

## IMAGES IN INTERVENTION

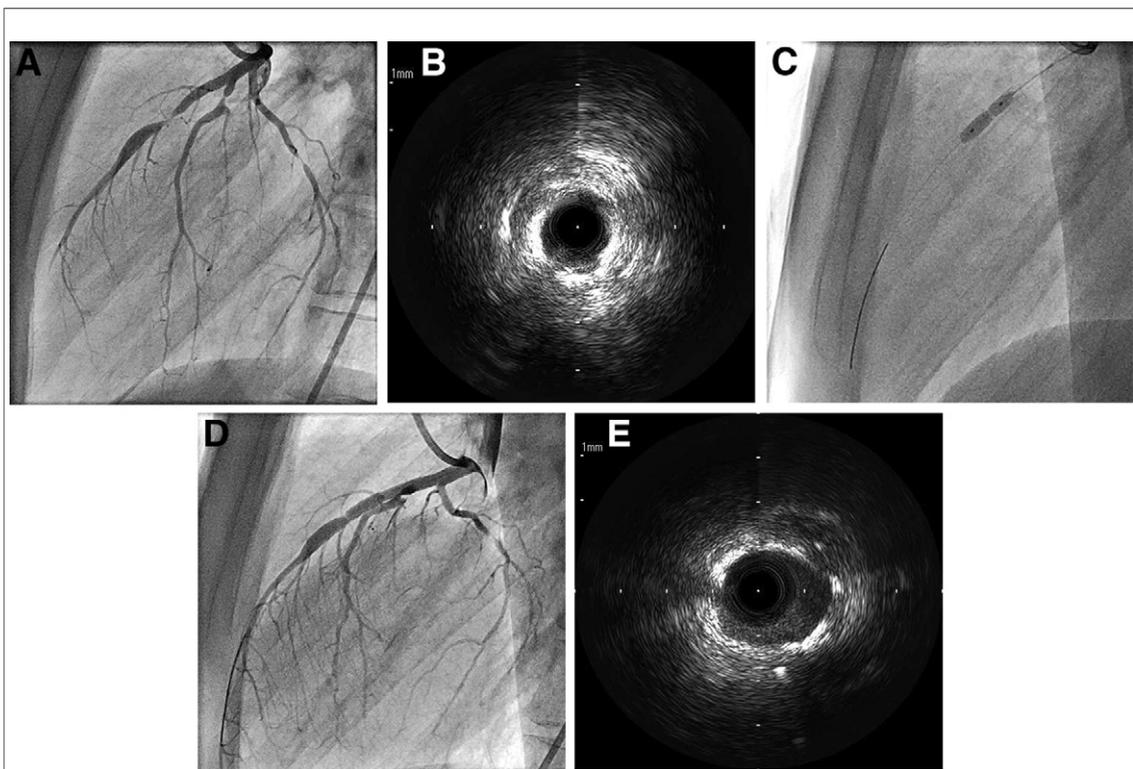
# Stiff Coronary Stenosis in a Young Female With Pseudoxanthoma Elasticum

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Pseudoxanthoma elasticum (PXE), which is caused by mutation in the *ABCC6* gene, is a heritable systemic disorder affecting elastic fibers, most

markedly in skin, retina, and blood vessels (1). To date, there is 1 case report that percutaneous coronary intervention (PCI) was performed in a



**Figure 1. Findings of the Left Anterior Descending Coronary Artery**

(A) Coronary angiogram revealed severe stenoses without visible calcification. (B) Intravascular ultrasound (IVUS) revealed a superficial high echoic component with several breaches around the vessel. (C) After post-dilation using a large noncompliant balloon (3.75-mm Quantum Maverick), optimum stent expansion could not be achieved. (D) Final angiography showed stent underexpansion of the lesion. (E) Final IVUS finding of the underexpanded lesion.

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patient with PXE (2). In that report, the stenosis was successfully treated by the conventional techniques. However, lesion pathology in PXE is reported to be different from atherosclerosis (3).

A 26-year-old Japanese woman was diagnosed with PXE. She had typical skin involvement (yellow cobblestone appearance of the neck and elastofibroma on skin histopathology) and eye involvement (angioid streak). She was referred to our cardiology division for examination of coronary artery disease. The multidetector computed tomography (MDCT) images revealed significant stenoses of the mid left anterior descending coronary artery (LAD) and the left circumflex coronary artery. However, no coronary calcification was visible. Coronary angiography revealed severe stenoses without visible calcification, consistent with the findings on MDCT (Fig. 1A).

Transfemoral PCI was tried to treat the LAD stenosis. Intravascular ultrasound (IVUS) (View It, Terumo, Tokyo) revealed a superficial high echoic component around vessels without acoustic shadowing (Fig. 1B). A 3.0 × 18-mm bare-metal stent (Driver, Medtronic, Minneapolis, Minnesota) was deployed (maximum inflation pressure of 16 atm). Post-dilations using several noncompliant balloons, such as the 3.75-mm Quantum Maverick (Boston Scientific Corporation, Natick, Massachusetts), were performed (maximum inflation pressure of 24 atm) (Fig. 1C). Unfortunately, the final angiogram showed stent underexpansion (Fig. 1D). Minimum stent area of the lesion was 4.4 cm<sup>2</sup> (Fig. 1E). We added bisoprolol for the treatment of other nondilated vessels.

Intimal calcification in the coronary artery is 1 of the characteristics of atherosclerosis. By contrast, intimal fibrosis in the coronary artery is an idiopathic change with PXE (3,4). Although there was no visible calcification on both MDCT and angiography, IVUS suggested the presence of dense intimal fibrosis. This dense fibrosis might impede the stent expansion. Our case suggests the necessity of taking a cautious approach in PCI of lesions in patients with PXE, because the presence of dense fibrosis, which is invisible by angiogram or MDCT, may inhibit the vessel expansion. Cautious and aggressive pre-dilation before stenting may be needed to prevent stent underexpansion.

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