Cerebrovasc Dis 2007;24:543–544
DOI: 10.1159/000111221

Spontaneous Carotid Artery Dissection in a Patient with Turner Syndrome

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Turner syndrome (TS) is the most common chromosomal abnormality in females, affecting 1:2,500 live female births [1]. It results from complete absence or partial deletion of one X chromosome. Aortic dissection is a well-recognized complication of TS [1, 2]. However, spontaneous internal carotid artery (ICA) dissection has not been previously reported.

Case Report

A 37-year-old woman presented to an outside hospital with acute onset of left-sided weakness and an NIHSS score of 22. She received intravenous tissue plasminogen activator and was transferred to our institution 6.5 h after symptom onset. Her medical history was remarkable for TS and hypothyroidism. Her medications included thyroid hormone and estrogen supplementation therapy. Her neurological examination revealed a left homonymous hemianopsia, left-sided sensory neglect and left hemiparesis.

Brain magnetic resonance imaging showed a large right middle cerebral artery territory infarction and minimal subarach-

noid hemorrhage (SAH). A neck CT angiography (CTA) showed marked narrowing of the right ICA distal to the bulb, with irregular and tapered appearance (fig. 1a, b) suggesting ICA dissection. Intracranial extension of the dissection was suspected due to the presence of SAH.

A transesophageal echocardiogram revealed a dilated ascending aorta and no evidence of aortic dissection. The remainder of the stroke workup was negative. The patient was treated with antiplatelet therapy and had an uncomplicated hospital course.

Discussion

Patients with spontaneous dissection of the carotid and vertebral arteries are suspected to have underlying structural defects of the arterial wall [3]. Approximately 5% of patients have at least one family member who had a spontaneous dissection of the aorta or its main branches [3]. However, the identification of a specific underlying arteriopathy remains elusive in most cases.

Cystic medial necrosis of the arterial wall is a common finding at postmortem examination of patients with spontaneous artery dissection [3]. This finding has been reported in 71% of 25 reported cases of TS complicated by aortic dissection in which histology was available [1]. The arterial wall abnormalities found in TS patients are not restricted to the aorta. Instead, patients with TS appear to have a more diffuse vasculopathy. Arterial dilatation, enlargement and increased intimal thickening similar to those seen in the aorta have been demonstrated affecting the carotid and brachial arteries [4]. Theoretically these underlying arterial wall abnormalities could lead to a higher risk for spontaneous dissection of the affected vessels of TS patients.

Fig. 1. **a** Neck CTA axial view shows right ICA dissection. **b** Neck CTA lateral view shows right ICA with irregular and tapered appearance. Recanalization extending to the petrous ICA is also noted.

We found of interest that despite the underlying diffuse arterial wall abnormalities observed in patients with TS, there are no prior reports in the literature of carotid dissection and there is a single case report of bilateral vertebral artery dissection associated with dissection of the aorta [5]. Further studies are needed to determine whether the occurrence of arterial dissection of the neck vessels in patients with TS presenting with stroke is an underdiagnosed disease or a rare association.

Conclusion

Arterial dissection of the neck vessels should be suspected in patients with TS presenting acute ischemic infarction. CTA can be useful in establishing the diagnosis.

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Cerebrovasc Dis 2007;24:544–546

DOI: 10.1159/000111222

USPIO-Enhanced MRI of Neuroinflammation at the Sub-Acute Stage of Ischemic Stroke: Preliminary Data

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Introduction

Postischemic cerebral inflammation may be involved in delayed ischemic brain damage. The development of noninvasive imaging methods is a critical issue and might allow the assess-

ment of neuroprotective therapies. According to experimental data, brain inflammation is present at the acute stage of ischemic stroke [1]. Ultra-small superparamagnetic particles of iron oxide (USPIO)-enhanced MRI may allow noninvasive monitoring of neuroinflammation [2–5]. In a previous study [5], we investigated 10 patients who presented with acute anterior circulation stroke. USPIO was administered on day 6 and signal alterations following USPIO injection were observed in 9/10 patients on day 9 T₁-weighted imaging. Hematogenous macrophage may act as the main target for USPIO uptake. As far as we know, no data investigating USPIO uptake at an earlier time window has been published in human stroke.

Patients and Methods

We designed a sequential MRI protocol in order to assess brain inflammation in patients with ischemic stroke. We enrolled patients who presented with documented acute anterior circulation stroke, with an NIHSS score ≥ 8 , and who were not eligible for thrombolytic therapy. Oral and written informed consent was obtained prior to inclusion. Major exclusion criteria were: (1) brain hematoma on initial brain CT scan or MRI; (2) Initial MRI performed more than 24 h previously; (3) ambiguous time of symptom onset; (4) lesion size below 1 cm³ on diffusion-weighted imaging on initial stroke imaging; (5) enrolment into other clinical studies; (6) Past history of neoplasia or known active liver disease; (7) Administration of gadolinium complexes within 24 h or iron particles within 6 months prior to this study; (8) Known allergy to dextran or drugs containing iron salts, and (9) current contraindications to MR imaging. The study design was approved by our ethical committee (CCPPRB Lyon B) [5].

The MRI protocol included T₁-weighted imaging (T1WI), gradient-echo T₂* imaging (T2*WI), diffusion-weighted imaging (DWI), perfusion-weighted imaging and MR angiography. Blood-brain barrier (BBB) disruption was defined as post-gadolinium enhancement on T₁-weighted images at day 2 before USPIO administration. BBB disruption was classified as absent, mild (<1/3 of the MCA territory), or severe (>1/3 of the MCA territory). USPIO (ferumoxtran, AMI-227, Sinerem®) was provided by Guerbet (Roissy CDG Cedex, France). It was administered by a single dose infusion (2.6 mg iron/kg body weight) through a 0.22- μ m pore filter at a rate of 4 ml/min immediately after day 2 MRI. Volume of signal changes attributed to USPIO were assessed both on T₁- and T₂*-weighted imaging at day 4. Volumes of USPIO-related signals were delineated manually, by the consensus of two neuroradiologists, in cerebral parenchyma, excluding large vessels. Since the analysis of USPIO-related signal changes could be affected by hemorrhagic transformation (HT) of brain infarction, the volume of HT-related signal changes was assessed at day 2 T₁- and/or T₂*-weighted imaging, and was removed from the day 4 USPIO signal change volume.

Results

The protocol was completed in 5 patients (4 women and 1 man): median age, 67 years (range, 52–75); mean day 2 DWI lesion volume, 111 cm³ (range, 28–201); median NIHSS score at day 0, 14 (range, 10–24). Three patients showed evidence of hemorrhagic transformation on day 2 T2*WI. A BBB disruption was observed in three patients. Parenchymal signal change attributable to USPIO was seen in a single patient on day 4 T1WI, and was confined within the ischemic lesion. A BBB disruption was ob-