperdense area in the right ambient and interpeduncular cisterns.



Figure 2: DSA shows that both ICA's are hypoplastic (white arrows) and that only the ECA's can be observed during right (A) and left (B) CCA contrast injection.

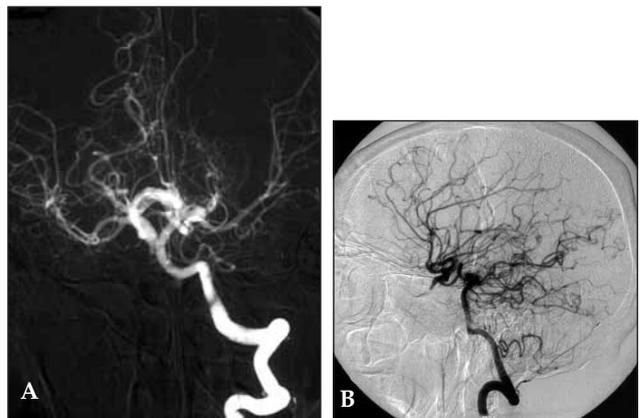


Figure 3: Vertebral angiography in the A-P (A) and lateral (B) positions shows that both PCA's, both ACA's, both MCA's and the supraclinoidal segments of ICA's derive their blood supply from the vertebral artery. However, the aneurysm is not seen due to superimpositions.

temporal) approach. A temporary clip was not used and the patient was kept normotensive during the surgery. The right ophthalmoplegia gradually recovered from the 3rd postoperative day. Postoperative 3DCTA showed that the clips were in the proper position with no remnants or vascular occlusion (Figure 6). The patient was discharged on the 15th postoperative day.



Figure 4: 3DCTA shows right and left ICA hypoplasia (white arrows).

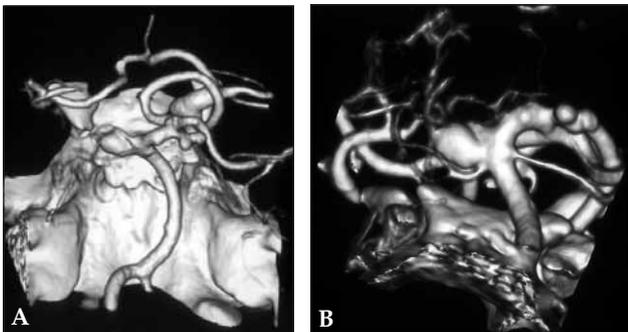


Figure 5 A,B: 3DCTA shows two aneurysms originating from the right PCA (P1) and between the left PCA origin and left superior cerebellar artery origin.

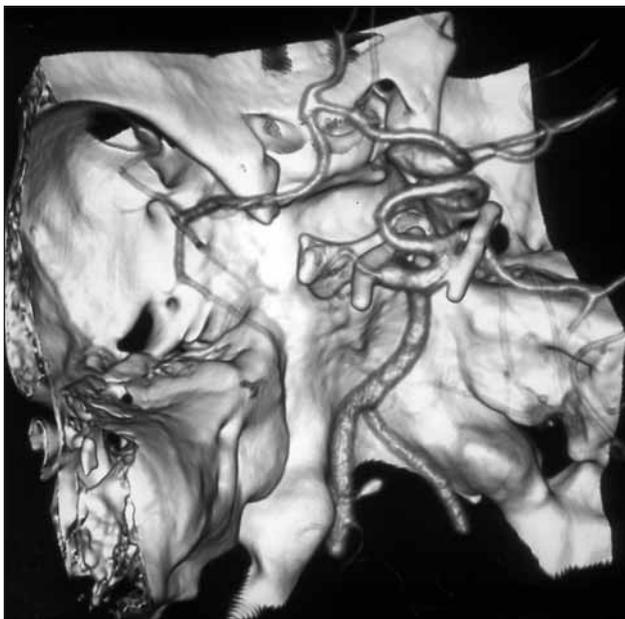


Figure 6: Postoperative 3DCTA showing the clips in the proper position with no remnants or vascular occlusion.

DISCUSSION

The terms ICA agenesis, aplasia and hypoplasia are generally used to denote the same condition in the literature. ICA hypoplasia is a rare congenital anomaly with an incidence below 0.01% (13,18). Unilateral ICA hypoplasia has been reported to be more frequent on the left (4,5,7,16). However, Lee (13) has reported hypoplasia mostly on the right in 9 reported cases. Bilateral ICA hypoplasia is very rare (2,13,15). The intracranial aneurysm rate is 2-4% in the normal population but has been found to be as high as 25-43% in cases with ICA hypoplasia (1,12,17). Lee (13) has reported a rate of 67% in a 9-patient study. Two mechanisms have been used to explain the association between ICA hypoplasia and intracranial aneurysm. Both pathologies may develop independently due to a developmental error during embryonic life or the aneurysm may be secondary due to an unsuitable hemodynamic status (1,4,17).

Collateral circulation is found in ICA hypoplasia cases. This collateral circulation may be in the form of a transcranial anastomosis with the external carotid artery (so called "rete mirabile"), with persistent embryonic arteries such as the trigeminal artery, etc. or with the normal anastomotic channels in the Circle of Willis (13,19). The collateral circulation was from the normal anastomotic arteries in the case presented here. Both supracavernous ICA's, both ACA's and both MCA's were supplied from the basilar artery via the posterior communicating artery. Patients with unilateral or bilateral ICA may be completely asymptomatic thanks to adequate collateral circulation. Despite a significant cerebrovascular anomaly in these cases, MRI and SPECT may not show any cerebral perfusion disorder (6). However, the majority of the reported cases have been diagnosed following a clinical presentation such as headache, epilepsy, cerebral ischemia, hemiplegia and SAH (2,3,7,12,13,15,22). DSA continues to play an important role in the radiological diagnosis of cerebral vascular pathologies. However 3DCTA has started to replace DSA in the diagnosis of cerebral vascular pathologies and especially intracranial aneurysms. The advantages of 3DCTA over DSA in aneurysm diagnosis are that it is a non-invasive technique not requiring artery puncture or catheterization, it can easily be performed by administering intravenous contrast material, it is possible to obtain multiplanar images, detailed information can be constructed on aneurysm morphology and vascular relations and it is less

expensive (10,11,20,21). 3DCTA is especially useful when the aneurysm cannot be seen with DSA due to superimposition as in the presented case. We also obtained very highly detailed images with postoperative 3DCTA that would not be possible with DSA.

In conclusion, symptomatic unilateral or bilateral ICA hypoplasia may not be a significant problem. However, other concurrent anomalies and especially aneurysms may be life-threatening conditions. These aneurysms should be treated even if they have not ruptured or are asymptomatic due to the marked hemodynamic stress. It is difficult to determine a size limit for aneurysms concurrent with ICA hypoplasia that have not ruptured due to the inadequate number of cases and follow-up duration in the literature. Endovascular methods or the open surgical method are used for treatment according to the surgical experience and technical adequacy of the clinic, and the patient's clinical status. Intraoperative hypotension and temporary clip usage should be avoided to prevent irreversible neurological deficits.

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