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AORTIC DISEASE

CASE REPORT: CLINICAL CASE

Segmental Arterial Mediolysis in a Patient With Ascending Aorta Dilatation

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ABSTRACT

A 70-year-old woman consulted us for dyspnea. Echocardiography revealed moderate aortic regurgitation secondary to ascending aorta dilatation. Study was completed with aortic angiography computed tomography, showing stenosis and dissection of several visceral arteries. There were no abnormal inflammatory or autoimmune markers, nor fluorine-18-fluorodeoxyglucose positron emission tomography uptake. Segmental arterial mediolysis was diagnosed.
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HISTORY OF PRESENTATION

A 70-year-old woman consulted us for dyspnea (NYHA functional class II). She was hemodynamically stable (blood pressure: 150/94 mm Hg, heart rate: 65 beats/min, saturated O_2: 98%). Physical examination revealed an early diastolic decrescendo murmur without signs of congestion. Electrocardiogram demonstrated atrial fibrillation with a controlled heart rate.

LEARNING OBJECTIVES

- To identify segmental arterial mediolysis, a rare nonatherosclerotic, noninflammatory, arteriopathic disease that is characterized by medial lysis of arterial walls.
- To understand the characteristic findings of segmental arterial mediolysis on CT images.
- To be able to make a differential diagnosis between segmental arterial mediolysis and other inflammatory vasculitis or atherosclerotic conditions.

PAST MEDICAL HISTORY

She was diagnosed with hypertension (and put on losartan 100 mg) 12 years ago.

INVESTIGATIONS

Transthoracic echocardiography showed a tricuspid aortic valve with a moderate regurgitation (vena contracta width: 0.5 cm, regurgitant fraction: 35%, effective regurgitation orifice area: 0.22 cm^2, pressure half time: 300 milliseconds) secondary to aortic dilatation (annulus: 23 mm, sinus: 36 mm, sinotubular junction: 33 mm, proximal ascending aorta: 47 mm). The left ventricle was mildly dilated (interventricular septum: 10 mm, left ventricle end-diastolic diameter: 55 mm) presenting a normal ejection fraction (left ventricular ejection fraction: 62%) (**Figure 1A**).

An aortic computerized tomographic (CT) angiography was performed to accurately measure aortic

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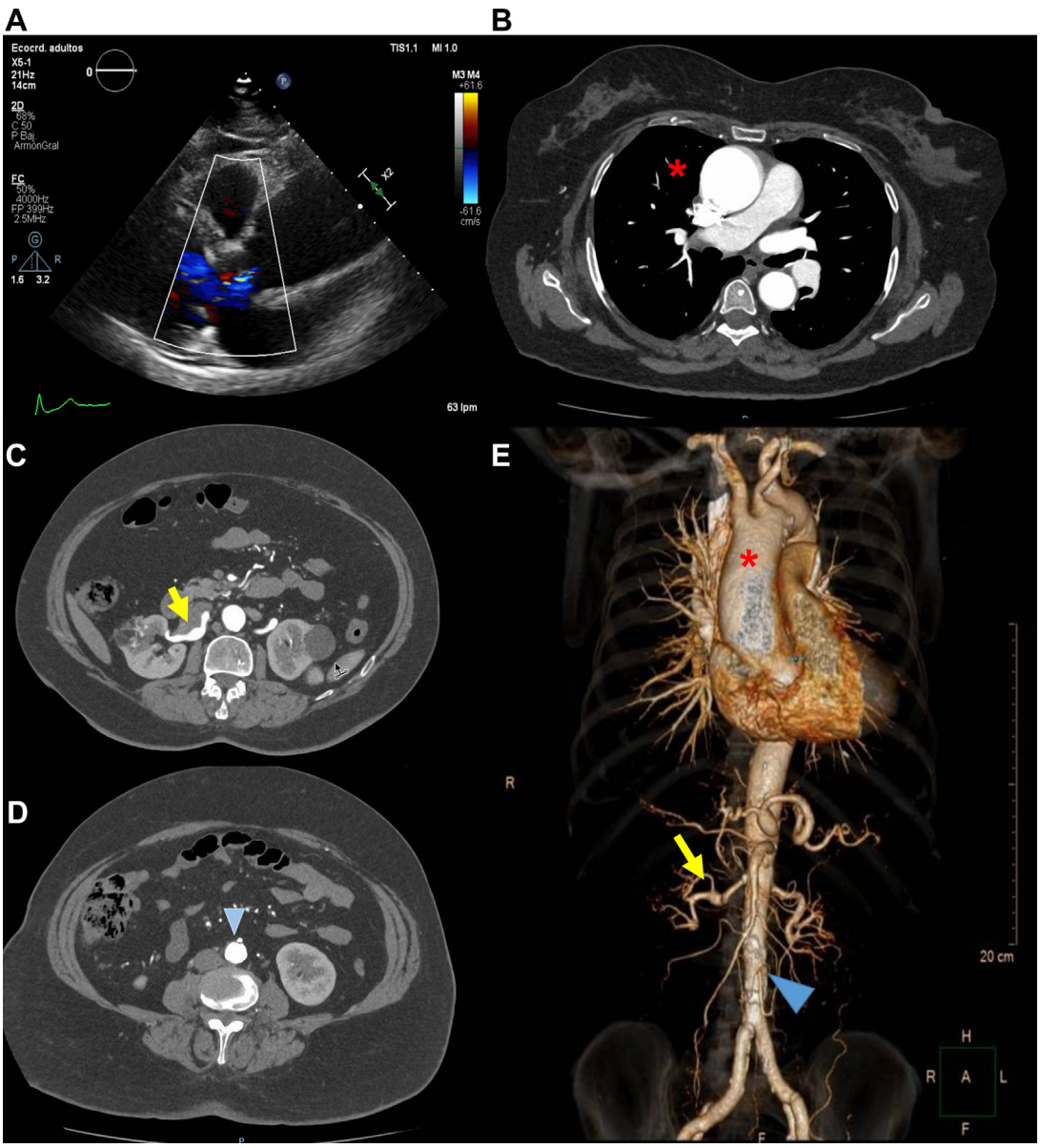
ABBREVIATIONS AND ACRONYMS

