ISOLATED HYPOPLASIA OF DISTAL BASILAR ARTERY: CLINICAL AND IMAGING FINDINGS

Nilda Turgut*, Gökhan Pekindil, Ufuk Utku*, Yahya Celik*, Semra Şengün***

ABSTRACT

Basilar artery hypoplasia usually includes entire or proximal part of the artery and commonly associated with hypoplastic vertebral arteries. Here we present a case of isolated hypoplasia of distal basilar artery associated with infarction of medulla oblongata, which has not previously been reported. MRI showed infarction and reduction in calibration of distal basilar artery whereas non-contrast cranial CT and color Doppler sonography of vertebral artery revealed normal findings. 3D-TOF MRA demonstrated regular uniform narrowing of distal part of basilar artery, which was confirmed by catheter angiography.
Keywords: angiography, basilar artery, hypoplasia, magnetic resonance imaging

DİSTAL BAZİLER ARTERİN İZOLE HİPOPLAZİSİ: KLİNİK VE GÖRÜNTÜLEME BULGULARI

ÖZET

Baziler arter hipoplazisi, baziler arterin tamamında veya proksimal bölümünde gelişmekte ve sıklıkla vertebral arter hipoplazisi eşlik etmektedir. Bu yazıda, medülla oblongatada infarktı olan, izole distal baziler arter hipoplazisi tespit edilen hasta sunulmuştur. MR'da infarkt gözlenmiş, distal baziler arterde kalibrasyon azalmış olarak tespit edilmiştir. Hastanın yapılan kontrastsız kranial BT'si ve vertebral arter renkli dopler ultrasonografisinde normâl bulgular elde edilmiştir. 3D-TOF MRA'de, kateter anjiografi bulgularına benzer olarak, baziler arterin distal bölümünde düzenli bir daralma tesbit edilmiştir.
Anahtar Kelimeler: anjiografi, baziler arter, hipoplazi, magnetik rezonans görüntüleme

INTRODUCTION

Hypoplasia of the basilar artery is rarely encountered anomaly of the vertebrobasilar system and frequently accompanied by persistant primitive trigeminal artery (PPTA) and/or hypoplastic vertebral artery (Boyko et al. 1996, Szdzuy and Lehmann 1972, Hegedüs 1985, Chaturvedi et al. 1999). Basilar artery hypoplasia includes proximal or whole part of the vessel in these cases. To date, isolated hypoplasia of distal basilar artery with normal vertebral arteries has not been reported, although incomplet fusion of distal part of the basilar artery has been described as distal hypoplasia in a case report by Szdzuy and Lehmann (Szdzuy and Lehmann 1972). Moreover, demonstration of these hypoplastic vessels may especially be of clinically important, since it has recently been suggested that hypoplastic vertebrobasilar vessels should be considered among the potential causes of cerebral ischemia in young adults (Chaturvedi et al. 1999). To date, symptomatic entire basilar artery hypoplasia has been described only in 13 cases by three previous reports (Szdzuy and Lehmann 1972, Hegedüs 1985, Chaturvedi et al. 1999).

Here we present computed tomographic, magnetic resonance imaging (MRI), MRA and digital subtraction angiographic (DSA) findings of a case with isolated hypoplasia of distal basilar artery associated with infarction of medulla oblongata.

CASE REPORT

A 50-year-old male patient with previous history of hypertension and smoking was admitted with acute onset of left hemiparesis, truncal ataxia, dysphagia and hiccup. Neurological examination revealed a left sided hemiparesis with truncal ataxia, right facial hypoestesia, and right-sided paralysis of pharinx. He was alert, cooperated and oriented. Non-contrast computed tomographic scan at admission was normal. However since the posterior circulation infarction was clinically thought, color Doppler sonography of the extracerebral carotid and vertebral arteries and cranial MRI was planned. Color Doppler sonography of the vertebral arteries revealed normal findings. Cranial MRI (using Siemens Magnetom 1 Tesla, with T1Weighted axial, T2Weighted axial and coronal, and proton density axial fast spin-echo images) performed four days later showed a small infarction in the right paramedian portion of the medulla oblongata (Fig.1.a). In addition, reduction in the size of distal part of basilar artery was suspected on T2-we-

