JVIN | Journal of Vascular and Interventional Neurology

OFFICIAL JOURNAL OF ZEENAT QURESHI STROKE INSTITUTE

Spontaneous Intracranial Artery Dissection in a Patient with Turner Syndrome

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Abstract

Background— Turner syndrome is a genetic disorder caused by partial or complete monosomy of the X chromosome. Although a few cases of ischemic stroke have been reported in Turner syndrome, but isolated intracranial artery dissection has not yet been reported. We report a case of a patient with Turner syndrome with middle cerebral artery dissection and, present the clinical and radiological features.

Case Description— A 34-year-old woman with Turner syndrome and diabetes mellitus was referred to our hospital with symptoms of left hemiparesis post appendectomy. Magnetic resonance imaging outside hospital showed diffusion restriction in the right middle cerebral artery territory with magnetic resonance angiography (MRA) identifying a right MCA occlusion. Digital subtraction angiography revealed a M1 segment occlusion. Mechanical thrombectomy of the right M1 segment was performed but a triangular flow-limiting stenosis for an underlying dissection. Intracranial stent placement was then performed with successful reperfusion.

Conclusions— Ischemic stroke in Turner syndrome can occur due to dissection of intracranial arteries and may require additional stent placement during mechanical thrombectomy. Further studies are required to investigate the mechanism for predisposition for dissection in Turner syndrome

Keywords— Turner syndrome, Ischemic stroke, Arterial dissection, Mechanical thrombectomy.

INTRODUCTION

Turner syndrome (TS) is a genetic disorder caused by partial or complete monosomy of the X chromosome. The partial or complete monosomy is usually the result of a sporadic chromosomal non-disjunction seen in 50 per 100,000 female births. This syndrome is associated with increased morbidity and mortality, but the affected individuals have an average intellectual level.[1] There are only a few reported cases of ischemic stroke in TS[2-8] but stroke due to isolated intracranial artery dissection has not yet been reported. We report a case of a patient with Turner syndrome with middle cerebral artery dissection and, present the clinical and radiological features.

CASE DESCRIPTION

A 34-year-old woman with TS and diabetes mellitus was referred to our hospital with left hemiparesis, which had developed post emergency exploratory laparotomy and appendectomy for a ruptured appendix. Post-operative, she was difficult to arouse and had left sided hemiparesis. Her computed tomographic scan of head did not identify any acute intracranial findings. She was not a candidate for intravenous tissue plasminogen activator due to recent laparotomy. Magnetic resonance imaging showed diffusion restriction in the right middle cerebral artery territory with fluid- attenuated inversion recovery mismatch (Figure 1) and magnetic resonance angiogram identified a right middle cerebral artery occlusion. The patient was intubated for airway protection and was transferred to our hospital for further management.

General examination revealed a mildly distressed woman with short stature, low hairline, short neck and poorly developed breasts. She was afebrile with a blood pressure of 134/93 millimeter of mercury, heart rate of 140 beats per minute, and respiratory rate of 18 breaths per minute. Neurological examination revealed a right gaze preference, left sided weakness, and sensory loss. We were unable to

Vol.12, No. 2, pp. 11-13, Published November, 2021.

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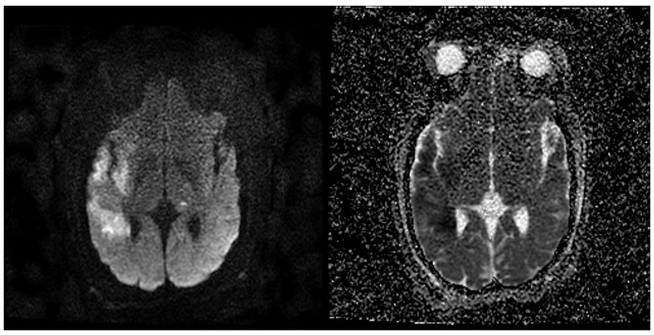

FIGURE 1: Diffusion weighted imaging showing hyper intensity in M2 distribution of right middle cerebral artery (left image) with apparent diffusion coefficient correlate (right image).

assess speech or language as the patient was intubated. Her National institute of health stroke scale score was 18 (Level of consciousness-1, month and age-2, Commands-2, partial gaze palsy-1, visual field-1, motor function left arm-3, motor function left leg-4, sensory-1, Language-3).

Her laboratory tests were unremarkable. On arrival, a computed tomography perfusion imaging was obtained which demonstrated no core infarct, and discrete areas of diminished regional cerebral blood flow specifically in the areas of abnormal diffusion restriction on the recent magnetic resonance imaging examination. The regional cerebral blood volume was relatively preserved and increased mean transit time. The patient was then taken to the neuro endovascular catheter laboratory. Digital subtraction angiography revealed a M1 segment occlusion (Figure 2a and 2b). Mechanical thrombectomy of the right M1 segment was performed but a triangular flow-limiting stenosis remained, suspicious for an underlying dissection. Intracranial stent placement was then performed with successful reperfusion. (Figures 3a & 3b). She was placed on aspirin and ticagrelor and was admitted to the Neurological Intensive Care Unit for further management. She eventually required tracheostomy and percutaneous gastrostomy and was discharged to a long-term care facility with modified Rankin scale of 5.

