A variation of vasculopathies in a patient with mild pulmonary artery hypertension

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DESCRIPTION

A 66-year-old woman with a 2-year history of rheumatoid arthritis (RA) that was treated with prednisolone was referred to our hospital due to a slight shortness of breath with normal vasculatures (figure 1A,B) and a small coin lesion (figure 1C,D). An estimated pulmonary artery pressure on echocardiography (figure 1E) of 44 mm Hg that increased to 65 mm Hg after 6 months. The mean pulmonary arterial pressure (PAP) was 37 mm Hg (figure 2) without a shuntvitium, the mean pulmonary capillary wedged pressure was 10 mm Hg, and the calculated pulmonary vascular resistance (PVR) was increased to 376 dyn/(s/cm^5). Because perfusion scintigraphy showed no abnormalities without clinical signs of systemic lupus erythematous, mixed connective tissue disease or systemic scleroderma, we diagnosed her as idiopathic pulmonary artery hypertension (IPAH). Administration of bosentan (125 mg) for 1 month improved her symptom and decreased the PAP and PVR to 27 mm Hg and 290 dyn/(s/cm^5), respectively. Because the coin lesion increased in size, right upper lobe was resected 2 months later (figure 3A), which revealed a granulomatous lesion with *Cryptococcus* spp. Although normal pulmonary arteries were also observed, medial hypertrophy, intimal thickening (figure 3B,C), microthrombosis (arrowheads; figure 3B,D), and few plexiform lesions (figure 3E) were observed in the same field, indicating that a variation was observed in a patient with mild PAH,[1] as well as severe PAH in autopsy or explantation cases.[2]

Since there were few reports[3] and clinical signs, we could not conclude whether the RA caused PAH, a very rare case or IPAH was coincidentally observed in a patient with RA.

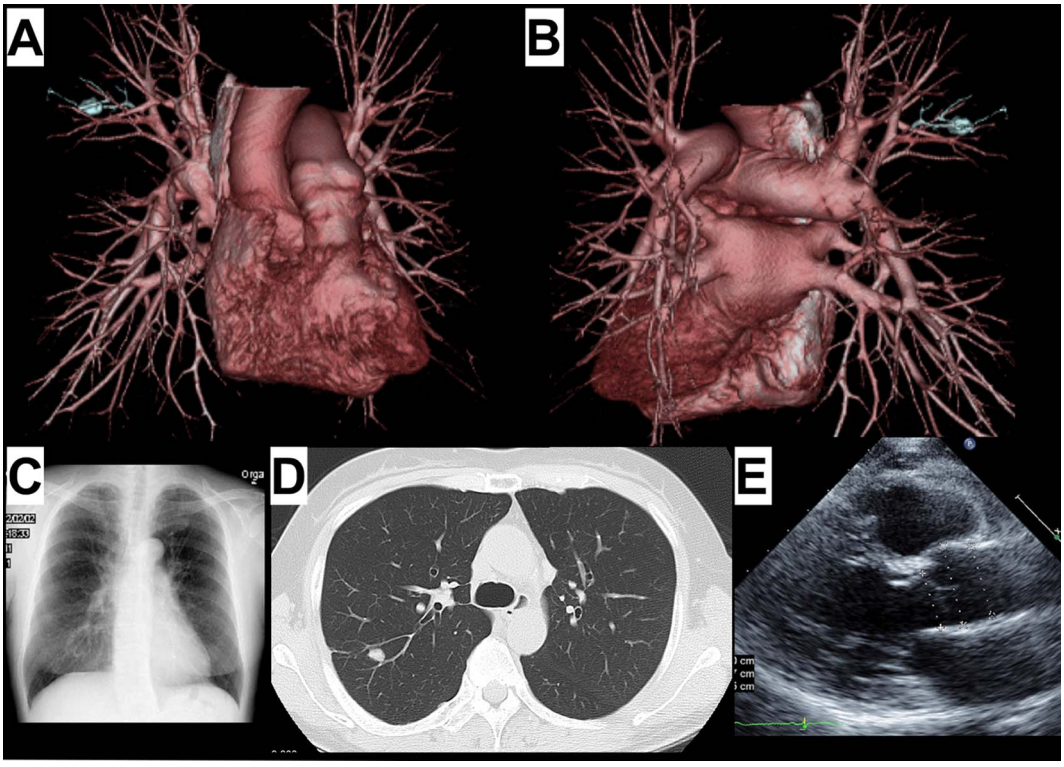

Figure 1 (A and B) Three dimensional reconstraction image of contrast-enhanced CT angiography. Anterior (A). Posterior (B). (C) A chest X-ray at admission. (D) Coin lesion in CT scans. (E) Echocardiography.