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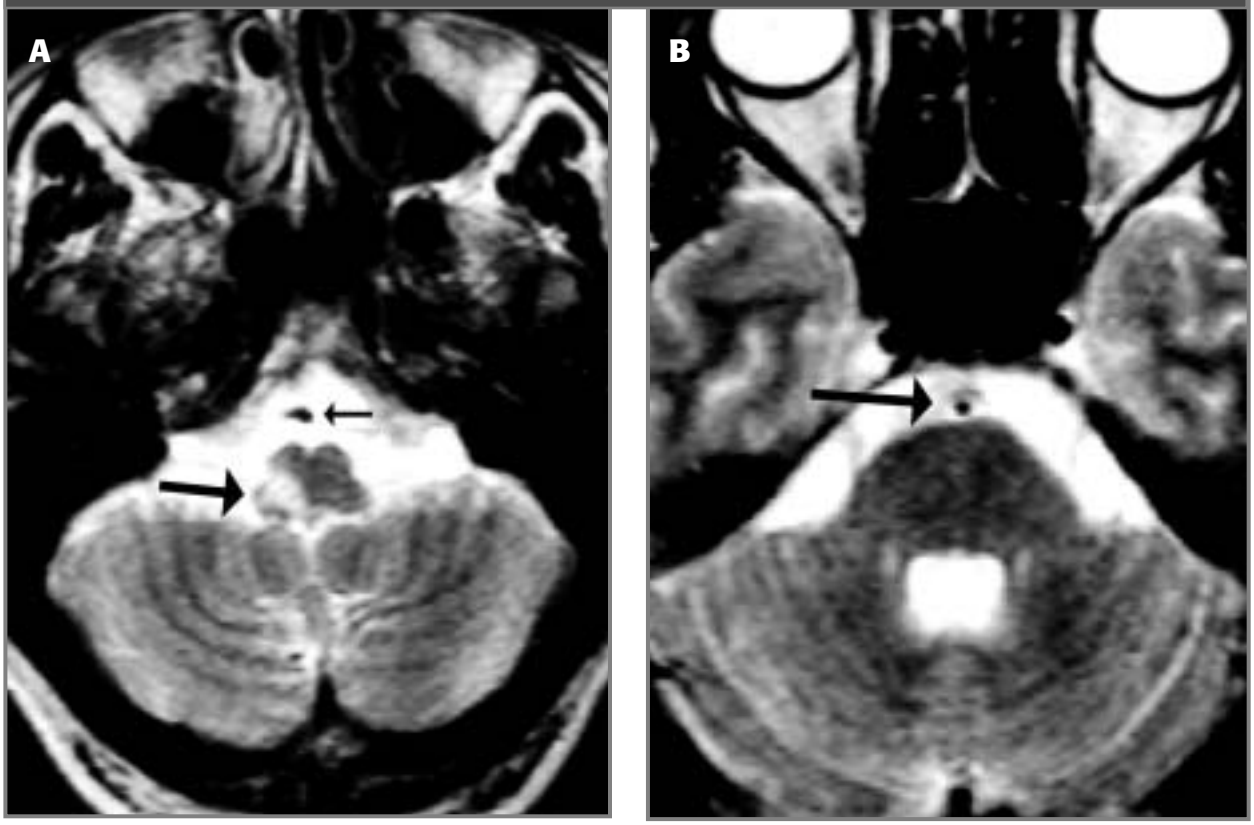
Figure 1.a) Axial T2-weighted MRI shows infarction on the right side of medulla oblongata (large arrow) and normal sized proximal basilar artery (small arrow) b) whereas the calibre of basilar artery (arrow) is decreased distally at the level of pons.



