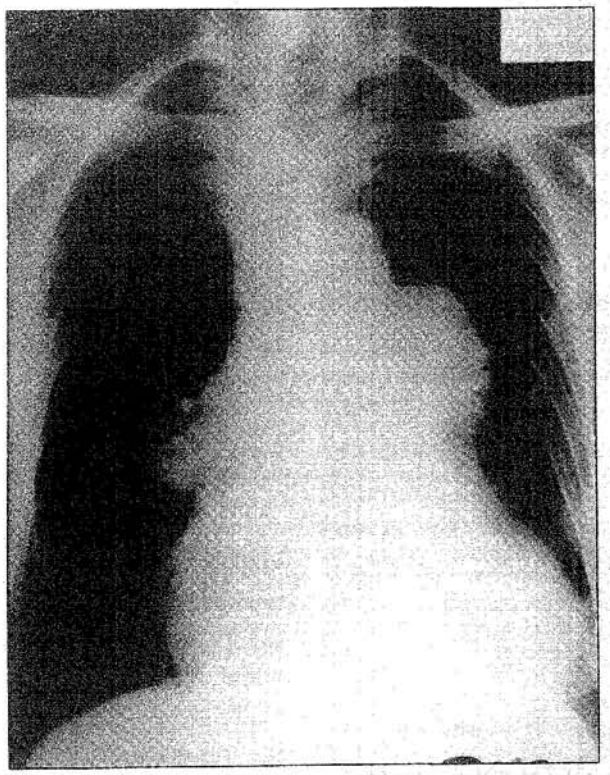
Clinical History:

An 81-year-old woman presented with a hip fracture following a fall. A pre-operative chest radiograph was performed (**Figure 1**). Comparison with previous chest radiographs performed over the past 10 years showed no significant change in appearances.

Figure 1: Posteroanterior chest radiograph



Answer on page 92

What is the diagnosis?

a) Sarcoidosis
b) Lymphoma
c) Tuberculosis
d) Silicosis
e) Pulmonary arterial hypertension

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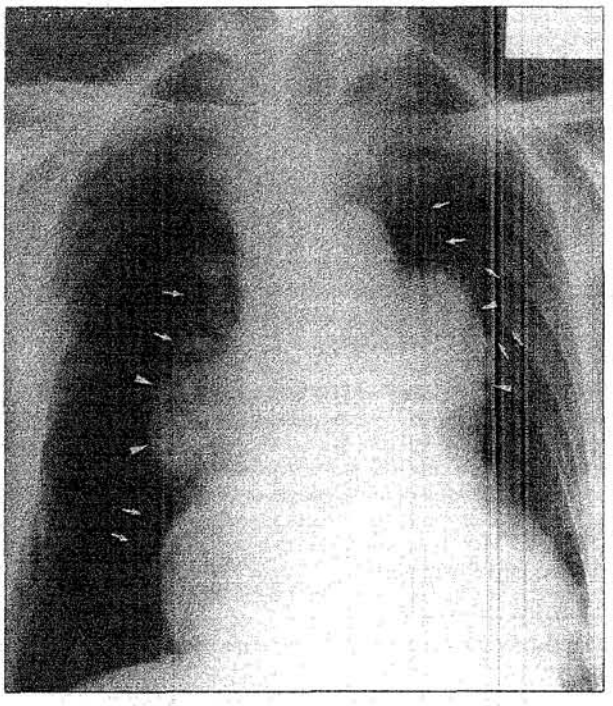
Answer:

e) Pulmonary arterial hypertension.

Radiological findings

The chest radiograph (**Figure 2**) shows an enlarged heart. The lung fields are hyperinflated with flattening of the hemidiaphragms, consistent with chronic obstructive airways disease (COAD). Prominent hilar shadows are due to dilated main and proximal pulmonary arteries, which taper rapidly distally with relative peripheral pulmonary oligaemia. The pulmonary arterial hypertension and cor pulmonale were secondary to underlying COAD.

Figure 2: **This figure is identical to Figure 1 with addition of arrows. The heart is grossly enlarged. The lung fields are hyperinflated, with the diaphragmatic domes located at the level of the posterior 12th ribs. Both the prominent hilar shadows (arrowheads) correspond in position to dilated proximal pulmonary arteries, to which the tapering distal vessels converge (small arrows)**



Discussion

Sarcoidosis

Sarcoidosis is a multisystem granulomatous disorder of unknown aetiology, with routine chest radiographic abnormalities being the presenting feature in 25% of cases. Other common presentations are erythema nodosum (30%), respiratory symptoms (20%), ocular symptoms (8%) and skin lesions (5%). Bilateral hilar lymphadenopathy is associated with "eggshell" calcification in long-standing disease, mediastinal lymph node enlargement and mid-zone parenchymal, commonly reticulo-nodular, opacities. The combination of right paratracheal and bilateral hilar lymphadenopathy has been described as a characteristic pattern of adenopathy in sarcoidosis, and has been termed the "one-two-three" sign. These clinical and radiographic features were not found in our patient.