DISCUSSION

Turner syndrome presents with a myriad of complications with cardiovascular disease as the cardinal trait. Ischemic stroke is infrequently reported in patients with Turner syndrome[6] The documented etiologies of ischemic stroke in TS are fibromuscular dysplasia,[2] congenital hypoplasia of the carotid artery,[3] premature atherosclerosis,[4] Moya Moya syndrome,[5] extracranial artery dissections of vertebral artery,[7] external carotid artery dissection,[8] and bilateral internal carotid arteries dissection,[9] while isolated intracranial dissections has not been previously reported. Aortic dissection is previously described in patients with Turner syndrome. The vascular structures of aorta and, head and neck arteries share a similar embryological origin from pharyngeal arches I-III thus suggesting a diffuse arteriopathy similarly affecting the aorta and cerebral vessels.[10] There appears to be a defect in the collagen of the vessel wall characterized by cystic medial necrosis similar to Marfan's syndrome[11,12] increasing vulnerability to dissection. The cerebral arteries differ from systemic arteries due to absence of an external elastic lamina, sparse elastic fibers in tunica media and a thin adventitia and therefore may be vulnerable to dissection.

CONCLUSION

Ischemic stroke in patients with Turner syndrome can occur due to dissections of either extracranial or intracranial arteries, with the former being more commonly reported. Since it occurred in the setting of sympathetic over activity post-surgery, it is interesting to theorize an association between the two. Intracranial dissection is a relatively rare cause of acute ischemic stroke due to large vessel occlusion and requires a different management paradigm. It is therefore imperative

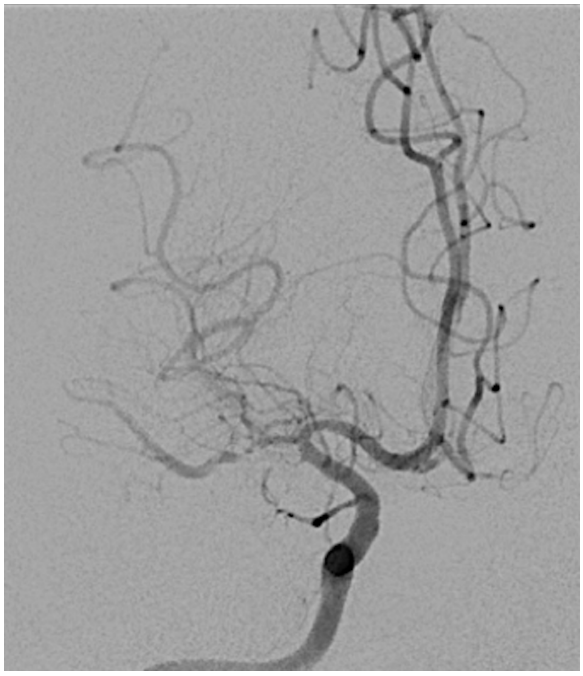

FIGURE 2A: Digital subtraction angiogram demonstrates right M1 occlusion.

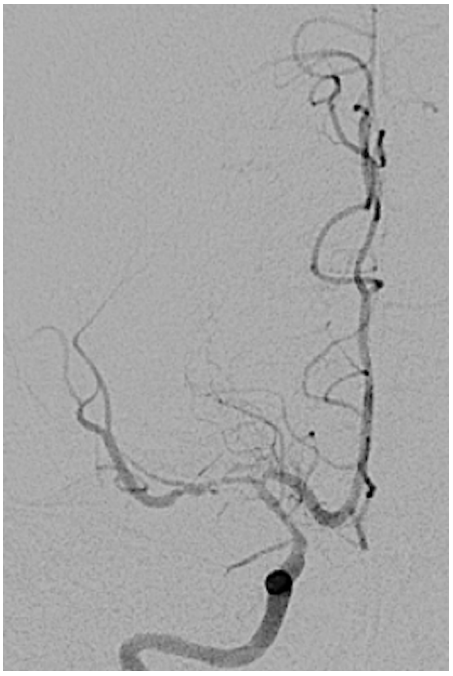

FIGURE 2B: Digital subtraction angiogram demonstrates right M1 segment flow-limiting arterial dissection post thrombectomy.