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Right heart catheterization

Cardiac output(L/min)				4.45	Cardiac index (L/min/m2)				2.77			
Oxymetry		SVC	71.30%	IVC	63.00%	RV	66.10%	mPA	66.70%	Aorta	97.90%	
Qp	4.1	Qs	4.4	Qp/Qs	0.9							

Echocardiography (transthoracic)

LVDd (mm)		37	LVDs (mm)		25	EF	64%	IVSTd (mm)		7	PWTd (mm)	7
LAD (mm)		28	LAVI (ml/m2)		16	E (m/sec)		0.72	A (m/sec)		0.75 E/A	0.96
TRPG	57mmHg		RVOT Act/ET		0.23	TAPSE(mm)		20.8	Tei index (RV)		0.38	
Shunt	none											

Laboratory findings

Anti-CCP-Ab	123	RF	55	ANA	40 (speckled)		U1-RNP-Ab	negative	
Anti-scl-Ab	negative	Anti-Centromere-Ab			negative				
BNP(pg/ml)	55								

Physical activity

NYHA stage	Ⅱ	6MWD(m)	375m	SpO2	95%

Respiratory function test

%VC	122.70%	FEV1.0%	74.90%	%DLCO	94.10%

SVC; superior vena cava, IVC; inferior vena cava, RV; right ventricle, mPA; main pulmonary artery, Qp; pulmonary flow, Qs; systemic flow, LVDd diastolic left ventricular dimension, LVDs; systolic LV dimension, EF; ejection fraction, IVSTd; diastolic inetrventricular septal thickness, PWTd; diastolic posterior wall thickness, LAD; left atrium diameter, LAVI; left atrium volume index, TRPG; tricuspid regurgitation pressure gradient, RVOT Act/ET; right ventricular outlet tract acceleration time/ ejection time, TAPSE; Tricuspid Annular Plane Systolic Excursion, RV; right ventricle, CCP; citrullinated protein, RF; Rheumatic factor, ANA; antinuclear antigen, .BNP; brain natriuretic peptide, NYHA; New York Heart Association, 6MWD; 6-minute walk distance, SpO2; blood oxygen saturation, VC; vital capacity, FEV1.0; expiratory volume after 1.0 second, DLCO; diffusing capacity for carbon monoxide.

Figure 2 Details of right heart catheterisation, laboratory findings, physical activity and respiratory function testing.

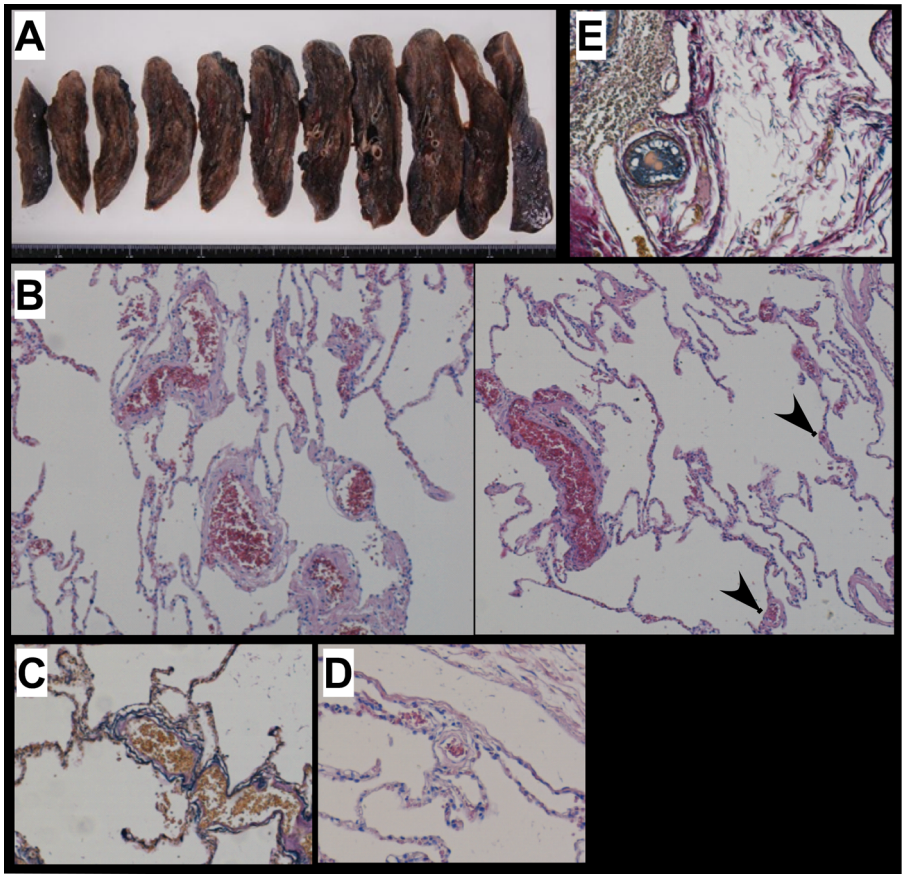

Figure 3 (A) Macroscopic examination of resected lung. (B) Medial hypertrophy, intimal thickening and microthrombosis (arrowheads) were observed by H&E stainings. In the same field, normal pulmonary arteries were also observed. (C) Medial hypertrophy by elastic-Van Gieson (EVG) staining. (D) Microthrombosis. (E) A plexiform lesion by EVG staining.

Learning points

- Pathological lesions in the pulmonary artery hypertension (PAH) are thought to belong to a similar spectrum in extension and distribution; however, a variation in the lesions was observed in a patient, even if it is very mild, as previously reported Wagenvoort, 1970.[4]
- Medial hypertrophy, intimal thickening, microthrombosis and a few plexiform lesions were observed and normal pulmonary arteries were also observed in the same field.
- Pulmonary hypertension with rheumatoid arthritis (RA) is rare, therefore we could not conclude whether the RA caused PAH, a very rare case, or idiopathic pulmonary artery hypertension was coincidentally observed in a RA patient.

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Competing interests None.

Patient consent Obtained.

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