CT = computerized tomography

diameters. Ascending aorta aneurism was diagnosed (maximal diameter: 47 mm). Aortic arch, descending aorta, and abdominal aorta were nondilated. CT showed a fusiform dilatation of celiac trunk (14 mm), superior mesenteric artery (11 mm), and right renal artery (10 mm). The distal region of the superior mesenteric artery showed a stenotic lesion. The inferior mesenteric artery was dissected at the origin (Figures 1B to 1E).

FIGURE 1 Segmental Arterial Mediolysis



(A) Transthoracic echocardiography. Parasternal long-axis view. Doppler color. Moderate aortic regurgitation secondary to aortic aneurism. (B) Computerized tomography, arterial contrast phase. Axial view. The asterisk indicates the ascending aorta aneurism. (C) Computerized tomography, arterial contrast phase. Axial view. Arrow shows the fusiform aneurism of the right renal artery. (D) Computerized tomography, arterial contrast phase. Axial view. Arrowhead marks the dissection of the inferior mesenteric artery. (E) Computerized tomography, multiplanar reconstruction. The asterisk indicates the ascending aorta aneurism. The arrow shows the fusiform aneurism of the right renal artery. Arrowhead marks the dissection of the inferior mesenteric artery.

There was no abnormal elevation of inflammatory markers (C-reactive protein, leukocytes, nor fibrinogen). Significant atherosclerosis was absent in the vasculature. Blood cultures and autoimmune panel resulted negative. Fluorine-18-fluorodeoxyglucose positron emission tomography showed no abnormal uptakes. A genetic panel (*ACTA2, FBN1, SMAD3, TGFBR1-2, COL3A1/5A2*) identified no pathogenic mutations. Segmental arterial mediolysis was diagnosed according to CT findings. Multifocal fibromuscular dysplasia was ruled out, because neither stenotic lesions nor a typical "string of beads" pattern were present. Histologic confirmation was not obtained because the patient refused invasive biopsy.

MANAGEMENT

Antihypertensive treatment was intensified. Although thromboembolic risk was high (CHA_2DS_2-VASc score = 3), hemorrhagic risk was of concern due to high risk of intra-abdominal hemorrhage. Left appendage closure was thus decided on, successfully implanting a 21-mm plug device.

DISCUSSION

Segmental arterial mediolysis is a rare non-atherosclerotic, noninflammatory, nonhereditary arteriopathic disease that is characterized by medial lysis of arterial walls.[1] The term "mediolysis" refers to the disruption of the smooth muscle cell membrane leading to arterial dilatation. Middle-age and elderly populations are predominantly affected. It most commonly involves large abdominal aortic branches, especially the coeliac axis/splenic arteries. The most frequent presentation is acute abdominal pain, secondary to arterial rupture and hemorrhage.[1] CT findings include aneurysms and stenosis in a segmental pattern, as well as arterial dissections. Differential diagnosis with vasculitis, fibromuscular dysplasia, or hereditary vascular diseases must be performed.[2] Its natural history is unpredictable: self-resolution, progression, or fluctuation, moving from some segments to others have been described. Continued imaging surveillance is thus recommended. Blood pressure control is recommended in most patients. Surgery/endovascular treatment should be considered in acute dissection or rupture.[3]

FOLLOW-UP

Lifetime imaging follow-up was decided, with no changes at 6-month follow-up compared to previous imagery. To date, no bleeding complications have arisen.

CONCLUSIONS

Segmental arterial mediolysis is a rare disease that is characterized by arterial aneurisms, stenosis, or dissections, especially of the splanchnic arteries. Although it may be an incidental diagnosis, this entity is potentially lethal. CT findings enable the diagnosis after excluding other vascular diseases, such as vasculitis and other mimics.

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KEY WORDS aortic disease, computed tomography, mediolysis, vasculopathy