



Intravascular ultrasound findings of immunoglobulin G4-related periarteritis

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Immunoglobulin G4 (IgG4)-related disease is an autoimmune disease that affects multiple organ systems including the endocrine, urinary, and cardiovascular systems. We were able to diagnose IgG4-related disease using intravascular ultrasound (IVUS).

A 77-year-old man underwent emergency invasive coronary angiography (CAG) for unstable angina. CAG showed severe stenosis in the mid left circumflex artery (LCX) (Fig. 1a) and the proximal right coronary artery (RCA) (Fig. 1b), and total occlusion of the proximal left anterior descending artery. IVUS findings showed low echoic regions not only on the inner side of the media but also on the outer side of the media, corresponding to stenosis in the mid LCX (Fig. 1a, A–D), and the proximal RCA (Fig. 1b, E–H). Drug-eluting stents were successfully deployed in the mid LCX and the proximal RCA. We suspected IgG4-related

disease from IVUS findings and performed further tests. The patient's serum IgG4 level was elevated at 774 mg/dL. We performed immunostaining with IgG on lymph node biopsy specimens obtained earlier. It revealed IgG4-positive cells > 200/high-power field and the ratio of IgG4 was > 50%.

Near-infrared spectroscopy intravascular ultrasound accurately can identify lipid core plaques. Stenotic lesions had two types of lesion, composed of atherosclerotic and non-atherosclerotic parts. We consider the low echoic regions without lipid component are plasma cell infiltration on the inner and outer sides of the media with previous histopathological report [1]. While IgG4-related disease is not rare disease, IgG4-related cardiovascular disorders are uncommon. Cases of sudden cardiac death in patients with coronary artery lesions caused by IgG4-related disease have

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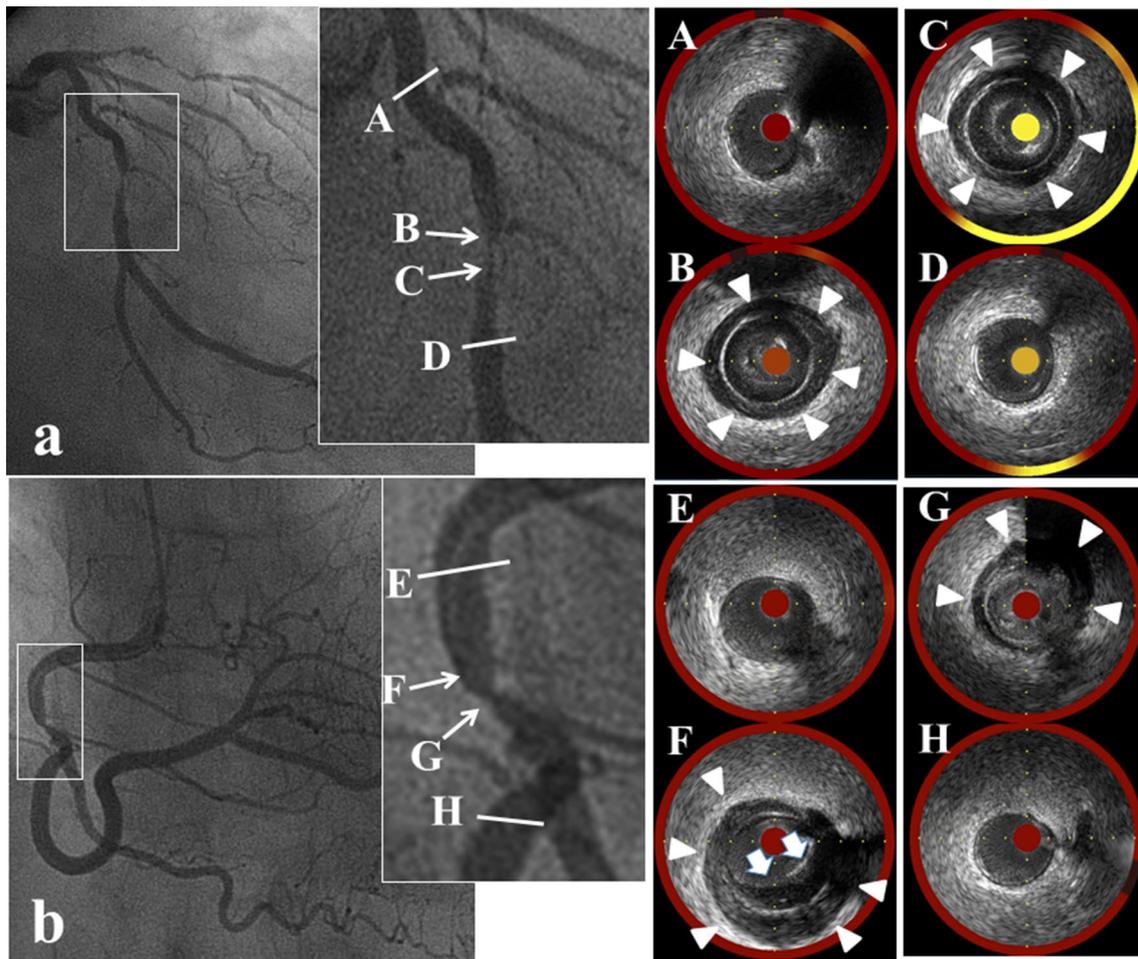


Fig. 1 **a** Coronary angiography (CAG) of the left coronary artery—CAG showed severe stenosis in the mid left circumflex artery (LCX) and total occlusion in the proximal left anterior descending artery. **A–D** Near-infrared spectroscopy intravascular ultrasound (NIRS-IVUS) imaging of the LCX showed a low echoic region on the outer side of the media (arrowheads), corresponding to stenosis. The sten-

otic lesion consisted of fibrous and lipid components. **b** CAG of right coronary artery (RCA)—CAG showed severe stenosis in the mid RCA. **E–H** NIRS-IVUS imaging of the RCA also showed a low echoic region on the inner (arrows) and outer sides (arrowheads) of the media, corresponding to stenosis. The stenotic lesion consisted of tissue without lipid components

been reported [2], suggesting that the disorder may be fatal. Awareness of IgG4-related disease is essential because the disorder is treatable.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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