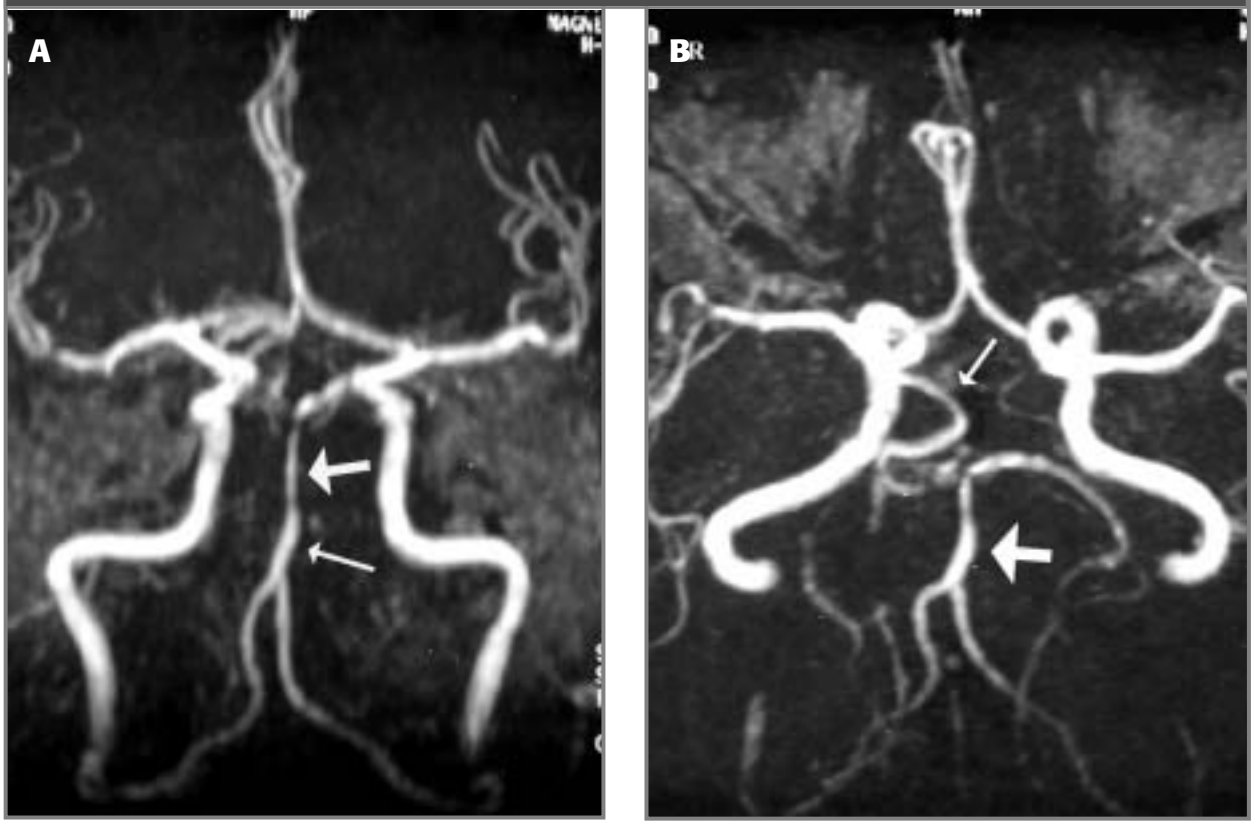
ighted axial MRI slices, therefore MRA was also performed to clarify this finding (Fig.1.b). Non-contrast-MRA using gradient echo technique (fast imaging with steady-steate precession (FISP), three-dimensional time-of-flight (3-D TOF), repetition time 30 miliseconds, echo time 9.6 miliseconds, flip angle 20º, field of view 165x220, matrix 200x512, slab thickness 38 mm, number of excitations 1, partitions 16) revealed narrowing in upper half of the basilar artery and elongated and large right posterior communicating artery whereas vertebral arteries were bilaterally patent and symmetrically in normal calibration (Fig.2 a,b). Catheter angiography performed using femoral approach and, two internal carotid and left vertebral arteries were injected. Angiography confirmed these findings and revealed regular, uniform narrowing of distal half of the basilar artery and additionally showed fetal type left posterior cerebral artery arising from left internal carotid artery (Fig. 3). The patient was treated with antiaggregant and antihypertensive therapy. Two weeks later, his sign's partially resolved and patient was discharged.

