Abstract

A 59-year-old Chinese patient was diagnosed with fibromuscular dysplasia (FMD) which is a rare genetic disorder among Asians. Neck vessels angiography showed a classic “string of beads appearance” affecting vertebral and internal carotid arteries. Carotid artery surgical reconstruction and vertebral intervention were proposed by vascular surgeon to prevent future strokes, but patient’s family refused for intervention/surgery. However, on optimal medical management his health improved during follow-up of over 8 months. This case shows unique multivessel FMD involving bilateral vertebral and one internal carotid artery which was managed successfully with medical management.

Key Words: Active intracranial hemorrhage with a midline shift, fibromuscular dysplasia, stroke in evolution

Introduction

Fibromuscular dysplasia (FMD) is an uncommon non-atheromatous, non-inflammatory arteriopathy of unknown origin affecting mostly the renal arteries (70-75%) in middle aged population having a “string of beads” appearance. Besides the renal arteries, the mid-cervical segment of the internal carotid artery (ICA) is the next most frequently affected region (25-30%) and 8-12% of patients with carotid involvement have associated vertebral artery involvement. This case presents a rare multivessel involvement of vertebral and carotid arteries.

Case Report

A 59-year-old male with one year history of poorly controlled hypertension (158/106 mmHg on presentation in the emergency department) presented with left side hemiparesis with headache, vertigo, and a Glasgow Coma Score of 15 points. Brain computed tomography (CT) revealed infarction of the right occipital lobe. Aspirin was started promptly. Three days later radiology disclosed infarction on the right thalamus (stroke-in-evolution) [Figure 1a]. Low molecular weight heparin was given for 3 days. Two weeks later, he had a suddenly unbearable headache, and his vision became blurred. His hemiparesis deteriorated. Brain CT showed postinfarction hemorrhage involving the thalamus and breaking into the lateral ventricle with a midline shift of 8.7 mm to the left side compressing the right optic nerve [Figure 1b]. Cranial nerve examination showed anisocoria (diameter of pupils; right: 2.5 mm, left: 4 mm). Digital subtraction angiography revealed a classic “beaded appearance” with multiple constrictions of the lumen affecting vertebral arteries and left internal carotid artery (ICA) [Figures 1c and 2]. He was diagnosed with fibromuscular dysplasia (FMD). He was discharged on day 21 without any neurologic sequelae, and aspirin (81 mg daily) was prescribed for primary stroke prevention.

Discussion

FMD, a genetic disorder, is rare among the Asian population.[1] This paper reports the first case from China. Less than 10% of FMD patients have problems with vertebral arteries.[1] The association of segmental FMD involving ICA, FMD affecting bilateral vertebral arteries causing subarachnoid hemorrhage is rare in medical literature. Research shows that alpha 1 antitrypsin deficiency is associated with arterial FMD.[2] However, despite having FMD our patient’s lung and liver functions were normal. Bleeding could be due to (a) posterior cerebellar artery dissection, (b) recanalization of the artery, or (c) rupture of a very small aneurysm.[3] Although
no rupture of the aneurysm was detected, we take this
eventuality under consideration. The neurosurgeon advised
draining the hematoma. The vascular surgeon suggested
surgical correction by end-to-end anastomosis or carotid,
and vertebral endovascular stenting. These measures were
expected to provide excellent long-term patency and help
prevent strokes in the future. However, such interventions
should ideally be carried out by high volume surgeons
as they have a higher success rate.\[^4\] Surgical approach
has only been adopted for symptomatic lesions.\[^5\] Artery
grafting has been advocated as the best treatment by
Van Damme \textit{et al.}\[^6\] His patient neither suffered from
postoperative mortality nor stroke after the artery
grafting. In the case of this patient, his family refused
surgical procedures. The patient was treated with calcium
channel blocker for appropriate hypertension control and
antiplatelets for stroke prevention. Interestingly, during the
8 months of follow-up, he reported improved health and
was asymptomatic. As a precautionary measure, the family
members of FMD patients should be screened in their 50
s (mean age 51.9 years) for arteries supplying the head and
neck, heart, kidneys, and pelvis to prevent catastrophic
complications because FMD is autosomal dominant.\[^5,6\]
This case demonstrates that such patients may have a
favorable prognosis even without any surgical procedure,
but the question is for how long. However, FMD should be
considered as a differential diagnosis for stroke patients.

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There are no conflicts of interest.

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