Giant internal carotid artery aneurysms and porcelain aorta in an elderly patient with Marfan syndrome

Hiroki Yagi1, NORIFUMI TAKEDA1, Yumiko Hosoya1, Haruo Yamauchi1, and Issei Komuro1

1The University of Tokyo Hospital

April 22, 2022

Abstract

Marfan syndrome (MFS) an inherited disorder caused by FBN1 gene mutations, is well known to cause lethal aortic aneurysm and dissections at a relatively young age. Here, we report giant internal carotid artery aneurysms and porcelain aorta in an elderly patient with MFS.

Category of manuscript: Clinical Images

Title: Giant internal carotid artery aneurysms and porcelain aorta in an elderly patient with Marfan syndrome

Hiroki Yagi, MD, PhD 1),2),#, Norifumi Takeda, MD, PhD 1),2), Yumiko Hosoya, MD, PhD 3)
Haruo Yamauchi, MD, PhD 2),4), Issei Komuro, MD, PhD1)

1)Department of Cardiovascular Medicine, 2)Marfan Syndrome Center, 3)Department of Therapeutic Strategy for Heart Failure, 4)Department of Cardiac Surgery, The University of Tokyo Hospital, 7-3-1 Hongo, Bunkyo-ku, Tokyo 113-8655, Japan

#Address for corresponding author: Hiroki Yagi, MD, PhD Department of Cardiovascular Medicine
Graduate School of Medicine
The University of Tokyo 7-3-1 Hongo, Bunkyo-ku, Tokyo 113-8655, Japan Phone: +81-3-3815-5411 Email: hiroki_yagi_19830414@yahoo.co.jp

199 words, 1 reference and 2 figures are included in this manuscript.

Key words: Marfan syndrome, internal carotid artery aneurysm, porcelain aorta, elderly patient

Patient is a 79-year-old obese woman with a BMI of 31.6, complicated with lifestyle-related diseases. At age 61, CT scan identified chronic thoracoabdominal aortic dissection (TAAD) involving multiple visceral and cervical arteries and with severe calcification (porcelain aorta) (Figure1A). At age 76, asymptomatic and marked enlarged bilateral internal carotid artery aneurysms (ICAAs) with tortuosity were incidentally depicted (Figure1B). The maximum right and left diameters were 2.2 cm and 5.6 cm, respectively. Surgical treatment options were considered, however conservative management was chosen because of her greater risk of operation. In April 2017, she was referred to our hospital and was diagnosed with Marfan syndrome (MFS) based on the presence of ectopia lentis and TAAD and a previously-reported FBN1 pathogenic variant (c.1709G>C; p.Cys570Ser). MFS is an autosomal dominant connective tissue disorder with genetic predisposition to aortic aneurysms and dissections at a relatively young age1). Recent advances in the medical and
surgical management have improved life expectancy. ICAAs and porcelain aorta had rarely been reported in MFS, however as the number of elderly patients increases, these unfamiliar late arterial complications might be kept in mind to develop more comprehensive management, because MFS arteries are considered to be easily influenced by lifestyle-related diseases.

Acknowledgments

None

Conflict of interest

The authors declare no conflict of interest.

Ethical Approval

The genetic analysis was approved by the University of Tokyo Hospital ethics committee (G-1538) and this case report was conducted in accordance with Declaration of Helsinki.

Consent

Written informed consent was obtained from the patient to use the data and pictures and publish this report in accordance with journal’s patient consent policy.

Author contributions

Hiroki Yagi (HY), Norifumi Takeda (NT), Yumiko Hosoya (YH), and Haruo Yamauchi (HY) were directly involved in management of the case. Issei Komuro revised the manuscript critically for important intellectual content. All authors approved the content of the manuscript and confirmed the accuracy or integrity of any part of the work.

Data Availability

The data that support the findings of this study are available on request from the corresponding author.

References


Figure Legend

Figure1

Three-dimensional CT angiography showing chronic thoracoabdominal aortic dissection involving multiple visceral and cervical arteries and with severe calcification (porcelain aorta) (A) and bilateral internal carotid artery aneurysms (B).