DISCUSSION AND CONCLUSION

Basilar artery anomalies are rare and include most commonly duplication or fenestration and so rarely hypoplasia, segmental aplasia, plexiform apperance etc. (Stehbens 1986, Yaşargil 1984, McCullogh 1962, Lasjaunias et al. 1979).

The cause of the hypoplasia of the basilar artery is not known as other arterial abnormalities. Embryologically, the posterior circulation begins as two paired plexiform longitudinal neural arteries and they start to fuse to form the basilar artery at ~5 weeks gestation while the trigeminal artery begins to involute (Padget 1948). The size of an artery depends on the area that ultimately supplies and an artery becomes unnecessary during development undergoes regression (Stehbens 1986). Therefore basilar artery hypoplasia is believed to be consequence of the PPTA (Fields 1968). It has also been suggested that large posterior communicating artery, which was commonly seen in these cases, may show persistent flow from carotid to vertebrobasilar circulation and this may cause vertebrobasilar hypoplasia (Hegedüs 1985). Although PPTA was not detected, there was large right pos-

Figure 2.a) MRA demonstrates normal sized proximal (small arrow) and uniformly-regularly narrowed distal part of the basilar artery (large arrow). b) Note elongated and prominent right posterior communicating artery (small arrow) and proximal basilar artery (large arrow).

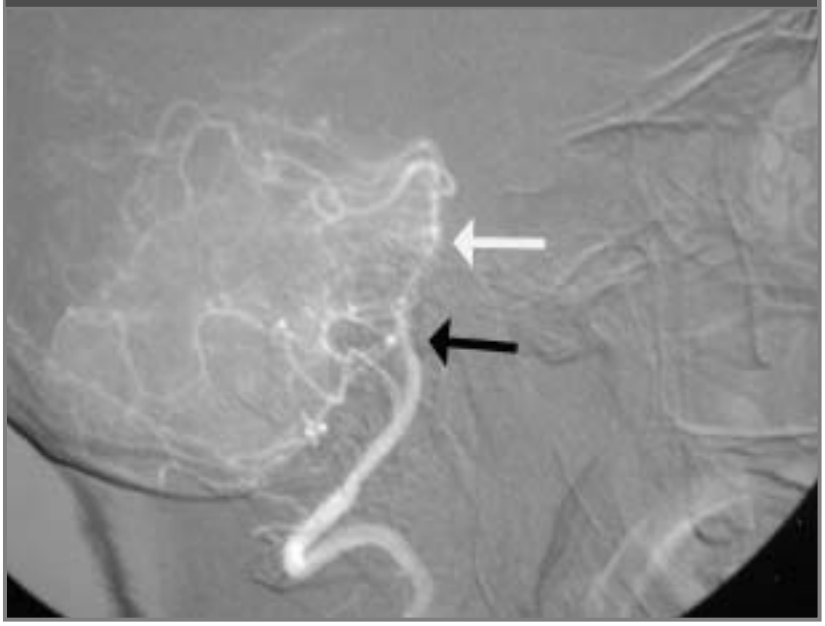


terior communicating artery in our case. In addition, malformations, injuries that effect acting either at the perinatal period (such as basal meningitis, arteritis, arterial occlusion) or in early childhood (trauma?) may impede the normal reproduction of the smooth muscle cells in the media from maintaining the capacity of the artery to grow with the brain have been suggested in development of hypoplastic arteries (Hegedüs 1985, Fisher 1959, Lhermitte et al. 1968). However, we could not detect any of these above-mentioned factors in the history of our case.

