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 chondroitin 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.

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).
DISCUSSION

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

SYNOPSIS

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.

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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REFERENCES


CLINICAL RESPONSE TO LONG-TERM PROPRANOLOL THERAPY IN HYPERTHYROIDISM


Propranolol has been shown to be effective in hyperthyroidism in a variety of situations. These include thyrotoxic crises, preparation for subtotal thyroidectomy, the control of residual thyrotoxic symptoms after radioactive iodine therapy and in some cases of thyrotoxic myopathy.

Standard antithyroid drugs are not without serious side-effects, and their prolonged use in hyperthyroidism may be complicated by the frequent need to adjust dosage to the changing clinical status of the patient. They also interfere with 131I tests of thyroid function during treatment. Propranolol, by contrast, causes few unpredictable side-effects if patients are appropriately selected, does not appear to induce hypothyroidism, and allows thyroid function to be tested during therapy. The present study was therefore undertaken to assess the effectiveness of this drug given as sole long-term treatment in hyperthyroidism.

PATIENTS AND METHODS

The trial group consisted of 27 patients (23 females and 4 males) attending the Thyroid Clinic, Groote Schuur Hospital, who were considered suitable candidates for long-term medical therapy and in whom there were no obvious contraindications to the use of propranolol, such as bronchospasm or overt cardiac failure. All were hyperthyroid by standard clinical and laboratory criteria. Their ages ranged from 21 to 63 years.

Propranolol (Inderal, ICI) was instituted as the sole therapeutic agent after assessment according to Wayne's therapy index; each patient had an initial score of 20 or more. The starting dose of propranolol was generally 40 mg. orally t.d.s. Patients were reassessed at monthly (or more frequent) intervals, and if clinical response was judged to be partial but inadequate the dose was increased (to a maximum of 120 mg. q.i.d.). Those who responded poorly were given Neo-Mercaptole after final clinical assessment for the trial.

After an average period of observation of 5 months (range 1-8 months), each subject was again carefully assessed clinically and a final therapy index score was assigned. Where possible, repeat radioactive iodine neck uptakes were also performed. On the basis of these 2 criteria, patients were then divided into 4 groups: those with an adequate clinical response (final therapy index score <5) and normal 131I studies (6- and 24-hour neck uptakes less than 40 and 50% respectively); those with an adequate response but elevated 131I uptakes; those with a partial clinical response (final therapy index score between 5 and 10)—all these had elevated 131I uptakes; and those with a poor clinical response (therapy index score >10).

RESULTS

Groups of Responders

As indicated in Table I, 47% of the series were considered to have had an adequate clinical response when finally reassessed. In all cases marked subjective and objective clinical improvement was seen within the first few weeks after commencement of therapy, and the dose of propranolol required was comparatively small. Indeed, just under half of these patients seemed to have become euthyroid again as indicated by normal repeat 131I neck uptakes. They could be differentiated from the patients who remained with elevated repeat uptakes, however, by their greater gain in weight.

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