

FAMILIAL MULTIPLE LIPOMATOSIS

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Abstract. A clinical study was made of 14 cases of multiple symmetrical lipomata in two families. There were 7 female and 7 male patients. In one family the members affected were observed in four generations. The disease set in during the third or fourth decade of life. The lipomata ranged in size from that of a pea to that of a hen's egg. They were limited to the forearms and trunk and were asymptomatic.

Key words: Lipomatosis; Hereditary diseases

Multiple symmetrical lipomatosis is a well-defined syndrome. It was first described by Brodie (8) in 1846. Lipomata are limited to the forearms and trunk. They usually appear in the third, fourth or fifth decade of life, rarely exceed 5 cm. in diameter and are generally asymptomatic.

Dietetic or environmental factors were considered to play an important role in the production of multiple lipomata. Hypercholesterolemia has been demonstrated in some patients (3, 7, 12, 17). Trauma (9, 19); endocrine disorders (4, 5, 20) and neurotrophic lesions (1, 22), have been considered important contributory factors.

Reports have appeared on the familial incidence of multiple lipomas (2, 6, 13, 14, 15, 16, 18, 21). The condition, however, is extremely rare (10, 16, 21). A survey of the literature shows that lipomatosis has been found about twice as often in males as in females. Touraine (24) found that the condition was transmitted as a dominant trait in 32 families out of 37. In 16 families it was sex-linked and limited to males.

Our observation of the occurrence of multiple lipomata in two families is worthy of report: so many members are affected that it is believed that genetic factors are undoubtedly involved.

MATERIALS AND METHODS

Two families with a total of 14 manifest cases of multiple lipomatosis were investigated (Figs. 1 and 2). A complete family analysis was carried out; all surviving members of these families were examined. In the majority of cases blood cholesterol and glucose levels were measured.

RESULTS

Clinical data

There were 7 females and 7 male subjects. In most of the cases the disorder appeared in the third or fourth decade of life.

Signs

The tumours were multiple and symmetrical. They ranged in size from that of a pea to that of a hen's egg. They were scattered over the trunk and forearms. In 2 patients a few tumours were excised and were demonstrated to be fibrolipomas.

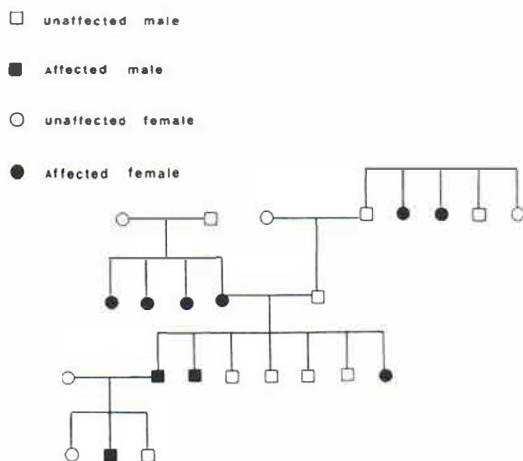


Fig. 1. Pedigree of family 1.

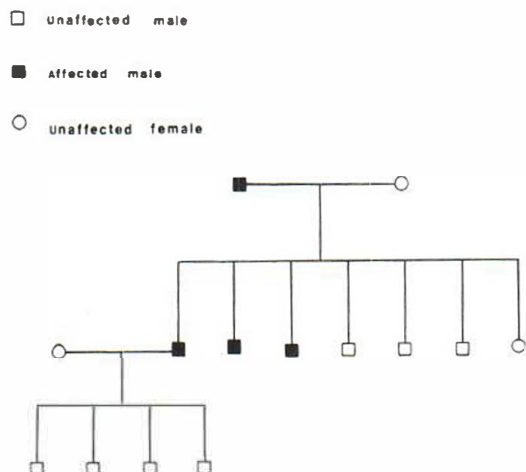


Fig. 2. Pedigree of family 2.

Symptoms

The tumours were asymptomatic, though some degree of anxiety or depression was present in 3 male and 4 female patients. Two subjects complained of pain due to cervical arthrosis.

The tumours grew slowly until a certain size was reached, after which they remained stationary indefinitely.

Laboratory findings

Only one patient was found to have diabetes. The other members of the two families definitely had no evidence of diabetes or hypercholesterolemia.

COMMENT

Family no. 1 appears to show autosomal recessive inheritance, the ratio of affected female to males being 7 to 3.

Some investigators (11, 16, 24) on the contrary, have suggested that the condition may be due to an autosomal dominant gene and that twice as many males as females are therefore affected. A critical scrutiny of the literature does not establish the mode of inheritance conclusively. In our opinion, however, the condition is not related to any single gene, but is probably polygenic. The role of exogenous, metabolic and emotional factors is difficult to assess, though it must be taken into account in order to explain the development of lipomas in adult years.

According to Wersheimer & Shapiro (25) the autonomic nervous system plays a role in the control of the lipid metabolism. The sympathetic system reduces the lipid synthesis, whereas the parasympathetic system increases lipid deposition in tissues. Sahara & Okinaka (23) were able to induce experimental lipomatosis in animals after abdominal sympathicectomy followed by pilocarpine and acetylcholine injections.

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