Visualization of Complete Regression of Pulmonary Arterial Remodeling on Optical Coherence Tomography in a Patient With Pulmonary Arterial Hypertension

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Figure 1. Serial optical coherence tomography (OCT) of the pulmonary artery during medical treatment. (A) The first OCT examination (12 August 2009) showed remodeling of the right anterior basal segmental artery, approximately 3.5 mm in diameter, which is consistent with OCT findings of intimal fibrosis (mean outer diameter, 3.45 mm; outer area, 9.35 mm²; wall-area ratio, 0.302; thickness-diameter ratio, 0.081; and thickness, 0.28 mm). Serial OCT showed that the medical treatment induced (B) progressive improvement of the pulmonary arterial remodeling on 25 November 2009 in the right middle lobe lateral segmental artery (mean outer diameter, 3.42 mm; outer area, 9.24 mm²; wall-area ratio, 0.247; thickness-diameter ratio, 0.067; and thickness, 0.23 mm), (C) complete regression on 13 August 2010 in the right lower lobe apical segmental artery (mean outer diameter, 3.43 mm; outer area, 9.27 mm²; wall-area ratio, 0.202; thickness-diameter ratio, 0.052; and thickness, 0.18 mm), and (D) sustained complete regression on 18 February 2011 in the right pulmonary artery with an undocumented specific location (mean outer diameter, 3.43 mm; outer area, 9.24 mm²; wall-area ratio, 0.187; thickness-diameter ratio, 0.049; and thickness, 0.17 mm). (Inset) High-power image of section of pulmonary arterial wall (dashed-line box). Arrows, wall thickness.
OCT (LightLab Imaging, Westford, MA, USA) showed remodeling with intimal fibrosis but no obstruction of the pulmonary arteries (Figure 1A). We thus diagnosed pulmonary arterial hypertension (PAH) associated with connective tissue disease according to the current guidelines, and prescribed oral beraprost (120 μg daily) in addition to oral prednisolone (5 mg daily). After initiation of corticosteroid pulse therapy combined with intensive immunosuppressive therapy with cyclophosphamide, she was re-evaluated in November 2009, with PAP 32/18/24 mmHg and improved pulmonary arterial remodeling on OCT (Figure 1B). After the corticosteroid pulse therapy, the dose of oral prednisolone was maintained at 10 mg daily. Sildenafil (60 mg daily) was then added to her prescription. On her next follow-up in August 2010, PAP was further improved to a normal range (30/12/19 mmHg), together with further improvement of pulmonary arterial remodeling as evidenced on OCT (Figure 1C). Six-minute walk test was also improved to 503 m with slight shortness of breath. On the patient’s next visit in February 2011, PAP remained within the normal range (30/12/20 mmHg), associated with normalization of pulmonary remodeling on OCT (Figure 1D). Since then, we have not performed OCT, given that the patient has remained...
asymptomatic at regular follow-up.

OCT images were quantified at the same levels with similar outer diameters and outer areas, using 3 morphometric parameters described in a previous study, including pulmonary arterial wall thickness, thickness-diameter ratio (defined as thickness divided by outer diameter) and wall-area ratio (defined as the wall area in the cross-section divided by the sum of area of both lumen and wall). These 3 structural parameters of the pulmonary arteries were progressively improved along with improvement of pulmonary hemodynamics during medical treatment (Figure 2). The lesion detected on OCT was similar to OCT findings of intimal fibrosis confirmed on histopathology in PAH in a previous report, but there is not sufficient evidence to completely deny other types of lesion in the present case, and further studies regarding comparison between OCT findings and histopathology are required. In the present study, the four OCT examinations were conducted at different locations in the pulmonary artery tree. Given that the pulmonary arterial lesions may not be homogenous with regard to location, there is a possibility that the reverse remodeling observed in the present study was only a coincidence, caused by the variation in location.

We have previously demonstrated the usefulness of OCT in the diagnosis of pulmonary hypertension. In the present case we were able to demonstrate, with serial OCT observations, that pulmonary arterial remodeling in PAH could completely regress in response to adequate medical therapy, indicating another aspect of usefulness of OCT in the management of the disorder.

Disclosures
None.

References