Corkscrew angiopathy of intracranial vessels in a young stroke patient: a case report

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Abstract

Introduction: We present a rare finding of a ‘corkscrew appearance’ of the distal cerebral vessels in a young Asian woman who presented with acute stroke.

Case presentation: A 32-year-old Asian woman presented with a 3-month history of recurrent right-sided transient ischemic attacks. Her clinical workup and brain imaging results were normal. A digital subtraction angiogram revealed an abnormal corkscrew appearance of all intracranial distal vessels. She was discharged on a single antiplatelet drug. She had no further transient ischemic attacks on clinical follow-up. A digital subtraction angiogram performed 1 year later revealed no changes in the appearance of these vessels.

Conclusion: To the best of our knowledge no similar previous reports exist in the literature. The present report describes a unique case of an unusual corkscrew appearance of the distal intracranial vessels. However, the underlying etiology in the present case remains unknown.

Keywords: Corkscrew angiopathy, Stroke, Digital subtraction angiography
in young patients. Osborn and Anderson [1] reported a 'string of beads' appearance of the vessels involving the intracranial internal carotid artery and middle cerebral artery in their series of 25 patients with fibromuscular dysplasia (FMD). Four patients in their series had FMD involving all four intracranial vessels. However, the present patient showed no evidence of FMD. The appearance of the angiogram suggested an abnormality of the small vessels. Results of a workup for associated vasculitis were negative, and superficial temporal artery biopsy results were normal. Puca et al. [2] reported a case of a 32-year-old woman with right brachioocular hemiparesis who had suffered from left hemispheric ischemic stroke with intracranial arterial dolichoectasia. On a three-dimensional angiogram they demonstrated dolichoectasia of the middle cerebral artery with an unusual corkscrew appearance. Isolated intracranial FMD presenting as a stroke in a 19-year-old woman was reported by Birnbaum et al. [3], and a cerebral angiogram showed a string of beads abnormality of the proximal middle cerebral artery and anterior cerebral artery, consistent with FMD. A corkscrew appearance of the retinal vessels was reported in 12 of 32 patients (n=32) with neurofibromatosis type 1 [4]. To the best of our knowledge a similar appearance of the small intracranial vessels associated with other conditions has not been reported in the literature.

Dissection as a cause of the present radiological findings was considered because it is an often underdiagnosed cause of stroke in young patients. However, diffuse involvement with a corkscrew appearance of almost all small intracranial vessels makes dissection a very unlikely etiology. Moreover, the typical angiographic findings of intracranial vessel dissection, such as alternating stenosis and dilatation, delayed arterial emptying, and anastomoses, a beaded appearance, or pseudoaneurysm [5], were not seen in any vessels in our case. The presence of normal MRI results usually rules out underlying vasculitis [6].

It is likely that the DSA findings were incidental and unrelated to the TIAs in the present case. However, in the absence of any other possible cause of the TIAs, a causative association cannot be ruled out. Coutts et al. [7] recently reported a case of intracerebral hemorrhage in a young man who presented with stroke. A cerebral angiogram was performed to rule out rare conditions and showed an abnormal corkscrew appearance of the distal vessels that was not typical of any condition. A literature review by these authors suggested the possibility of COL4A1 mutation, and further genetic consultation was done with deoxyribonucleic acid (DNA) sequencing of a COL4A1 complementary DNA product obtained by reverse transcriptase polymerase chain reaction. This investigation evidentially showed a mutation-associated lack of the exon 25 sequence between exons 24 and 26. However, such genetic studies were not performed in the present case because of difficult access to clinical genetic testing centers.

**Conclusion**

We report a possibly unique case of an unusual corkscrew appearance of the small vessels of the intracranial circulation in a young woman who presented with stroke and without any underlying known clinical conditions or associations.
Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

Authors’ contributions
AA analyzed and interpreted the patient data. LSPK also analyzed the data and performed the literature review on these findings and contributed to preparation of the manuscript. SPO assisted in the clinical workup and follow-up of the patient as well as the editing and review of the manuscript. All authors read and approved the final manuscript.

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