When associated with persistent primitive trigeminal artery, basilar artery hypolasia occurs commonly at proximal part of the vessel and usually associated with vertebral artery hypoplasia (Boyko et al. 1996). Atherosclerotic stenosis most commonly involve middle portion of basilar artery with irregular and non-uniform appearance (Djang and Drayer 1990). Fibromuscular dysthrophy usually demonstrates multifocal stenosis with adjacent dilatations, the so-called string of beads (Healthon 1986). Basilar artery fenestration most commonly occurs in the lower half of the vessel, extending for a lenght of less than 5 mm (Goldstein et al. 1999).

Prior studies of stroke in young adults have not included hypoplastic cerebral vessels among the potential causes of cerebral ischemia (Caplan 1996, Adams et al. 1995, Lisovski F and Rousseaux 1991). The rate of symptomatic entire basilar artery hypoplasia that was detected from ~ 4000 cases evaluated for stroke or transient ischemic attack was recently reported as (1/500%) by Chaturvedi et al (Chaturvedi et al. 1999). They suggested that hypoplastic basilar artery might be a predisposing factor for ischemic stroke since the mean age of all cases was 49.8 (Chaturvedi et al. 1999). Hegedüs demonstrated autopsy findings of three cases of entire basilar artery hypoplasia associated with vertebral hypoplasia in which two of them had neurological symptoms characteristic of insufficiency in the vertebrobasilar system (Hegedüs 1985). Szdzuy and Lehmann (1972) described angiographic findings of incomplete fusion of distal part of the basilar artery associated with vertebral artery hypoplasia in two cases presented with symptoms of brainstem ischemia and termed this condition as distal hypop-

Figure 3. Catheter angiography lateral view confirms regularly narrowed distal (white arrow) and normal sized proximal basilar artery (black arrow).



lasia. However the appearence of our case is different from all these descriptions and appears to reflect an original condition.

Although vertebral or posterior inferior cerebral artery lesion was clinically thought for medullar infarction, these vessels were patent on both MRA and angiography in our case. Therefore microtrombotic occlusion of medullary branches of these arteries was proposed. It was speculated that poor retrograde flow due to hypoplastic distal basilar artery might make easy occurence of infarction in our case. Demonstration of these hypoplastic narrowing is also of importance since atherosclerotic disease may also appear at an earlier age if the native vessel is hypoplastic and would become stenosed sooner than a large vessel (Fischer et al. 1965). Moreover, embolic occlusions tend to involve the distal basilar segment and usually result fatally (Djang and Drayer 1990).

It has been reported that MRA successfully shows atherosclerotic stenoses of arterial system in cases with vertebrobasilar ischemia, although the degree of stenosis was difficult to evaluate and was both over- and underestimated with similar percentage (Röther et al. 1993, Ruggieri et al. 1994). It has recently been reported that MRA can also demonstrate hypoplasia of entire basilar artery (Chaturvedi et al. 1999). Additionally, our case confirms that MRA can also reveal partially (distally) hypoplastic vessel in the basilar system. It should be noted that color Doppler sonographic examination of vertebral arteries, and non-contrast cranial CT may be normal in distal basilar hypoplasia. Moreover, transcranial Doppler sonography has not found sensitive to lesions of the mid and distal basilar artery (Tettenborn et al 1990). This entity should be suspected on the basis of MRI, if basilar artery calibre is decreased distally. However angiography is required to provide definitive diagnosis. We also conclude that arterial abnormalities as a predisposing factor should always be investigated in young stroke cases, and as a non-invasive method, MRA should be considered in diagnosis of basilar abnormalities.

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