BILATERAL TUMOUR-FORMING PULMONARY AMYLOIDOSIS

AN UNUSUAL AND INTERESTING CASE

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A European male, aged 83 years, was admitted to the Uitenhage Provincial Hospital on 5 February 1959 suffering from developing gangrene of his left foot, which was found to be cold, discoloured and slightly oedematous.

He was a free patient, and the house surgeon in charge did the usual routine tests; except for a blood pressure of 180/110 mm.Hg and signs of emphysema, nothing was found in his urine, blood, heart, nervous system, and abdomen to suggest any serious disease. His clinical condition was described as fairly good.

On X-ray examination, however, a rare condition was found. The patient was questioned regarding his lungs and he stated emphatically that he did not remember ever having suffered from any complaint of his chest, but his house doctor stated that he had treated the patient some years ago for pleurisy; this was evidently correct because on postmortem examination there was no doubt that at some time during his life he had definitely suffered from pleurisy.

The hospital radiologist showed me the X-ray film and I decided to show it to the doctors in charge of the tuberculosis section. I was advised that whatever happened I had to retain the lungs at death. Arrangements were then made with the next-of-kin for a postmortem examination in the event of the patient's death.

The patient lived for about a year after this and died in Graaff-Reinet. His body was returned to Uitenhage. At the postmortem examination I found both lungs equally affected; hence I removed the right lung only. I also found that he had a pleural effusion and old pleuritic adhesions of both lungs. The heart appeared to be normal. I was informed that death was due to a stroke.

The case proved so interesting to me and others that it was suggested that I follow it up by sending the lung for a pathological report. This is submitted herewith, together with the X-ray report.

PATHOLOGICAL REPORT ON LUNG

Macroscopic Features

The lung shows the presence of chronic pleurisy. The lung substance is studded with multiple, various-sized, calcified irregular masses, and bears a superficial resemblance to a pneumolithiasis. The calcified masses vary from very small ones to masses several centimetres in diameter. They can be readily enucleated. On dissection they are found to be attached to the walls of the pulmonary veins and grow outwards in irregular positions compressing, but not invading, the contiguous lung substance. They have not been found in the lumina of vessels, nor attached to bronchi.

Chemistry

Chemical examination shows the bulk of the masses to consist of a protein matrix impregnated with calcium, magnesium, and sodium salts (carbonates and chlorides). Carbohydrate is present in traces — tests for fat are negative.

Histology

Histological examination shows that the masses are amorphous, irregular, hyaline masses which consist of *paramyloid* in which calcification has occurred. Tests for amyloid show a red metachromasia with gentian violet and a red colouration with Congo red.

Histological examination of several sections shows that the condition begins as an amyloid degeneration of the media of blood vessels, especially veins. Large collections of plasma cells are found in the vicinity of many of the lesions.

Diagnosis

Primary pulmonary tumour-forming amyloidosis.

RADIOLOGICAL REPORT

11 March 1959 — Chest (Fig. 1)

There are multiple irregular densities scattered throughout both lung fields. These opacities are extremely dense and there is evidence of involvement of some of the bronchi and possibly of some of the blood vessels in the lungs. There is surprisingly little distortion of the lung anatomy. These appearances are consistent with a diagnosis of pneumopathia osteoplastica racemosa in which bone formation occurs within the lung tissue and bronchial tube walls. Note also the wedging of the body of one of the lower dorsal vertebrae. This probably represents senile osteoporosis with wedge compression of the involved vertebra.

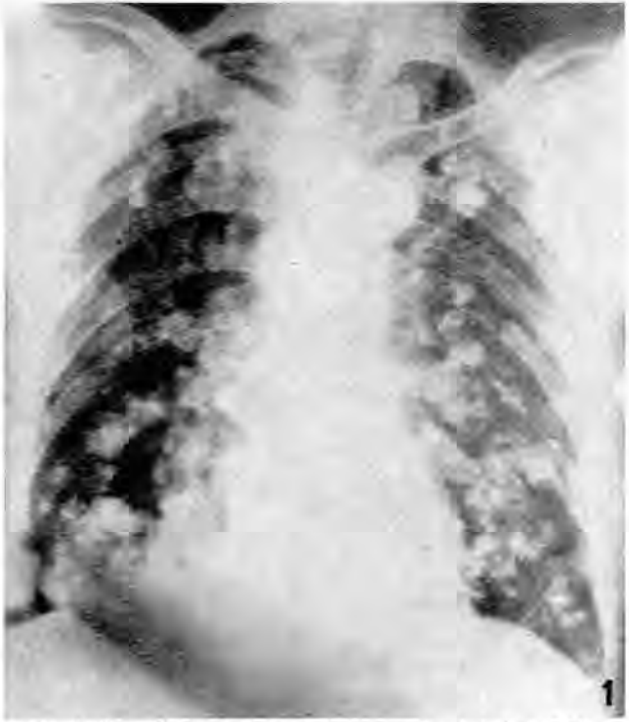

Fig. 1. Chest X-ray showing calcified masses in both lung fields.

DISCUSSION

Frank Bergman and Erland Linder[1] give the following information about amyloidosis:

'Incomplete knowledge of the aetiology and nature of amyloid degeneration makes classification of amyloid diseases difficult. Research has recognized three groups in addition to the secondary amyloidosis of chronic infectious diseases, namely (1) Primary atypical amyloidosis characterized among other things by the absence of any disposing basal disease; (2) nodular solitary or multiple local amyloidosis; and (3) amyloidosis, associated with multiple myeloma amyloid tumours, has been the subject of research for many years, especially in Germany where Leopold emphasized the value of publishing cases of localized amyloidosis and stressed the importance of descriptions of the tumours in the elucidation of problems bearing on amyloidosis.'

The factors considered of importance for the development of secondary amyloidosis can nearly always be excluded in nodular amyloidosis.

Another important difference is that amyloid tumours are seldom seen in parenchymatous organs. They show a liking for smooth muscle, skin, lymph nodes, and the mucous membrane of the digestive and urinary tracts. This form of amyloidosis is rarely seen in the lung tissue. As far as the lung itself is concerned, changes sometimes produce symptoms of pneumonia and pleurisy, but at other times no clinical signs are present and are only discovered incidentally by X-ray examination. It will be noticed from the above history that this was what occurred in the present case, when it was only by X-ray examination that the strange condition was discovered. This form of amyloidosis is rarely seen in the lung parenchyma, and a search in the literature failed to reveal reports of more than 13 cases.

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REFERENCE

1. Bergman, F. and Linder, E. (1958): J. Thorac. Surg., **35**, 628.