Lymphoma

Intra-thoracic lymphadenopathy is present in the majority of patients with lymphoma. Anterior mediastinal, paratracheal and tracheobronchial nodal involvement occurs more frequently than does hilar nodal disease. The distribution of lymphadenopathy tends to be bilateral and asymmetrical. Extension into the lungs produces a mixed nodular and interstitial pattern of spread. This diagnosis can be excluded based on clinical and radiographic features.

Tuberculosis

Hilar lymph node enlargement may be found in many infective conditions such as tuberculosis. The hilar lymphadenopathy is, however, usually unilateral and is rarely bilateral and symmetrical. In primary tuberculosis, there is usually associated paratracheal lymph node enlargement and pulmonary consolidation of the upper or middle lobe. If the hilar lymph nodes are large enough, they may obstruct bronchi by external compression or by discharging caseous material into the lumen. None of these findings were present in our case.

(Continued on page 95)

RADIOLOGICAL CONFERENCE

Silicosis

Silicosis may occur in people exposed to free silica, for example, miners, quarry workers, pottery workers and sand blasters. The time of onset of disease depends upon the duration and degree of dust exposure. The silica causes a fibrotic reaction in the lung, which may progress even after cessation of exposure. The predominant chest radiographic abnormality is the presence of multiple small nodular opacities of fairly uniform size, which may be complicated by development of progressive massive fibrosis in the upper lobes. Hilar lymph node enlargement is often minor, and is usually obvious only when calcification occurs. This diagnosis can be excluded in our patient on the basis of lack of a relevant occupational history and the radiographic appearances.

Pulmonary arterial hypertension

Massive dilatation of the main and proximal pulmonary arteries can mimic bilateral hilar lymphadenopathy. Unlike the former condition, enlarged lymph nodes do not taper distally and instead produce distinct lumpy masses, likened to a "sack of potatoes". The "hilum convergence" sign is of value in differentiating hilar masses from vascular structures. If the pulmonary arterial branches converge towards the mass rather than towards the heart, this implies that the mass represents an enlarged pulmonary artery. On the other hand, convergence towards the heart rather than towards a hilar mass indicates that the mass is extravascular. In pulmonary arterial hypertension, besides proximal pulmonary arterial dilatation, "pruning" due to rapid distal tapering and peripheral vascular narrowing is typically seen. Pulmonary arterial calcification is rare but is considered characteristic, if present.

Pulmonary arterial hypertension is defined as mean pulmonary arterial pressure exceeding 20 mmHg (and when the systolic pressure is greater than 30 mmHg and the diastolic pressure is more than 15 mmHg). Pulmonary arterial hypertension can be classified into the rare primary form, which affects young women in an idiopathic manner, and the much commoner secondary form. The pathogenesis of the secondary form includes primary pleuropulmonary disease, primary vascular disease and pulmonary venous hypertension (see **Table 1**).

Table 1: Classification of pulmonary arterial hypertension

A. Primary (idiopathic)
B. Secondary
- i. lung parenchymal disease
  - e.g. COAD
    chronic bronchitis
    end-stage fibrotic disease
    cystic fibrosis
- ii. alveolar hypoventilation
  - e.g. sleep apnoea
    chronic high altitute
- iii. pleural disease
  - e.g. thoracoplasty
    kyphoscoliosis
- iv. primary vascular disease
  - e.g. congenital heart disease
    chronic pulmonary thromboembolism
    pulmonary veno-occlusive disease
- v. pulmonary venous hypertension

(Adapted from reference 1)

In our patient, the chest radiograph shows background features of COAD. Most of the other differential diagnoses can be excluded on basis of the long-standing static radiographic appearances. In COAD, pulmonary arterial hypertension is caused by a combination of hypoxaemia and destruction of the microvasculature, leading to obliteration of the small pulmonary arteries and arterioles. Persistently raised pulmonary artery pressure would eventually result in right ventricular hypertrophy and cor pulmonale. ■

References

1. Dähnert W. *Radiology Review Manual*. 2nd ed. Baltimore: Williams & Wilkins. 1993; pp 360-361.
2. Paré JAP, Fraser RG. Pulmonary hypertension and edema. In: Paré JAP, Fraser RG. *Synopsis of Diseases of the Chest*. Philadelphia: WB Saunders Co. 1983; pp 482-492.
3. Eisenberg RL. *Atlas of Signs in Radiology*. Philadelphia: JB Lippincott. 1984; pp 224, 248.
4. Chapman S, Nakielny R. *Aids to Radiological Differential Diagnosis*. 3rd ed. London: Baillière Tindall. 1995; pp 126-143.