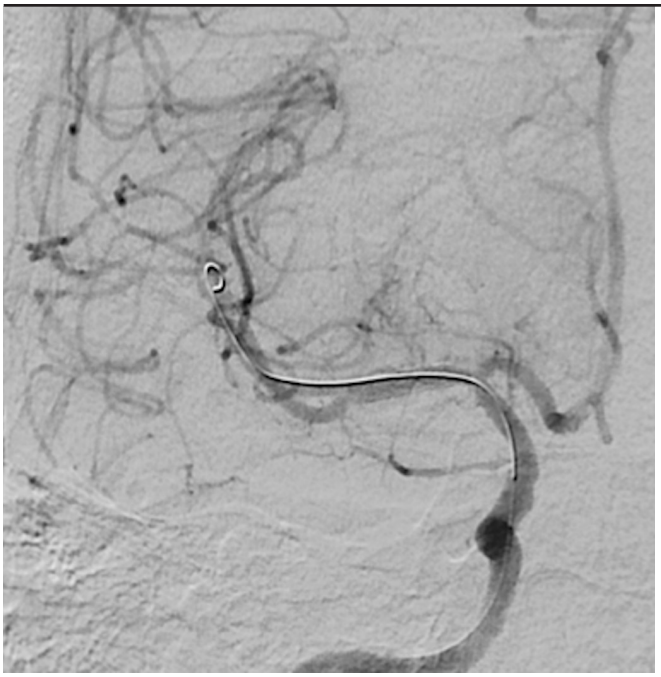

FIGURE 3A: Digital subtraction angiogram shows stent deployed.

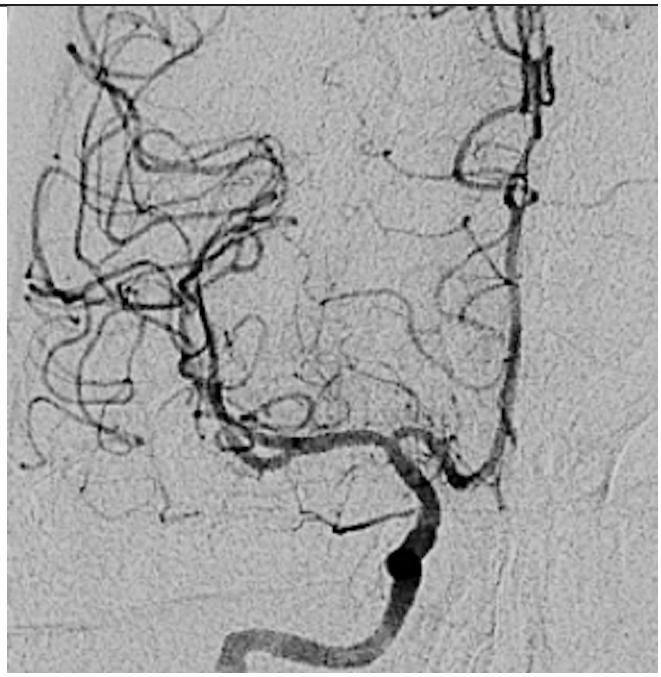

FIGURE 3B: Digital subtraction angiogram shows complete recanalization post stent placement.

to keep an eye out for ischemic stroke due to spontaneous intracranial artery dissection in Turner syndrome and manage it appropriately.

FINANCIAL SPONSORSHIP

None

DISCLOSURE

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

REFERENCES

1. Gravholt CH. Clinical practice in Turner syndrome. *Nat Clin Pract Endocrinol Metab* 2005;1(1):41–52.
2. Lancman M, Mesropian H, Serra P, Granillo R. Turner's syndrome, fibromuscular dysplasia, and stroke. *Stroke* 1991;22(2):269–71.
3. Komori H, Matsuishi T, Abe T, et al. Turner syndrome and occlusion of the internal carotid artery. *J Child Neurol* 1993;8(4):412–5.
4. Irioka T, Mizusawa H. Ischemic stroke in a young adult with Turner syndrome. *Neurol Sci* 2011;32(2):317–9.
5. Spengos K, Kosmaidou-Aravidou Z, Tsivgoulis G, et al. Moyamoya syndrome in a Caucasian woman with Turner's syndrome. *Eur J Neurol* 2006;13(10):e7-8.
6. Yoon CW, Lee E, Yoon B-N, Park H-K, Rha J-H. A Case of Turner Syndrome with Multiple Embolic Infarcts. *Case Rep Neurol* 2016;8(3):199–203.
7. Muscat P, Lidov M, Nahar T, Tuhrim S, Weinberger J. Vertebral artery dissection in Turner's syndrome: diagnosis by magnetic resonance imaging. *J Neuroimaging* 2001;11(1):50–4.
8. Fuentes K, Silveira DC, Papamitsakis NIH. Spontaneous carotid artery dissection in a patient with Turner syndrome. *Cerebrovasc Dis* 2007;24(6):543–4.
9. Laskay NMB, Estevez-Ordonez D, Atchley TJ, et al. Report of Spontaneous Internal Carotid Dissection in a Patient with Turner Syndrome with a Systematic Review of the Literature. *World Neurosurgery* 2019;128:340–6.
10. Gittenberger-de Groot AC, Azhar M, Molin DGM. Transforming growth factor β-SMAD2 signaling and aortic arch development. *Trends Cardiovasc Med* 2006;16(1):1–6.
11. Kostich ND, Opitz JM. Ullrich-Turner syndrome associated with cystic medial necrosis of the aorta and great vessels: case report and review of the literature. *Am J Med* 1965;38:943–50.
12. Price WH, Wilson J. Dissection of the aorta in Turner's syndrome. *J Med Genet* 1983;20(1):61–3.