Marked Dilatation of Pulmonary Arteries in Mixed Connective Tissue Disease

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A 59-year-old woman was diagnosed with mixed connective tissue disease 25 years ago, along with pulmonary hypertension (85/35 mm Hg) measured directly by right heart catheterization. Pulmonary arterial pressure was inferred from echocardiography. The systolic blood pressure of her pulmonary artery had been between 90 and 110 mm Hg since then, although it decreased slightly to between 70 and 90 mm Hg recently. She has been treated with digoxin, furosemide, and azathioprine since the initial diagnosis. She has been receiving home oxygen therapy for the past 4 years. Glucocorticoids were never administered. A marked enlargement of cardiac silhouette and dilatation of pulmonary arteries were seen in her recent chest radiograph (Figure 1), compared with one taken 25 years ago (Figure 2). Despite such remarkable radiographic changes, brain natriuretic peptide (BNP) in her plasma was relatively low at 87.5 pg/ml (normal, < 18.4 pg/ml). BNP is considered a prognostic indicator in primary pulmonary hypertension[1]. She remains well, with mild shortness of breath on moderate exertion, although she can walk, shop, and do housework with no difficulty. She is in functional class II proposed by the New York Heart Association. Similar radiographic changes were reported previously in a patient with primary pulmonary hypertension[2]. In both cases the reason for unusually long survival is unknown.

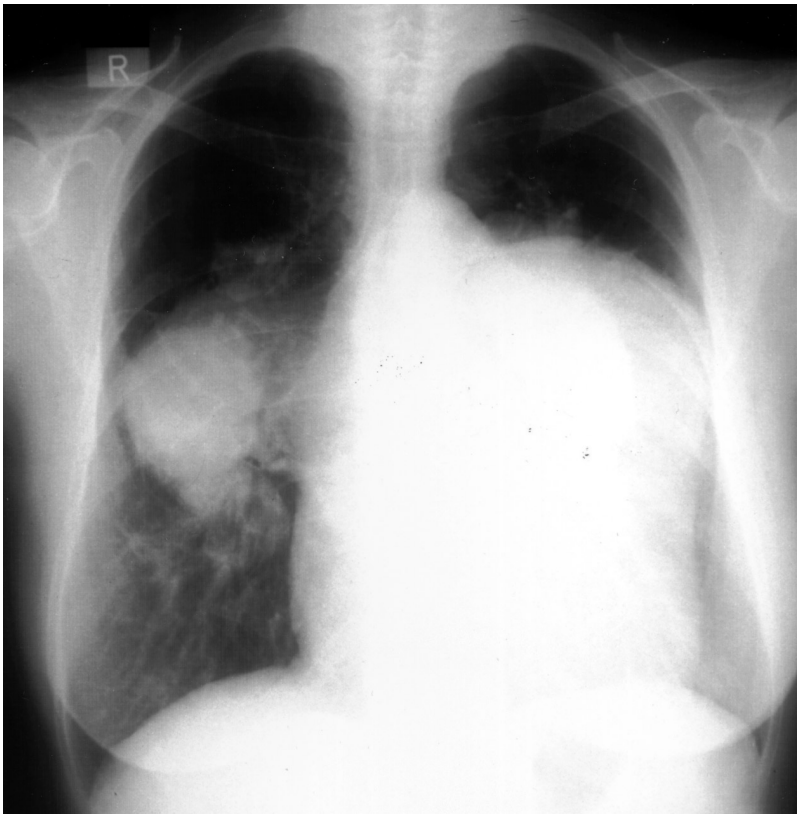

Figure 1. Recent chest radiograph shows marked enlargement of cardiac silhouette and dilatation of pulmonary arteries.

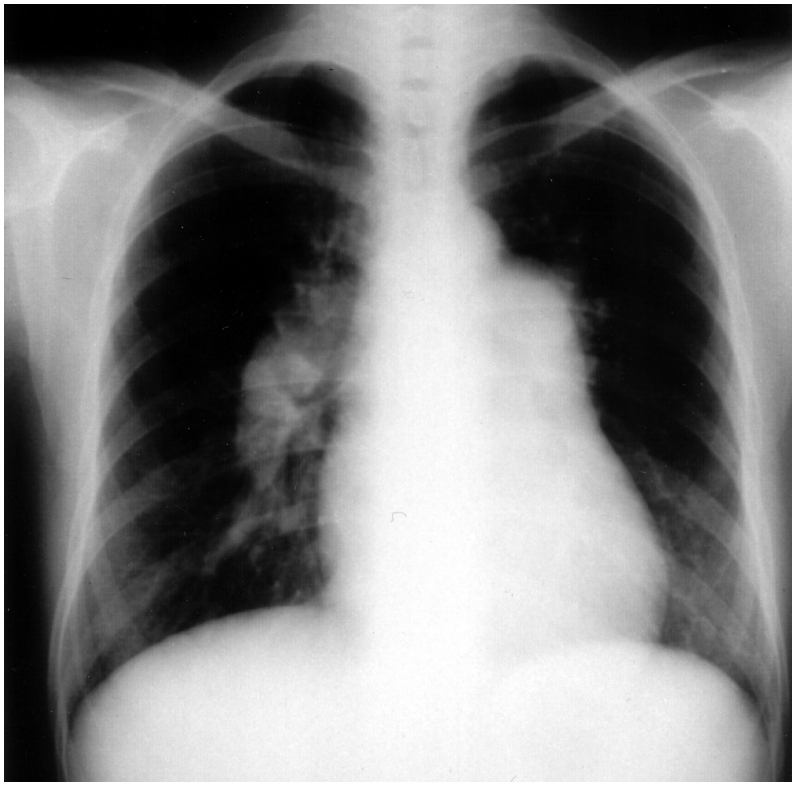

Figure 2. Chest radiograph at the diagnosis of pulmonary hypertension 25 years ago.

REFERENCES

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