Case report

Hypoplasia of the internal carotid artery: a noninvasive diagnosis

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Abstract. We present the characteristic imaging findings of hypoplasia of the internal carotid artery (ICA) in two cases, one accompanied by an intracranial aneurysm. Finding of a diffuse luminal narrowing of the ICA on MR angiography or digital subtraction angiography (DSA) could wrongly evoke severe acquired diseases such as dissection or atherosclerosis. Absence of associated wall thickening and flow disturbances on color Doppler sonography (CDS) should suggest carotid hypoplasia. Confirmation of the diagnosis is obtained by CT of the skull showing a small carotid canal. Non-invasive procedures are sufficient to differentiate this rare congenital anomaly from acquired string signs.

Key words: Carotid arteries – CT – MRI – US – Hypoplasia

Introduction

Hypoplasia of the internal carotid artery (ICA) is a very rare anomaly of the embryonic development. Since 1968, approximately 30 cases have been reported in the literature, 20 of them being bilateral, and most of these were based on conventional angiographic studies [1, 2, 3]. This congenital anomaly is usually asymptomatic because of sufficient collateralization from the circle of Willis [2]; however, it has to be recognized because of its frequent association with intracranial aneurysms [4, 5, 6] and not to be confused with more serious carotid conditions.

We present two cases of unilateral hypoplasia of the ICA assessed by color Doppler sonography (CDS), MR angiography, and CT, for the first one; and by CDS, digital subtraction angiography (DSA), and CT for the second one. The former is the first case of carotid hypoplasia to be elucidated without the performance of invasive imaging procedures. In this article we discuss the value of the different imaging modalities, especially non-invasive ones, and emphasize the differential diagnosis with acquired carotid diseases such as dissection and atherosclerosis.

Case reports

Case 1

A 74-year-old woman was admitted to our radiologic department because of suspected dementia. Physical and neurological findings were normal except for cognitive deterioration. Magnetic resonance performed with a 1-T unit (Magnetom Expert, Siemens, Erlangen, Germany) excluded any curable cause to the patient’s dementia. On the transverse T2-weighted turbo spin-echo images, lack of normal flow void in the left cavernous sinus suggested the possibility of an anomaly of the ICA. A dynamic contrast-enhanced MR angiography of the carotids was then carried out in the coronal plane using a fast three-dimensional (3D) sequence, a dedicated head and neck coil, and a bolus injection of 20 ml of gadopentetate dimeglumine performed by hand and followed by a 20-ml saline solution flush. Parameters of the sequence included: TR 40 ms; TE 1.65 ms; flip angle 25°; number of slices 64; section thickness 1 mm; matrix 105 × 256; field of view (FOV) 260 mm; and number of excitations 1. A series of four consecutive 3D scans of 10 s each was obtained, starting at approximately the same time as the bolus injection, the total duration of which did not exceed 15 s. The first precontrast data set was subtracted from the data set of the second scan obtained during arterial phase to eliminate background noise, and subsequent subtracted images were chosen for maximum intensity projection (MIP) reconstruction. The MIP images showed a smaller left common carotid artery (CCA), a normal external carotid artery (ECA), and an abnormally thin left ICA (Fig. 1a). In order to rule out
severe stenotic disease of the ICA, color Doppler imaging was performed immediately: It showed the left ICA to be patent without any parietal thickening. A pulsed Doppler sonogram demonstrated normal flow waveform within the left ICA, with a peak systolic velocity (PSV) of 30 cm/s and a normal resistance index (Fig.1b). B-mode sonography confirmed reduction of size of both left ICA and CCA but revealed that this reduction not only involved their internal but their external diameter as well. The transversal external diameters of the left CCA measured 6.6 mm (vs 8.9 mm on the right; Fig.1c). Furthermore, an intracranial aneurysm was suspected on the first MR angiography and, the following day, another MR angiography was performed in the transverse plane on the circle of Willis using time-of-flight (TOF) technique and no contrast injection, which confirmed a 6-mm aneurysm on the supraclinoid segment of the right ICA (Fig. 1d). The left A1 segment

Fig.1a–e. Hypoplasia of the left internal carotid artery (ICA) in a 74-year old woman. a Maximum intensity projection (MIP) reconstruction of dynamic contrast-enhanced MR angiography (frontal view) demonstrates a diffusely thin left ICA (arrows). b Color Doppler sonography shows a smaller-than-normal left ICA with normal flow. c Color Doppler sonography shows that the external diameter of the ipsilateral common carotid artery (CCA) is also reduced compared with the right CCAs. d Three-dimensional time-of-flight MR angiography MIP projection (frontal view) obtained the day after the first MR angiography confirmed and better depicted the prior suspected aneurysm on the supraclinoid segment of the right ICA (arrow). e A CT of the skull base exhibits a congenitally narrowed left carotid canal (arrow)