AMYLOID DISEASE—AN UNUSUAL CAUSE OF MEGALO-OESOPHAGUS*

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A megalo-oesophagus may be caused by numerous pathological conditions. However, the cause is not always easily determined. Often only a differential diagnosis can be presented, and then, in conjunction with the clinical syndrome, a tentative diagnosis suggested. Confirmation can only be obtained if biopsies are performed. The purpose of this presentation is to serve as a reminder that amyloidosis is usually a generalized disease, and that it is often associated with, or secondary to, other disease. An isolated form, though rare, is also described.

Amyloidosis, 'the widespread deposition of a homogeneous material throughout many organs of the body', was first described by Rokitansky in 1842. Virchow, in 1854, noted that, like starch, this material stained blue with iodine and sulphuric acid, and termed it amyloid. Actually it has no relation to starch, but is a protein of variable composition, usually associated with a sulphate-bearing polysaccharide, similar to if not identical with chondroiton sulphuric acid. Amyloid disease has been found to affect most organs of the body. The distribution of affected organs is shown in Fig. 1.

CASE REPORT

A Bantu woman, born in 1910, first presented at Groote Schuur Hospital in March 1964. She complained of a mass in the left groin and an ulcer on the left leg (venous stasis). The diagnosis of an irreducible inguinal hernia was made and she underwent operation on 20 March 1964, when only a large mass of lymph glands was found. This was removed, and examined histologically. These glands showed typical manifestations of amyloid disease.

The patient was a relatively fit and healthy person with no history or clinical evidence of chronic suppurative disease; this was therefore unlikely to be secondary amyloidosis. X-ray examination of the skeleton did not reveal any lytic lesions of bone, nor were any lesions evident in the surrounding soft tissues. However, the blood count and protein electrophoresis investigations showed a mild megaloblastic anaemia with 3 - 4% of plasmocytes and reversal of the albumin-globulin ratio, associated with the presence of myeloma proteins and also increased myeloid activity in the bone marrow. The blood picture was suggestive of multiple myeloma; but no abnormal plasmocytes could be found, nor were Benz-Jones proteins present in the urine.

The patient was discharged and followed-up as an outpatient. She had one recurrence of the groin mass which was again removed on 13 February 1967. Otherwise she kept relatively well except for mild disability caused by cardiac decompensation due to amyloid infiltration of the myocardium (non-specific changes compatible with amyloid infiltration of the myocardium were found on ECG).

In April 1968 she complained of retrosternal pain, nausea, vomiting and diarrhoea. These symptoms had started in September 1967, and had been intermittent since. She was therefore referred to the X-ray department

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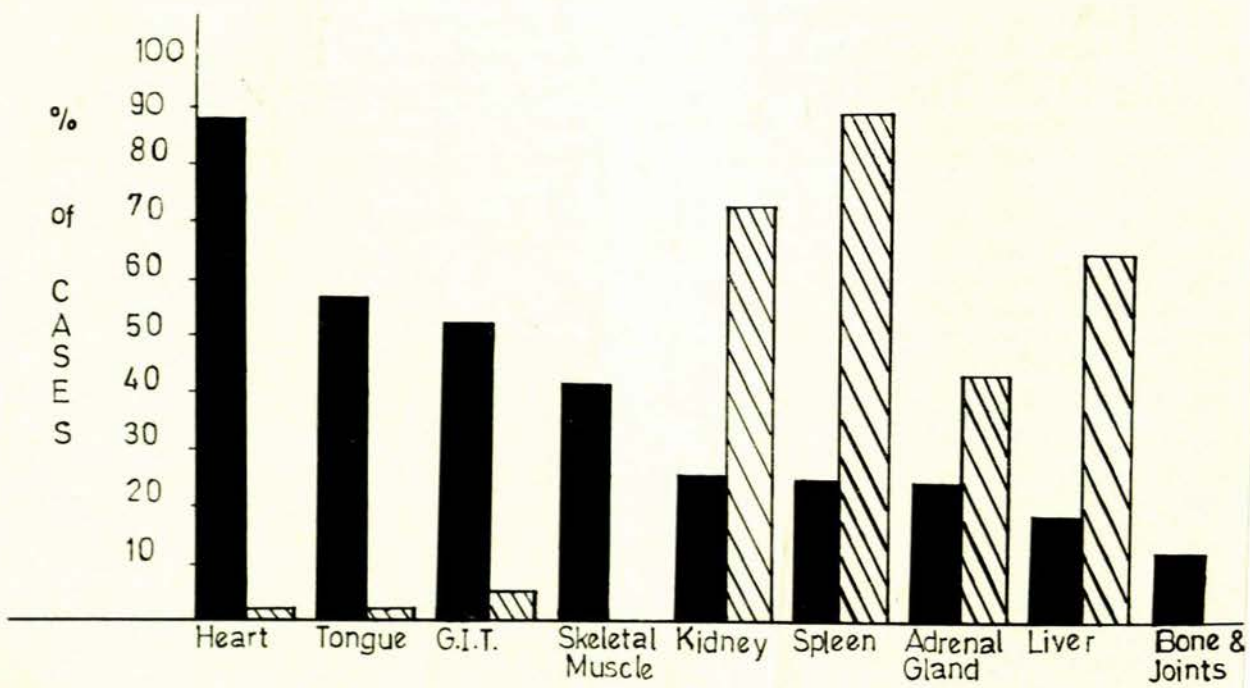

Fig. 1. The distribution of affected organs in primary and secondary amyloidosis is supportive evidence of the diagnosis of primary amyloid disease. ■ = distribution of affected organs in primary amyloidosis (Eisen, 46 cases). ▨ = distribution of affected organs in secondary amyloidosis (Rosenblatt, 110 cases).

for a barium swallow and meal. On radiological examination a very large and atonic oesophagus was demonstrated. There was also a suggestion of muscular inco-ordination at the initiation of deglutition, which could be the result of a pseudobulbar palsy. However, as she was known to have amyloid disease and did not have any other signs of bulbar palsy, this disease and other conditions such as scleroderma and senile degeneration could be reasonably left out of the differential diagnosis. Uniform enlargement of the small bowel was also found.

DISCUSSION

Although there have been few radiological studies of the oesophagus in primary amyloidosis,[3] these features correlate well with most findings described. Korelitz and Spindell[4] showed abnormal retention of barium in the valleculae and narrowing of the gullet. Hertzman and others[5] reported, 'The oesophagus appeared dilated and aperistaltic'. They also mentioned associated gastro-oesophageal reflux, which was not seen in this case. Toriola *et al.*[6] recorded a patient who required a bypass operation for amyloid disease of the oesophagus: the barium swallow showed dilation of the upper two-thirds of the oesophagus and a complete hold-up in the lower third.

SUMMARY

The absence of long-standing chronic illness, the multifocal nature and the failure to prove a definite association with a myelomatous state suggest that this is a case of primary amyloidosis involving (so far as is known) the heart, gastrointestinal tract (oesophagus and small intestine) and the lymph nodes in the groin.

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